

Original article

Cross Effect of Neurofibromatosis type 1

by

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From toshichan-man hospital

I statistically analyzed 27 patients with neurofibromatosis type 1.

Since the 1930's, it has been said that the neurofibromatosis type 1 born to affected mothers is very serious. It is called "maternal effect". But many are against the opinion. The result of my examination is as follows.

The male cases born to affected mothers or the female cases born to affected fathers have extremely possibility of having malignant tumors.

The male cases born to affected fathers or the female cases born to affected mathers or those who were new mutations didn't have malignant tumors. They were all benign.

"When the tumor is found in the male cases born to affected mothers or female cases born to affected fathers, it is extremely possible that the tumor is malignant."

I newly name it "cross effect".

【key words】 neurofibromatosis type 1, cross effect, maternal effect, malignant tumor

【firstly】

When a male patient of the neurofibroma type 1 more derived from mother than the 1930s was seriously ill, and a tumor was found, it has been said that the probability that was a malignant tumor was very high as for it. When a male patient of the neurofibroma type 1 derived from mother was seriously ill, and a tumor was surely found when the writer took the statistics of 27 neurofibroma type 1, it became clear that malignant probability

was very high as for it. When a female patient of the neurofibroma type 1 derived from father was seriously ill, and a tumor was found in the same way, it became clear that the probability that it was malignant was very high. It became clear that the malignant thing might not have a female patient derived from mother, a male patient derived from father and proband even if a tumor was caught in the neurofibroma type 1 of the patient. However, this is consideration about 27 that the writer collected and needs a following supplementary examination.

[case]

[example as follows derived from mother for men]

<< case 1 >> Mother origin, man, 38 years old

There are a few neurofibromas, but the occipital bone has a bone defect. It is healthy by nature. An iris nodule (+).

In about last September, nystagmus appears at the time of fatigue. It is October, and nystagmus becomes strong. I have a tumor of the hemisphere of cerebellum pointed out in head CT. The craniotomy tumor enucleation enforcement. It was a pleomorphic glioblastoma pathologically. The abnormality is not discovered by the examination of chromosome.

It was two brothers, and there was an older brother of the neurofibroma type 1, but I slept, but it was discovered in futon what died in the evening after the older brother learned strong fatigue at noon, and he came back from the workshop in 43 years. The cause of death was settled concerning heart failure. It was healthy, and this older brother has not had heart problem pointed out by nature, too. The dissection to check the cause of death was not performed. This older brother is thought to have been more likely to be a tumor of cerebellum or the brainstem, too.

<< case 2 >> Mother origin, man, 24 years old

I recognize café au lait spot on the back, the buttocks from time for childhood. An iris nodule (+).

It takes effect, and, on October 2, 1984, there is no head CT in the views that should mention specially. There are no views that should mention specially in the brain waves. A flexible mass (-).

The older brother who is 2 years old older intelligence quotient 32 (sb type). After being going to wake you up in the morning, during House of nursing hospitalization, he died in 12 years. As a result of dissection, it was brainstem malignant neurologic sheath tumor.

<< case 3 >> Mother origin, man, 19 years old

It enforces four times of tumor enucleation of the left orbit bottom at time for childhood. An iris nodule (+).

On November 13, 1987, I extract a right clavicle superior fovea tumor. It was a malignant star cell tumor pathologically.

On December 7, 1987, I extract a right elbow joint part tumor. Similarly it was a malignant star cell tumor.

On January 10, 1988, I extract left orbit bottom tumor. This was a malignant star cell tumor in the same way, too.

I introduce a university hospital in the reposition purpose of the brain lump with the loss of the left orbit wing this time.

<< case 4 >> Mother origin, man, 33 years old

Mother has several things like the small cafe au lait spot (it is said that she appeared from 40 seasons and years).

There is one older sister, but I am operated on by dislocation of the hip joint and take effect.

There is a tumor to the right hemisphere of cerebellum. An iris nodule (+). It is a bone defect to the right suboccipital region. I protrude the right back of the head. The left side is approximately totally blind. It was a malignant star cell tumor (grade 3) by a craniotomy operation pathologically.

<< case 5 >> Mother origin, man, 7 years old

Five countrymen. Three older sisters are healthy. The older brother switches over from pneumonia to meningitis in 1 year and dies.

A congenital aqueduct of midbrain obstruction is unidentified. An iris nodule (+). Is both sides eyesight drop; the eyesight drop is a tendency to progress.

<< case 6 >> Mother origin, man, 1 year old

Mind campaign development delay, congenital heart disease, an iris nodule (+). It is a lot of cafe au lait spot to a lower part of the body after life right more. In head CT brain atrophy (+), a tumor (-).

Two countrymen. As for the older sister, spotted dark brown on the left buttocks (4*4cm).

<< case 7 >> Mother origin, man, 14 years old

There is more left fibula false joint than the young time. An iris nodule (+). In 14 years, appeal for dizziness; the next House. In head CT, there is a tumor to the left hemisphere of cerebellum. Craniotomy tumor enucleation takes effect in a university hospital. It was a pleomorphic glioblastoma pathologically.

<< case 8 >> Mother origin, man, 11 years old

Because there are nausea, a headache from approximately two years ago, I consult our House. In head CT, there is tumor which seems to be a star cell tumor to left both sides of the head parietal area. Size increases, and tumor introduces a university hospital. The craniotomy operation enforcement. It was a malignant star cell tumor pathologically. An iris nodule (+).

There is cafe au lait spot for a little younger sister in three brothers (two younger sisters) immediately.

<< case 9 >> Mother origin, man, 23 years old

It is cafe au lait spot more-prone to a whole body than straight bottom time. There is the back in a flexible mass subcutaneously. I had a diagnosis of neurofibroma type 1 by biopsy of the last year. An iris nodule (+).

In 23 years, it is noticed the existence of the brain tumor in head CT. The volume of tumor is a tendency to increase. The plan of the craniotomy operation.

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[example as follows derived from mother for women]

<< case 10 >> mother origin, woman, 23 years old

Three women, Sueko of the one man. Two older sisters are healthy.

A right-sided auditory nerve tumor. It is a benign tumor pathologically. Both sides blindness. Sense of smell suffers a loss with right and left.

The older brother dies of a spinal cord tumor at 15 years old (as for this older brother, type 1 a neurofibroma).

<< case 11 >> mother origin, woman, 38 years old

Cerebral infantile paralysis, neurogenic bladder.

I came by walking alone until junior high school time. Low back pain develops and, in 37 years, becomes both lower limbs paralysis. In CT, there is tumor shadow to a lumbar part. It is a benign tumor pathologically.

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[example as follows derived from father for men]

<< case 12 >> father origin, man, 27 years old

Though the human trunk has a flexible mass, in a junior high student, I notice. I

gradually write 増数 and consult our House nine years ago and have a diagnosis of neurofibroma type 1 afterwards. An older sister dies by asthmatical bronchitis.

There is no abnormality in head CT. There is a cauda equina nerve tumor. Cauda equina nerve tumor was a benign tumor pathologically.

<< case 13 >> father origin, man, 32 years old

There are three countrymen (all man), absence attacks.

In head CT, there is tumor shadow on the right head. Craniotomy was a benign star cell tumor though I was operated on.

<< case 14 >> father origin, man, 31 years old

Left pulsatility drawing of the eyes with marking dots, left orbit bottom bone defect.

<< case 15 >> father origin, man, 18 years old

On back of the head midline a bone defect (3.5*3.5cm). From the young time a speech impediment.

It is the next House for a medical examination for Self-Defense Forces enlistment.

<< case 16 >> father origin, man, 2 years old

Right sella turcica expansion (J type). The expansion of the ear canal in the right. The elevation of the right wing-like scale.

The expansion of the right eye nerve way. Expansion of the orbit.

<< case 17 >> father origin, man, 11 months

There is the right cranial bones loss. There is not the neurologic loss. There is a tumor to cervical vertebrae. It is a benign tumor pathologically.

<< case 18 >> father origin, man, 10 years old

The cafe au lait spot of 10 several appears to a human trunk, limbs from one month after birth. There are not increase of the number of cafe au lait spot and the expansion of the size. The neurofibroma developed in the whole back mainly on hips for one year, but six elasticity tumors which began to resolve in the summer from the time (natural regression), and slightly rose to hips at the time of this House outpatient department first medical examination on (September 5, 1992) existed. Almost none of the parts could confirm it at the time of hospitalization on (December 16, 1992), too and resolved.

I am hospitalized for the inspection of the neurofibroma this time. In head CT, there are no views that should mention specially.

Cafe au lait is spotted for father. The countryman has an older sister in two people, but there is no abnormality now.

[example as follows derived from father for women]

<< case 19 >> father origin, woman, 34 years old

It was noticed café au lait spot from the small time. However, there was a little café au lait spot, and there were not most of the flexible masses. An iris nodule (+). Appeal for a headache; a senior Pope next House. The bone defect was not clear, but there is a tumor to the left hemisphere of cerebellum. A university hospital introduction. Craniotomy was operated on, but a pathologically malignant star cell tumor. After an operation, he dies.

<< case 20 >> father origin, woman, 21 years old

The right parietal area, a right back of the head ache develop from about October, 1992. When a pain is strong, there are feeling like vomiting, a radiation ache to the right shoulder. I do not recognize it in head CT abnormally. A neurofibroma of the right cervical vertebrae transverse process is diagnosed. An iris nodule (+).

In 1992, I have star cell tumor doubt pointed out by left both sides of the head parietal area in head CT. Size increases, and tumor enforces a craniotomy operation. A pathologically malignant star cell tumor.

<< case 21 >> father origin, woman, 28 years old

There was café au lait spot from time for childhood and hemispheric flexible masses occurred frequently more systemically than 16 years old and came to increase. An iris nodule (+). Right ear mixture characteristics hearing loss.

There is a stenosis in rectum. However, tumor marker (-).

There is the flexura which is a whit to lower part thoracic vertebra. There is no abnormality in blood, biochemistry.

I recognize calcification and low-attenuation foci to hemisphere of cerebellum in head CT. With contrast media a reinforcement effect (-), a cerebellum symptom (-). I have a diagnosis of cerebellum atrophy or arachnoid cyst and become the follow-up without the need of the operation.

Nystagmus appears and, in head CT, has a tumor pointed out by hemisphere of cerebellum, and a craniotomy operation takes effect recently. A pathologically pleomorphic glioblastoma.

<< case 22 >> father origin, woman, 16 years old

A countryman has two older brothers, younger brother one, but café au lait spot or the flexible mass are not detected. I detected large-scale café au lait spot and the elasticity mass of the part from the back to the buttocks after life more immediately. An iris nodule (+). I operate for a flexible mass in S General Hospital plastic surgery. It is the next House

by looking at the both eyes, and having dripped down from 2.0 in 0.9 recently. In head CT, there is a tumor to occipital lobe. I look at the both eyes more and fall in 0.1 recently. I introduce me and am operated on, and it is taken effect to a university hospital. A pathologically malignant star cell tumor.

<< case 23 >> father origin, woman, 13 years old

There is an epilepsy attack to a younger brother. An older sister until about 7 years old febrile convulsion (do abnormally now). Purpura (I do abnormally now) allergic as for the older brother.

There is almost café au lait spot to a whole body. A number and the area are tendencies to increase. The flexible mass is recently, too

"Case 23" Father-derived, female, 13 years old

My younger brother has epileptic seizures. My older sister is fever seizures (now no abnormality) until about 7 years old. My older brother has allergic purpura (now there is no abnormality).

Most of the body has caffè ore spots. The number and area tend to increase. Recently, a soft mass begins to appear. Iris nodule (+).

I have a stomachache since March this year. There are 2 clots of bloody stools in June.

Laparotomy surgery is enforced. Pathologically, it was a colon tumor with a high degree of malignancy.

No particular noticeable findings on head CT.

[The following is unknown origin. That is, the case itself is a proband male]

Unknown from "Case 24", Male, 57 years old

Café aule plaque and soft tumor mass are remarkable. At the age of 30, a lumbar soft mass reconstruction was performed.

We have undergone surgery 12 years ago, 11 years ago, 8 years ago under diagnosis of hydronephrosis in bilateral kidney 12 years ago. Pathologically, benign hamartoma.

Strong left side vision disorder continues from 10 years ago, but it has not progressed.

No abnormality was found by chromosome examination.

August last year, nystagmus, appearance. Similar nystagmus, emergence in September (however, nystagmus only occurred once in August, September). It is pointed out suspected astrocytoma which is performed partly on the right side parietal region by MRI. Suspected astrocytoma in the right parietal region was excised by craniotomy but the part considered to be a tumor that was imaged by MRI was a benign tumor with no vascular hyperplasia and neuroglial tumorigenesis.

Unknown from "Case 25", Male, 39 years old

There is a cafe au lait on the left buttocks at birth. Emergence of soft tumor from 4 to 5 years ago. Two years ago I noticed a hard, marginally clear, movable hardening with a diameter of 8 mm on the left third intercostal sternum left edge. The soft mass of the left buttocks is (7 × 4 cm) large soft-type, pigmented. There is abnormal electroencephalogram in the back of the head. There is abnormal shadow in the basal ganglion of the cerebrum, on the head CT.

Unknown from "Case 26", Male, 21 years old

Sister is SLE. When I was in elementary school, I noticed brown spots on the upper right lip. It was a cafe au lait. In case

When a junior high school student, the abdomen, notice the light brown spots of both upper limbs. In July 1985, a brain tumor was found in the head CT, and the tumor was excised.

Pathologically, the tumor was benign.

In case

Unknown from "Case 27", Male, 14 years old

At the age of 5, emergence of soft tumor in upper left arm. A soft mass exists also in the right neck, right wrist. Brown spot on front chest from early childhood. It is diagnosed as cafe aule spot in dermatology.

At the age of 13, left upper arm soft tissue mass extraction.

At the age of 15, surgery was performed on the extramedullary tumor of the third cervical vertebra and fifth cervical vertebra. Pathologically, it was a benign tumor. There is also cerebral sinus meningioma (fibrous).

[The following is unknown origin. That is, the case itself is a proband woman]

1 case none

【Summary of cases】

[Example of male from mother]

Case 1 died of glioblastoma multiforme (38 years old), older brother died (43 years old) in the tumor (suspicion) of the brain stem

In case 2, the elder brother died of brain stem malignant schwannoma (12 years old), the patient himself was alive (24 years old) on the head CT, tumor (-)

Case 3 is malignant astrocytoma (19 years old)

Case 4 is a malignant astrocytoma (grade 3) (33 years old), although one has an older sister, he undergoes surgery with hip joint dislocation.

In case 5, older brother dies after transitioning from pneumonia to meningitis at 1 year of age. The patient himself is congenital midbrain water obstruction obstruction, vision deterioration

tends to progress (7 years old), three older sisters health

Case 6 is delayed psychomotor development, congenital heart disease (1 year old)

Case 7 is glioblastoma multiforme (14 years old)

Case 8 is malignant astrocytoma (11 years old)

Case 9 is an increasing trend of brain tumor (23 years old)

[Example of female from mother]

Case 10 is right auditory nerve tumor (benign) (23 years old), older brother died of spinal cord tumor at age 15

Case 11 shows a soft tumor (38 years old)

[Example of male from father]

Case 12 is a cauda equina neoplasm (benign) (27 years old), older sister died of childhood asthma

Case 13 is astrocytoma (benign) (32 years old)

Case 14 was left orbital bottom bone defect and left pulsating eyestrain (31 years old)

Case 15 was bone loss (3.5 x 3.5 cm) on the medial line of the occipital region, language disorder (18 years old) from young age,

Case 16 shows right Turkish saddle expansion (J type), enlargement of right middle ear canal, elevation of right winged scales (2 years old)

Case 17 was tumor (benign) in the cervical spine (11 months)

In case 18, neurofibroma appeared all around the back centering around the lumbar region from 1 year ago but it disappeared (spontaneous withdrawal) (10 years old) from around the summer

In case

[Examples of female from father]

Case 19 died of malignant astrocytoma (34 years old)

Case 20 was malignant astrocytoma with surgery (21 years old)

Case 21 is surgery (28 years old) with glioblastoma multiforme

Case 22 was malignant astrocytoma with surgery (16 years old)

Case 23 is a colorectal tumor with a high degree of malignancy (13 years old)

[Cases themselves are male cases of probands]

Case 24 is a benign brain tumor (57 years old)

Case 25 shows abnormal shadow (39 years old) in basal ganglia of the cerebrum,

Case 26 is a benign brain tumor (21 years old)

Case 27 is an extramedullary tumor (benign) of the third cervical vertebra and fifth cervical vertebra (14 years old)

[Case itself is a female example of a proband]

I could not find one case

【Discussion】

There are few references in which origin is clearly described in brain tumors of type 1 neurofibroma. There is a literature of a malignant brain tumor in which the origin was specified, but it was a case of multiple malignant schwannoma and was derived from a mother, and the patient was a male. In case

In addition, there is a case in which another origin is clearly stated, which is a document that suggested spontaneous resolution of the orbital glioblastoma multiforme of a 2 - year - old father-born girl [5].

When examining 27 cases, neurofibroma 1 type of a male-derived male patient tends to be malignant. And female patients from fathers also have a strong tendency to malignant to the same extent.

Mother-derived effects in neurofibroma 1 type, that is, maternal effect (6, 7, 8, 10, 11, 13) have been conventionally mentioned. However, there are also many objections (1, 2, 3, 12, 16), and unification of opinion has not been made. I propose a new cross effect, here a cross effect. Mother-derived effects exist reliably in this survey. It is strongly related whether neurofibroma type 1 is a father-derived female patient, a mother-derived male patient, or another case for canceration. As described above, malignant tumors are highly likely to develop in male patients and father-derived female patients in neurofibroma 1 type, and mother-derived female patients, male-derived male patients, and patients themselves are probands I believe that the absence of malignant tumors will give great implications for cancer gene therapy.

In case

【Reference】

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----- Summary -----

Cross effect of neurofibroma 1 type

Statistical analysis of 27 neurofibroma 1 types was performed.

Patients with mother-derived neurofibroma 1 type have been said to be more severe than in the past. This is called maternal effect. However, there are many objections. As a result of my investigation, it is very likely that a tumor is malignant in a mother-derived male patient or a father-derived female patient, and if a mother-derived female patient, a father-derived female patient, or a case where the mother is a proband It was never malignant, all were benign.

"When a tumor is found in a mother-derived male patient, a father-derived female patient, the possibility that it is malignant is extremely high." This is called a new cross effect.

Someone please translate it into English. You can post to a foreign medical journal. It is useless in Japan. I am not good at English very much. Thanking you in advance.

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(Telephone is useless, thank you for your consideration.)

<< case 23 >> father origin, woman, 13 years old

There is an epilepsy attack to a younger brother. An older sister until about 7 years old febrile convulsion (do abnormally now). Purpura (I do abnormally now) allergic as for the older brother.

There is almost cafe au lait spot to a whole body. A number and the area are tendencies to increase. The flexible mass begins to develop recently, too. An iris nodule (+).

There is more abdominal pain than this March. In June, there is blood in stool twice. The open surgery enforcement. It was strong large intestine tumor of the malignancy pathologically.

In head CT, there are no views that should mention specially.

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[missing an origin as follows. In other words, case oneself an example of the proband for men]

<< case 24 >> origin ignorance, man, 57 years old

As for the cafe au lait spot and the flexible mass, it is significant. In 30 years, it enforces hips elasticity mass resection.

I operate on both sides kidney under the diagnosis of the hydronephrosis with eight years ago 11 years ago 12 years ago from 12 years ago. A blunder tumor benign pathologically.

A strong left-sided visual disorder continues from ten years ago, but does not progress.

The abnormality is not discovered by an examination of chromosome.

In last August, it is appeared nystagmus. Nystagmus, an appearance (however, there was the nystagmus by 9 once a month in August) that are similar in September. I have the star cell

tumor doubt that MRI takes effect, and is contrasted partly by the right parietal area pointed out. I extracted the right calvarial star cell tumor doubt by craniotomy, but there was a blood vessel hyperplasia, and there was not malignant transformation of the glia, and the part which seemed to be a tumor contrasted by MRI was a benign tumor.

<< case 25 >> origin ignorance, man, 39 years old

There is more cafe au lait spot on the left buttocks than birth. A mass appearance more flexible than before four or five years. From two years ago on the left sternal border between the left third rib of 8mm in diameter a border is clear, and notice movable induration firmly. There are (7*4cm) great weak bunch type, pigmentation in the flexible mass of the left buttocks. There are abnormal brain waves at the back of the head. In head CT, there is abnormal shadow in the basal nuclei part.

<< case 26 >> origin ignorance, man, 21 years old

A younger sister is SLE. Though the right Uekuchi labial part has brown spot, in a primary schoolchild, I notice. It was cafe au lait spot.

In a junior high student, I notice the abdomen, ecru spot of both arms. In July, 1984, a brain tumor is found in head CT, and tumor enucleation takes effect. Tumor was benign pathologically.

<< case 27 >> origin ignorance, man, 14 years old

5 seasons and years, left upper arm part elasticity mass appearance. There is the flexible mass on the right neck, the right wrist. There is brown spot in the precordial region after childhood. I have a diagnosis of cafe au lait spot with dermatology.

13 seasons and years, left upper arm part elasticity mass extraction.

I am operated on for a marrow outside-related tumor of third cervical vertebrae, the fifth cervical vertebrae and, in 15 years, take effect. It was a benign tumor pathologically. In addition, there is a brain sickle part meningioma (fibrousness).

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One, too and

Original article

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by

Toshiro Takami, M.D.

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Since the 1930's, it has been said that the neurofibromatosis type 1 born to affected mothers is very serious. It is called "maternal effect". But many are against the opinion. The result of my examination is as follows.

The male cases born to affected mothers or the female cases born to affected fathers have extremely possibility of having malignant tumors.

The male cases born to affected fathers or the female cases born to affected mothers or those who were new mutations didn't have malignant tumors. They were all benign.

"When the tumor is found in the male cases born to affected mothers or female cases born to affected fathers, it is extremely possible that the tumor is malignant."

I newly name it "cross effect".

【key words】 : neurofibromatosis type 1, cross effect, maternal effect, malignant tumor

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When a male patient of the neurofibroma type 1 more derived from mother than the 1930s was seriously ill, and a tumor was found, it has been said that the probability that was a malignant tumor was very high as for it. When a male patient of the neurofibroma type 1 derived from mother was seriously ill, and a tumor was surely found when the writer took the statistics of 27 neurofibroma type 1, it became clear that malignant probability was very high as for it. When a female patient of the neurofibroma type 1 derived from father was seriously ill, and a tumor was found in the same way, it became clear that the probability that it was malignant was very high. It became clear that the malignant thing might not have a female patient derived from mother, a male patient derived from father and proband even if a tumor was caught in the neurofibroma type 1 of the patient. However, this is consideration about 27 that the writer collected and needs a following supplementary examination.

[case]

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It was two brothers, and there was an older brother of the neurofibroma type 1, but I slept, but it was discovered in futon what died in the evening after the older brother learned strong fatigue at noon, and he came back from the workshop in 43 years. The cause of death was settled concerning heart failure. It was healthy, and this older brother has not had heart problem pointed out by nature, too. The dissection to check the cause of death was not performed. This older brother is thought to have been more likely to be a tumor of cerebellum or the brainstem, too.

<< case 2 >> Mother origin, man, 24 years old

I recognize cafe au lait spot on the back, the buttocks from time for childhood. An iris nodule (+).

It takes effect, and, on October 2, 1984, there is no head CT in the views that should mention specially. There are no views that should mention specially in the brain waves. A flexible mass (-).

The older brother who is 2 years old older intelligence quotient 32 (sb type). After being going to wake you up in the morning, during House of nursing hospitalization, he died in 12 years. As a result of dissection, it was brainstem malignant neurologic sheath tumor.

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Mother has several things like the small cafe au lait spot (it is said that she appeared from 40 seasons and years).

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Five countrymen. Three older sisters are healthy. The older brother switches over from pneumonia to meningitis in 1 year and dies.

A congenital aqueduct of midbrain obstruction is unidentified. An iris nodule (+). Is both sides eyesight drop; the eyesight drop is a tendency to progress.

<< case 6 >> Mother origin, man, 1 year old

Mind campaign development delay, congenital heart disease, an iris nodule (+). It is a lot of cafe au lait spot to a lower part of the body after life right more. In head CT brain atrophy (+), a tumor (-).

Two countrymen. As for the older sister, spotted dark brown on the left buttocks (4*4cm).

<< case 7 >> Mother origin, man, 14 years old

There is more left fibula false joint than the young time. An iris nodule (+). In 14 years, appeal for dizziness; the next House. In head CT, there is a tumor to the left hemisphere of cerebellum. Craniotomy tumor enucleation takes effect in a university hospital. It was a pleomorphic glioblastoma pathologically.

<< case 8 >> Mother origin, man, 11 years old

Because there are nausea, a headache from approximately two years ago, I consult our House. In head CT, there is tumor which seems to be a star cell tumor to left both sides of the head parietal area. Size increases, and tumor introduces a university hospital. The craniotomy operation enforcement. It was a malignant star cell tumor pathologically. An iris nodule (+).

There is cafe au lait spot for a little younger sister in three brothers (two younger sisters) immediately.

<< case 9 >> Mother origin, man, 23 years old

It is cafe au lait spot more-prone to a whole body than straight bottom time. There is the back in a flexible mass subcutaneously. I had a diagnosis of neurofibroma type 1 by biopsy of the last year. An iris nodule (+).

In 23 years, it is noticed the existence of the brain tumor in head CT. The volume of tumor is a tendency to increase. The plan of the craniotomy operation.

[example as follows derived from mother for women]

<< case 10 >> mother origin, woman, 23 years old

Three women, Sueko of the one man. Two older sisters are healthy.

A right-sided auditory nerve tumor. It is a benign tumor pathologically. Both sides blindness. Sense of smell suffers a loss with right and left.

The older brother dies of a spinal cord tumor at 15 years old (as for this older brother, type 1 a neurofibroma).

<< case 11 >> mother origin, woman, 38 years old

Cerebral infantile paralysis, neurogenic bladder.

I came by walking alone until junior high school time. Low back pain develops and, in 37 years, becomes both lower limbs paralysis. In CT, there is tumor shadow to a lumbar part. It is a benign tumor pathologically.

[example as follows derived from father for men]

<< case 12 >> father origin, man, 27 years old

Though the human trunk has a flexible mass, in a junior high student, I notice. I gradually write 増数 and consult our House nine years ago and have a diagnosis of neurofibroma type 1 afterwards. An older sister dies by asthmatical bronchitis.

There is no abnormality in head CT. There is a cauda equina nerve tumor. Cauda equina nerve tumor was a benign tumor pathologically.

<< case 13 >> father origin, man, 32 years old

There are three countrymen (all man), absence attacks.

In head CT, there is tumor shadow on the right head. Craniotomy was a benign star cell tumor though I was operated on.

<< case 14 >> father origin, man, 31 years old

Left pulsatility drawing of the eyes with marking dots, left orbit bottom bone defect.

<< case 15 >> father origin, man, 18 years old

On back of the head midline a bone defect (3.5*3.5cm). From the young time a speech impediment.

It is the next House for a medical examination for Self-Defense Forces enlistment.

<< case 16 >> father origin, man, 2 years old

Right sella turcica expansion (J type). The expansion of the ear canal in the right. The elevation of the right wing-like scale.

The expansion of the right eye nerve way. Expansion of the orbit.

<< case 17 >> father origin, man, 11 months

There is the right cranial bones loss. There is not the neurologic loss. There is a tumor to cervical vertebrae. It is a benign tumor pathologically.

<< case 18 >> father origin, man, 10 years old

The cafe au lait spot of 10 several appears to a human trunk, limbs from one month after birth. There are not increase of the number of cafe au lait spot and the expansion of the size. The neurofibroma developed in the whole back mainly on hips for one year, but six elasticity tumors which began to resolve in the summer from the time (natural regression), and slightly rose to hips at the time of this House outpatient department first medical examination on (September 5, 1992) existed. Almost none of the parts could confirm it at the time of hospitalization on (December 16, 1992), too and resolved.

I am hospitalized for the inspection of the neurofibroma this time. In head CT, there are no views that should mention specially.

Cafe au lait is spotted for father. The countryman has an older sister in two people, but there is no abnormality now.

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[example as follows derived from father for women]

<< case 19 >> father origin, woman, 34 years old

It was noticed cafe au lait spot from the small time. However, there was a little cafe au lait spot, and there were not most of the flexible masses. An iris nodule (+). Appeal for a headache; a senior Pope next House. The bone defect was not clear, but there is a tumor to the left hemisphere of cerebellum. A university hospital introduction. Craniotomy was operated on, but a pathologically malignant star cell tumor. After an operation, he dies.

<< case 20 >> father origin, woman, 21 years old

The right parietal area, a right back of the head ache develop from about October, 1992. When a pain is strong, there are feeling like vomiting, a radiation ache to the right shoulder. I do not recognize it in head CT abnormally. A neurofibroma of the right cervical vertebrae transverse process is diagnosed. An iris nodule (+).

In 1992, I have star cell tumor doubt pointed out by left both sides of the head parietal area in head CT. Size increases, and tumor enforces a craniotomy operation. A pathologically malignant star cell tumor.

<< case 21 >> father origin, woman, 28 years old

There was cafe au lait spot from time for childhood and hemispheric flexible masses occurred frequently more systemically than 16 years old and came to increase. An iris nodule (+). Right ear mixture characteristics hearing loss.

There is a stenosis in rectum. However, tumor marker (-).

There is the flexura which is a whit to lower part thoracic vertebra. There is no abnormality in blood, biochemistry.

I recognize calcification and low-attenuation foci to hemisphere of cerebellum in head CT. With contrast media a reinforcement effect (-), a cerebellum symptom (-). I have a diagnosis of cerebellum atrophy or arachnoid cyst and become the follow-up without the need of the operation.

Nystagmus appears and, in head CT, has a tumor pointed out by hemisphere of cerebellum, and a craniotomy operation takes effect recently. A pathologically pleomorphic glioblastoma.

<< case 22 >> father origin, woman, 16 years old

A countryman has two older brothers, younger brother one, but cafe au lait spot or the flexible mass are not detected. I detected large-scale cafe au lait spot and the elasticity mass of the part from the back to the buttocks after life more immediately. An iris nodule (+). I operate for a flexible mass in S General Hospital plastic surgery. It is the next House by looking at the both eyes, and having dripped down from 2.0 in 0.9 recently. In head CT, there is a tumor to occipital lobe. I look at the both eyes more and fall in 0.1 recently. I introduce me and am operated on, and it is taken effect to a university hospital. A pathologically malignant star cell tumor.

<< case 23 >> father origin, woman, 13 years old

There is an epilepsy attack to a younger brother. An older sister until about 7 years old febrile convulsion (do abnormally now). Purpura (I do abnormally now) allergic as for the older brother.

There is almost cafe au lait spot to a whole body. A number and the area are tendencies to increase. The flexible mass begins to develop recently, too. An iris nodule (+).

There is more abdominal pain than this March. In June, there is blood in stool twice. The open surgery enforcement. It was strong large intestine tumor of the malignancy pathologically.

In head CT, there are no views that should mention specially.

<< case 24 >> origin ignorance, man, 57 years old

As for the cafe au lait spot and the flexible mass, it is significant. In 30 years, it enforces hips elasticity mass resection.

I operate on both sides kidney under the diagnosis of the hydronephrosis with eight years ago 11 years ago 12 years ago from 12 years ago. A blunder tumor benign pathologically.

A strong left-sided visual disorder continues from ten years ago, but does not progress.

The abnormality is not discovered by an examination of chromosome.

In last August, it is appeared nystagmus. Nystagmus, an appearance (however, there was the nystagmus by 9 once a month in August) that are similar in September. I have the star cell tumor doubt that MRI takes effect, and is contrasted partly by the right parietal area pointed out. I extracted the right calvarial star cell tumor doubt by craniotomy, but there was a blood vessel hyperplasia, and there was not malignant transformation of the glia, and the part which seemed to be a tumor contrasted by MRI was a benign tumor.

<< case 25 >> origin ignorance, man, 39 years old

There is more cafe au lait spot on the left buttocks than birth. A mass appearance more flexible than before four or five years. From two years ago on the left sternal border between the left third rib of 8mm in diameter a border is clear, and notice movable induration firmly. There are (7*4cm) great weak bunch type, pigmentation in the flexible mass of the left buttocks. There are abnormal brain waves at the back of the head. In head CT, there is abnormal shadow in the basal nuclei part.

<< case 26 >> origin ignorance, man, 21 years old

A younger sister is SLE. Though the right Uekuchi labial part has brown spot, in a primary schoolchild, I notice. It was cafe au lait spot.

In a junior high student, I notice the abdomen, ecru spot of both arms. In July, 1984, a brain tumor is found in head CT, and tumor enucleation takes effect. Tumor was benign pathologically.

<< case 27 >> origin ignorance, man, 14 years old

5 seasons and years, left upper arm part elasticity mass appearance. There is the flexible mass on the right neck, the right wrist. There is brown spot in the precordial region after childhood. I have a diagnosis of cafe au lait spot with dermatology.

13 seasons and years, left upper arm part elasticity mass extraction.

I am operated on for a marrow outside-related tumor of third cervical vertebrae, the fifth cervical vertebrae and, in 15 years, take effect. It was a benign tumor pathologically. In addition, there is a brain sickle part meningioma (fibrousness).

[missing an origin as follows. In other words, case oneself an example of the proband for

women]

[summary of the case]

[example derived from mother for men]

As for case 1, as for the death (38 years old), the older brother, as for death (43 years old) case 2, the death (12 years old), the person are in good health for brainstem malignancy nerve sheath tumor an older brother for a tumor (doubt) of the brainstem for a pleomorphic glioblastoma (24 years old); a star cell tumor (19 years old) malignant in head CT as for tumor (-) case 3

Case 4 has bad star cell tumor (grade 3) (33 years old), older sister one, but I am operated on by dislocation of the hip joint and enforce it.

An older brother transfers case 5 to meningitis from pneumonia in 1 year and dies. Brain tumor (23 years old) that mind exercise develops, and health case 6 delays as for tendency to progress (7 years old), older sister three as for the eyesight drop for a congenital aqueduct of midbrain obstruction as for the person, and increase shows a tendency as for malignant star cell tumor (11 years old) case 9 as for pleomorphic glioblastoma (14 years old) case 8 as for congenital heart disease (1 year old) case 7

[example derived from mother for women]

Mass (38 years old) more flexible as for auditory nerve tumor (good nature) (23 years old), the older brother right as for case 10 at 15 years old for a spinal cord tumor as for death case 11 than the infancy to a whole body

[example derived from father for men]

Cauda equina nerve tumor (good nature) (27 years old), an older sister were asthmatical bronchitis, and, as for case 12, as for death case 13, as for star cell tumor (good nature) (32 years old) case 14, as for left pulsatility drawing of the eyes with marking dots (31 years old) case 15, as for speech impediment (18 years old) case 16, as for expansion of the ear canal, elevation (2 years old) case 17 of the right wing-like scale, as for tumor (good nature) (11 months) case 18, a neurofibroma appeared in the whole back mainly on hips to cervical vertebrae for left orbit bottom bone defect in right sella turcica expansion (J type), the right on back of the head midline from one year ago from a bone defect (3.5*3.5cm), the young time, but in the summer from the time regression (natural regression) (10 years old) [example derived from father for women]

Large intestine tumor (13 years old) that the malignancy is strong as for operation (16 years old) case 23 for a malignant star cell tumor as for operation (28 years old) case 22 for a pleomorphic glioblastoma as for operation (21 years old) case 21 for a malignant star cell tumor as for death (34 years old) case 20 for a malignant star cell tumor as for case 19

[case oneself an example of the proband for men]

A marrow outside-related tumor (good nature) of cervical vertebrae, the fifth cervical vertebrae third as for case 24 as for brain tumor (57 years old) case 25 of the good nature in the basal nuclei part as for abnormality shadow (39 years old) case 26 as for brain tumor (21 years old) case 27 of the good nature (14 years old)

[case oneself an example of the proband for women]

I cannot discover one, too.

[consideration]

There are few documents that an origin was specified for a brain tumor of the neurofibroma type 1. 6) that there were the documents of the malignant brain tumor that one origin was specified, but derived from mother in the case of multiple malignant nerve sheath tumors as for it, was a man as for the patient.

In addition, 5) that there is the case that one origin was already specified, but is the documents which showed regression naturally of the orbital part polymorphism glioblastoma of the 2-year-old girl derived from father as for it.

When I consider 27 cases, the neurofibroma type 1 of the male patient derived from mother is strong in a tendency of the malignant transformation. And the female patient derived from father is strong in a tendency of the malignant transformation at the same level, too.

Effect derived from mother in the neurofibroma type 1 namely maternal effect^{6,7,8,10,11,13}) is that it has been said conventionally. However, there are many arguments, and one or two, ^{3,12,16}), the unification of the opinion are not accomplished. The writer proposes intersection effect namely cross effect newly here.

The effect derived from mother exists in this investigation surely. That neurofibroma type 1 is a female patient derived from father is a male patient derived from mother or is other cases or is greatly related to canceration. When the male patient derived from mother, the male patient derived from a female patient, father derived from a malignant tumor being full of it to a female patient derived from father, and being easy to occur and mother and the patient gives that malignant tumor does not occur in the case of proband the big suggestion to a cancer gene therapy in neurofibroma type 1 in this way, I am convinced.

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Intersection effect of the neurofibroma type 1

I performed statistical analysis of 27 neurofibroma type 1.

Conventionally, it has been said that a patient of the neurofibroma type 1 derived from

mother had high disease severity. maternal effect means this. However, there is much dissenting opinion, too. When male patient derived from mother, female patient that the possibility that a tumor was malignant in a female patient derived from father was extremely high and was of mother origin, male patient derived from father, oneself were proband, it was not malignant and was all benign as a result that the writer checked it.

I name this which "the possibility that it is malignant is full of it when a tumor is found in a female patient derived from a male patient, father derived from mother, and is high" an intersection effect (cross effect) newly.

Please translate it into English. I can post it on a foreign medical journal. It is no use in Japan. I am extremely weak in English. Thank you in advance.

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