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原 著

DEGENERATION OF THE DESCENDING FIBERS FROM THE FRONTAL LOBE TO THE LOWER PART OF THE BRAIN STEM IN CASES OF CONGENITAL AND ACQUIRED LESION OF THE FRONTAL LOBE

by

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INTRODUCTION

In each case of cystic malformation of the bilateral frontal lobes and enormous meningioma situated at the base of the right frontal lobe, the brain obtained at autopsy was studied on changes of the descending fibers from the frontal lobe to the lower part of the brain stem. Serial sections were made from the brain stem and stained by SCHNITZLER-KURODA's modification of WEIGERT-PAL's myelin sheath staining.

MATERIALS

Case 1.

Cystic malformation of the bilateral frontal lobes

C.H., a daughter of an engineer, two years and eight months old was admitted to our hospital on May 19, 1947, with a complaint of abnormal enlargement of the head.

Present history: Born spontaneously and at full term. On the third neonatal day, during lactation she suddenly had a fit of deadly pallor and stopped sucking for about five minutes. Such a fit came on repeatedly during the subsequent week. From three months after birth it was noticed that the head was becoming much greater and the systemic development was notably tardy; she could neither fix her head, nor sit. Speaking and grasping were impossible. Sleep was disturbed. There was no convulsion. She had no history of trauma in the head and febrile diseases.

Family history : Her parents were both healthy and their marriage was not consanguineous. Their blood tests after WASSERMANN were negative.

Physical examination : 1. She is underdeveloped, weighing six kilograms. The head is markedly enlarged. The sutures of the skull and the fontanels are not yet closed. Cracked pot resonance is proved. 2. Bilateral optic nerve atrophy are noticed. 3. The upper extremities are held in flexed position, and the lower extremities in pes equinovarus position. With her right hand she performs no skilled movement, only grasps an object. 4. She is apathetic and indifferent. She seems to be able to recognize her mother, but she is unable to talk. 5. The initial pressure of cerebro-spinal fluid shows 140 mm H₂O; the final one, 120 mm H₂O after drawing two milli-liters. It was aqueously transparent; cell count 15.4; negative in globulintest. 6. Both superficial and deep reflexes are normal, and there are no abnormal reflexes.

Operative findings : For the purpose of clipping the chorioidal artery in the right lateral ventricle, a frontotemporal craniotomy was performed. The skull was thin, but the dura mater was found to be normal. The parenchyma of the brain was thinned and membranous, the gyri and sulci being not clear. Laterally to the ventricle, a large oval cavity containing the cerebro-spinal fluid extended from the frontal lobe to the anterior part of the parietal lobe, pushing the temporal lobe downward. The size of the right lateral ventricle itself was almost normal. The right chorioid plexus in the ventricle was rudimentary. The right MONRO's foramen was thumb-tip-sized, and a part of the septum pellucidum failed. Through this septal defect the left lateral ventricle was reached, which was markedly enlarged, and laterally to which several paraventricular cysts were found communicating with one another through openings in their thin walls. Four silver clips were put on the left chorioidal plexus near the posterior horn and eight hundred milli-liters saline solution was injected into the ventricle. After closure of the dura mater and the skin

Fig. 1

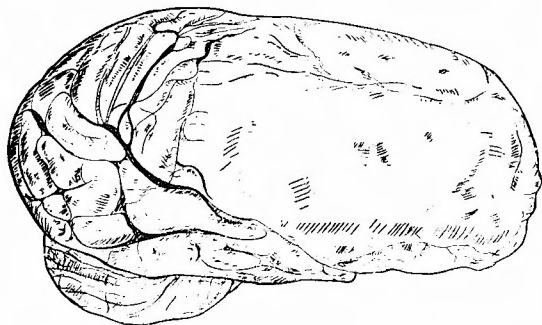
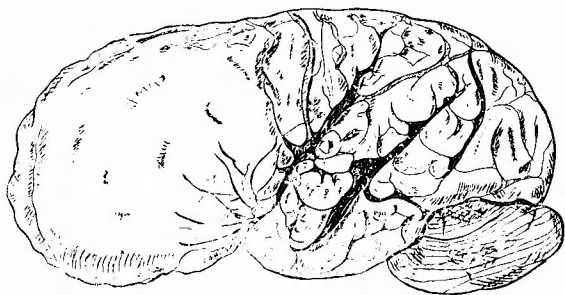


Fig. 2



her general condition suddenly deteriorated and died.

Autopsy findings : Sutures of the skull is not completely closed. Gyrations and sulci rostral to the precentral gyrus are uniformly lost on both sides. The brain substance of bilateral frontal lobes is thin like paper, because just beneath the surface a single large cyst on the right and several large cysts on

the left are present communicating with the corresponding lateral ventricle situated medially, which is dilated on the left. The left chorioid plexus is seen to have been clipped. The third ventricle is slightly dilated. Aqueductus Sylvii is not obliterated. The fourth ventricle is normal. The cerebral surface except in frontal lobes appear to be normal. In the thin cortex of the frontal lobe, the existence of ganglion cells was confirmed histologically but the regular lamellar structure was entirely lost.

Case 2.

The right olfactory groove meningioma

K T., a thirty four year-old male, an office employee.

Admitted to our hospital on Jan. 19, 1950.

C. C. : Visual disturbance.

Past history : In 1945, he had fracture of the right femur,

contusion of chest and was unconscious for 24 hrs. by an explosion accident of mines.

Present history : In 1946, without any causative moment, he complained of visual disturbance, occipital pain, and numbness and motor disturbances in the left upper and lower extremities. In March of 1947 he underwent subtotal extirpation of the right olfactory groove meningioma, weighing 100 grams. After the operation, he regained the sight so as to resume his previous work. In 1949 again the vision was gradually impaired, and finally dropped to the mere perception of light. He became melancholic and needed much care by his family. He is a moderate smoker and drinker, but denies venereal diseases.

Family history : His parents and three brothers are all healthy.

Physical examination : 1. He is well built and in good nutrition, clearly conscious and cooperative with examination, but his face is apathetic and emotionless. 2. Bilateral olfactory disturbances are present. 3. Bilateral visual acuity is decreased to the degree of perception of light. 4. Blood pressure 98/60. Incontinentia albi and nocturnal enuresis. 5. The initial pressure of cerebrospinal fluid is 250 mm H₂O; the final one 120 mm H₂O, after removing 15 milli-liters. Clear and two in cell countings. Positive globulin reaction. 6. Exaggerated bilateral knee jerks. No

Fig. 3

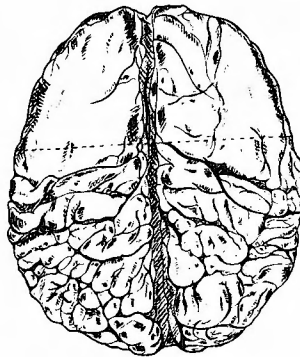


Fig. 4

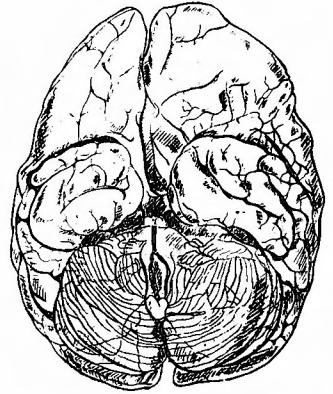


Fig. 5



other pathological reflexes. 7. Excessive alimental and sexual desire.

Operative findings : The operations were performed in two stages at an interval of two weeks. The first was the right frontal craniotomy; the reoperation in the same way as three years before. The dura mater was remarkably thickened. It was found that the recurrent tumor extended to the anterior cranial fossa on the left side. The right-side portion of the recurrent tumor weighing 62 grams was removed from the base of the right frontal lobe. Two weeks later, by the bilateral frontal craniotomy the tumor was removed from the left anterior cranial fossa. The left-side portion of the tumor was 100 grams in weight. Total weight of the excised tumor was 162 grams. The patient died on the following day of the second operation.

Autopsy findings : The right frontal pole was completely lacking and the anterior part of the upper frontal gyrus was also destroyed. Viewed from the basal surface, the lesion involved orbital gyri, olfactory sulci, and rectal sulcus on the right side. Slight damages to the same areas of the left frontal lobe were also seen. In the other portions, no abnormalities were observed except slight enlargement of the right lateral ventricle.

Fig. 6

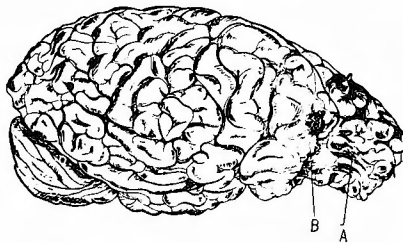


Fig. 7

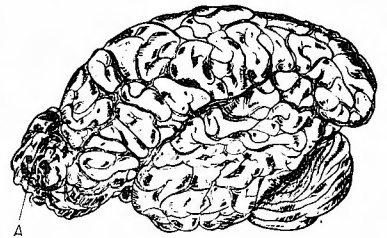


Fig. 8

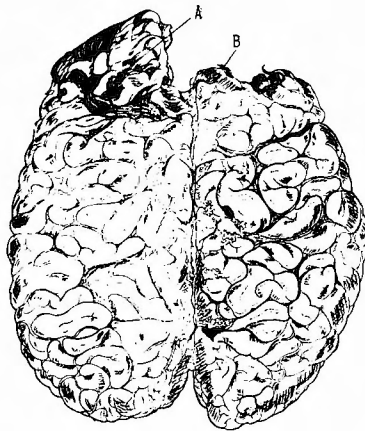
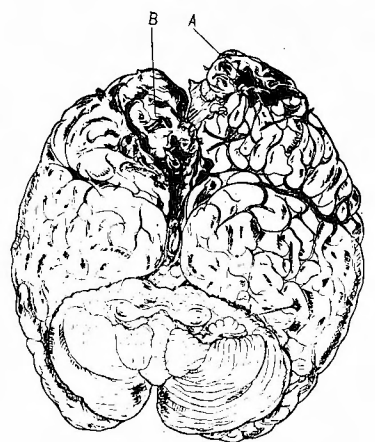


Fig. 9



STAINING METHODS

Serial sections were made from the brain stem and stained by SCHNITZLER-KURODA's modification of WEIGERT-PAL's myelin sheath staining. These methods are my own one.

MICROSCOPIC FINDINGS OF THE SPECIMENS

Serial sections were made and one out of each three sections was picked up and stained, which was given a serial number from cranial to caudal. The descri-

ption of findings was limited to a small number of sections and the abbreviations according to RILEY were used :

BPO	Brachium pontis
NOI	Nucleus olivaris inferior
PSP	Pes pedunculi
TCS	Tractus cortico-spinalis
TFPO	Tractus fronto-pontinus
TPTPO	Tractus parieto-temporo-pontinus

I. Case 1.

(Specimen number) **No. 260** (The level of the pyramidal decussation) Fig. 10.

- (1) The shape is distorted.
- (2) Right TCS is generally poorly stained, and the myelin sheath of each fiber is markedly atrophic, irregular and smaller in size than the left.

No. 246 (The upper level of the caudal limit of the fourth ventricle) Fig. 11.

- (1) The shape is, as a whole, distorted.
 - (2) TCS : the same as above.
 - (3) The right NOI is smaller in size and illchromatic.
- The same finding is obtained from this to No. 226. The left NOI is larger in size and good in stainability.

No. 226 (The caudal limit of the fourth ventricle) Fig. 12.

- (1) Both sides are symmetrical.
- (2) TCS : the same as above.
- (3) Both NOI are the same in size. Their stainability is equal on both sides.

No. 166 (The root of the facial nerve) Fig. 13.

- (1) The root of the facial nerve is seen.
- (2) On the right a part of the abducens nerve has appeared.
- (3) TCS : the same finding as above.
- (4) The NOI has disappeared.

No. 142 (The root of the trigeminal nerve) Fig. 14.

- (1) The right BPO is markedly larger in size than the left.
- (2) TCS : the same as above.
- (3) The TFPO and TPTPO are both indistinguishable.

No. 111. (The middle part of the pons) Fig. 15.

- (1) Both BPO are almost equal in size and symmetrical in shape.
- (2) In respect to the other structures the same findings as above.

No. 61 (The cephalic limit of the inferior colliculus) Fig. 16.

- (1) A gentle curvature of the right PSP is seen.
- (2) TCS : the same as above.
- (3) Bilateral TFPO seem defective.
- (4) The myelin sheath of the right TPTPO is atrophic, but that of the left

is uncertain (the left outer portion of the pes pedunculi has been cut away).

No. 56 (Oculomotor nerve) Fig. 17.

- (1) The oculomotor nerve is clearly seen.
- (2) The right PSP is in a steep curvature. The left is thick and strong.
- (3) TCS is the same as above.
- (4) TFPO is defective on both sides.
- (5) TPTPO : the same as above.

Summary of the histologic findings in case 1 :

The right TCS is markedly atrophic throughout the brain stem, and the left is also atrophic but to a lesser degree. The changes of the TFPO and TPTPO are hardly discernible in the lower brain stem except in the PSP. In the PSP, the TFPO are bilaterally definitely defective. The right TPTPO is supposed to be atrophic (the specimen of the left lacks). To summarise the findings the TFPO is so much atrophic that it appears to fail bilaterally. The TCS is much more atrophic on the right side than on the left. The TPTPO is supposed to be slightly atrophic unilaterally on the right. In the medial one-third of the PSP, marked atrophy is noticed bilaterally, but more intense on the right.

II. Case 2.

(Specimen number) **No. 256** (The upper level of the pyramidal decussation)

Fig. 18.

TCS : bilaterally normal.

No. 216 (The middle level of the medulla) Fig. 19.

All important tracts seem to be normal on both sides.

No. 121 (The level of the abducens nerve) Fig. 20.

No changes are seen.

No. 19. (The level of the red nucleus) Fig. 21.

TCS, TFPO, TPTPO are bilaterally normal.

No. 11 (The mesencephalo-diencephalic junction) Fig. 22.

There seem to be no changes.

Summary of the histologic findings in case 2 :

No change is observed on both sides and throughout the whole course of the TCS, TFPO, TPTPO.

DISCUSSION

(1) In the first case the bilateral entire frontal lobes and the anterior parts of the basal ganglia are occupied by large hydrocystic malformations. At what developmental stage did the cysts appear? Considering that asphyxia or other serious events did not take place at the time of birth, and that the neurological signs appeared as early as on the 3rd neonatal day, though the head remarkably enlarged after three months of age, these malformations developed probably in the later period of the fetal life. However the frontal lobes seem to have developed to a certain degree prior to the development of the cysts, in view of the fact that they still preserve roughly their characteristic outline, showing the definite topographic relat-

ionship to the lateral cerebral fissure and to the central sulci, though frontal gyri and sulci are missing.

Therefore, it may be asserted with a more probability that the TFPO, after once formed, was later destroyed than that it failed *a priori*. Also the fact that there remains an empty space in the medial part of the PSP seemingly corresponding to the seat of the TFPO, may suggest that the TFPO disappeared, after once formed, rather than that it was not formed *a priori*. In the TCS, the change on the right side is much more striking in contrast to that on the left. This change may originate from the internal capsule affected by the cysts, and may be responsible for the striking motor disturbance of the left upper and lower extremities. Though the cystic changes may seem grossly equal in the bilateral frontal lobes, the right internal capsule is affected far more seriously than the left.

The changes of the TPTPO are not definitely confirmed because of the asymmetrical excision of the midbrain specimen (the outer part of the left PSP is cut away), but on the right side the atrophic changes similar to those of the TCS are supposed to exist. This supposition is based on the fact that the PSP on the right side shows an undulating configuration and is strongly curved. Its appearance is more slender than the left. This change is too remarkable to be regarded as being due to the oblique direction of the microscopic sections. In addition, if the intensive atrophic change of the right TCS is considered, the deformation of the right PSP may be the result of the atrophy of the TPTPO in the right lateral part of the PSP. As wrinkles in the skin of the aged, the right PSP becomes wrinkled, because these descending tracts atrophy after having once been formed. Supposed that they were underdeveloped from the beginning, such wrinkles as this would not be produced.

In the specimens at the level of NOI, the difference of the size and stainability between both sides may be regarded not necessarily as pathological findings, but as a failure to adjust exactly the direction of the section (the right half of the specimen is larger at the caudal part of the medulla, but on the contrary, smaller at the rostral part). In the specimens at the level of the BPO, it is uncertain whether the great difference in the size between both sides is of pathological significance or not.

The motor disturbance in the first case seems to be of the type of LITTLE's disease. This should be attributed to the subcortical change, especially that of the internal capsule including the basal ganglia rather than that of the cortex of the frontal lobes.

(2) In the second case, the right frontal lobe, which had developed normally, was later widely destroyed at the basal and the anterior portion by the compression of an enormous olfactory groove meningioma and also by the operative procedure for it. It was supposed that the destruction must have resulted in complete degeneration of projection fibres from these areas during these three years after the first operation, and in addition during two weeks after the recurrence operation. In the findings of the specimen, however, no change was observed in the descending

fibers of the brain stem. Supposed that the TFPO originates from the anterior part of the frontal lobe, as C.V. MONAKOW said, it would appear that some changes should be expected in the TFPO in this case. Thus the origin of the TFPO is considered to be not in the anterior frontal region, but probably in the posterior frontal areas as SUNDERLAND, RILEY and others stated.

CONCLUSION

Following findings were obtained in my study of the descending fibers from the frontal lobes, in the two cases, (1) cystic malformation of the bilateral frontal lobes (clinically atypical LITTLE's disease) and (2) the right frontal lobe damaged by operations for the meningioma arising from the basal surface.

Serial sections were made and stained by the SCHNITZLER-KURODA modification of WEIGERT-PAL myelin sheath staining.

(1) Case 1. Cystic malformation of the bilateral frontal lobes

Frontal lobes of both sides were totally occupied by large cysts which were considered to have developed in the later period of the fetal life and further enlarged postnatally.

Bilateral defect of the tractus frontopontinus and marked changes of the right tractus cortico-spinalis were observed. In this case, which seemed to be of LITTLE's disease type, the motor disturbance was caused by the subcortical changes especially the internal capsule rather than those of the cortex of the frontal lobe.

(2) Case 2. The right olfactory groove meningioma (recurrence)

Marked changes were observed in the basal surface and the pole area of the right frontal lobe at autopsy. Because the operative damage was done two years ago, the degeneration of nerve fibers, if any, should be demonstrable in the specimen. But no changes were observed in the tractus fronto-pontinus, tractus cortico-spinalis and tractus parieto-temporo-pontinus.

From these findings, it would appear that the tractus fronto-pontinus should originate not from the frontal pole, but from the posterior part of the frontal lobe.

Note :

In concluding the paper, the author wishes to express his sincere thanks to professor CHISATO ARAKI for his many valuable suggestions and also to Professor KO HIRASAWA, Professor SUEO EGUCHI of Gifu Medical School and Professor Kozo Ito of Tottori Medical School for their instructions.

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Figures I. H. Kuroda.

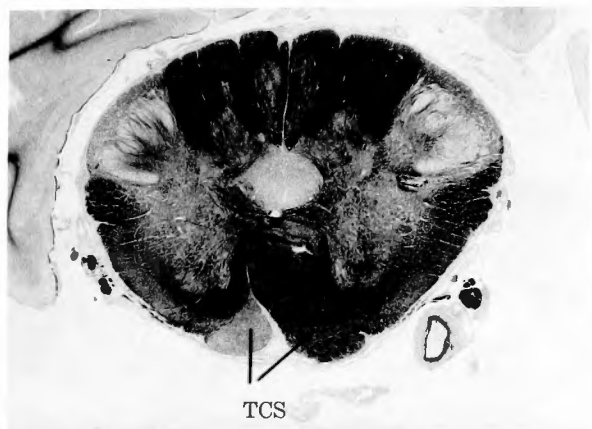


Fig. 10

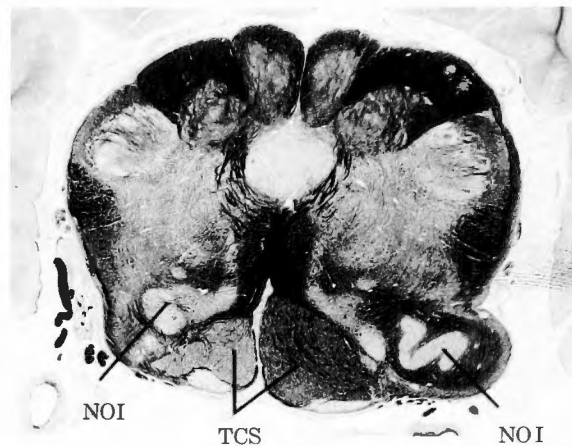


Fig. 11

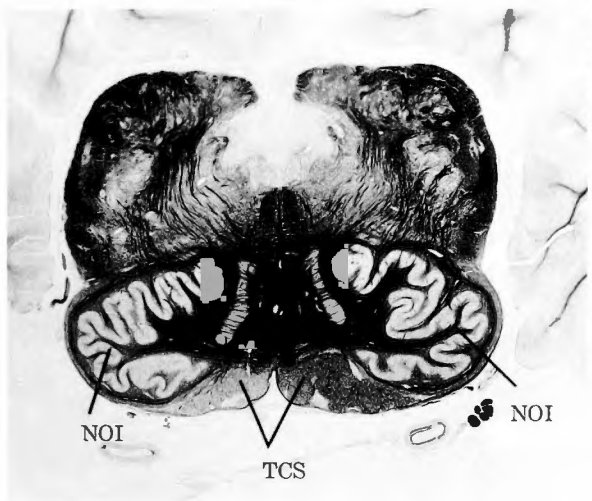


Fig. 12

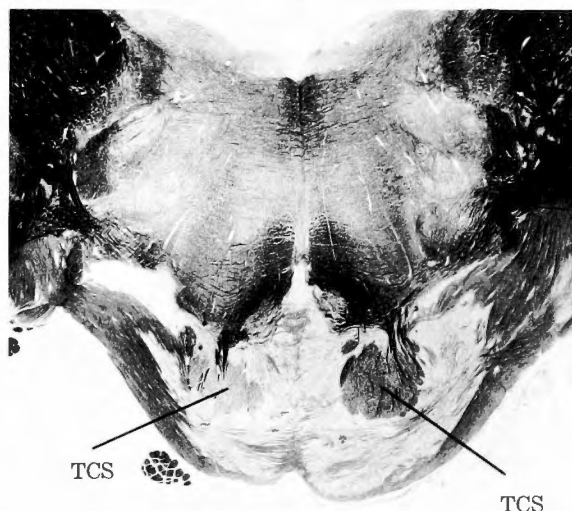


Fig. 13

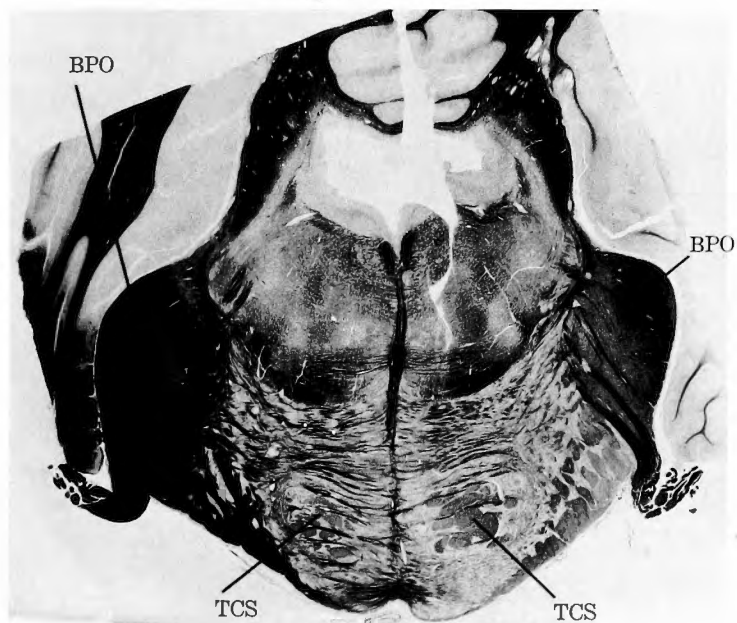


Fig. 14

TCS:

Tractus cortico-spinalis

NOI:

Nucleus olivaris inferior

BPO:

Brachium pontis

Figures II. H. Kuroda.



Fig. 15

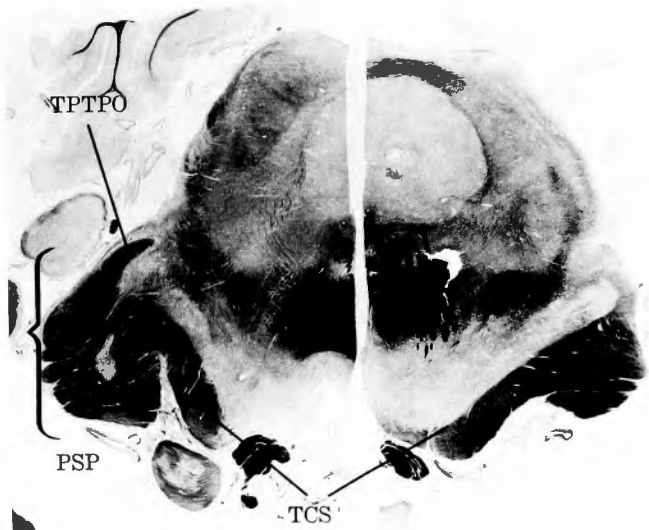


Fig. 16

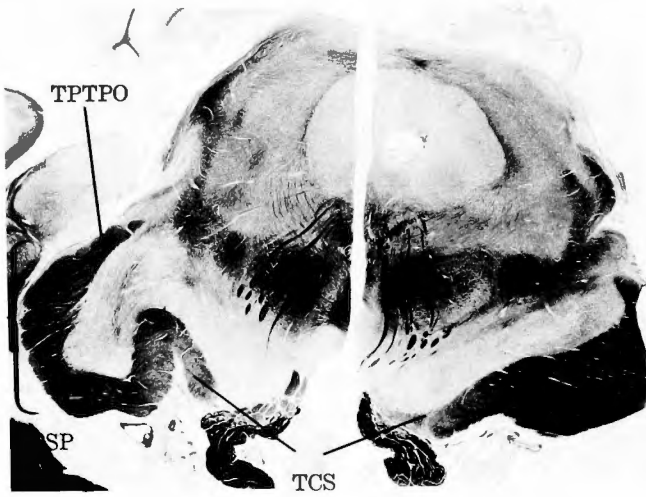


Fig. 17

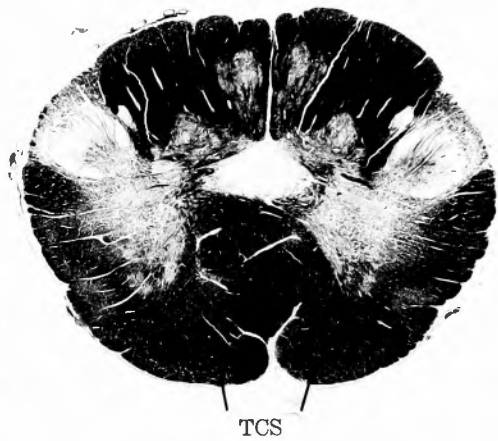


Fig. 18

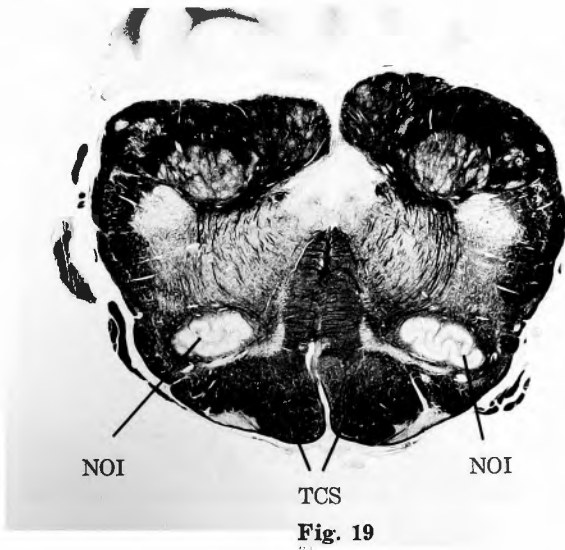


Fig. 19

TCS: Tractus cortico-spinalis

NOI: Nucleus olivaris inferior

BPO: Brachium pontis

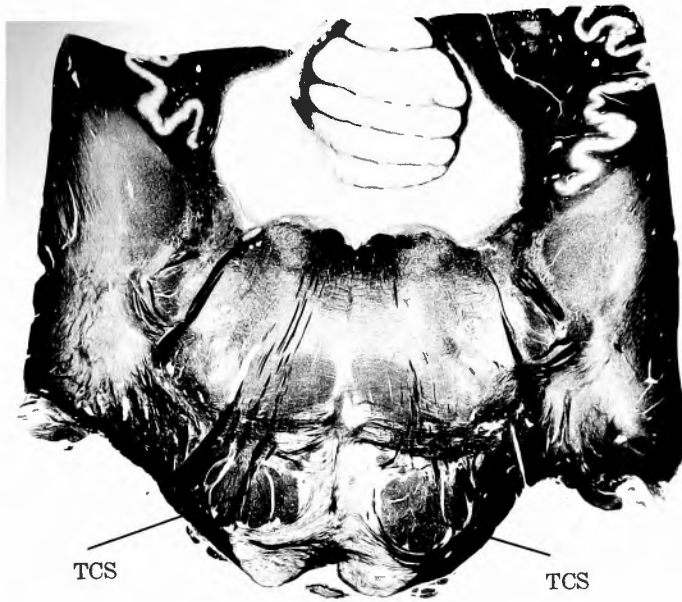


Fig. 20

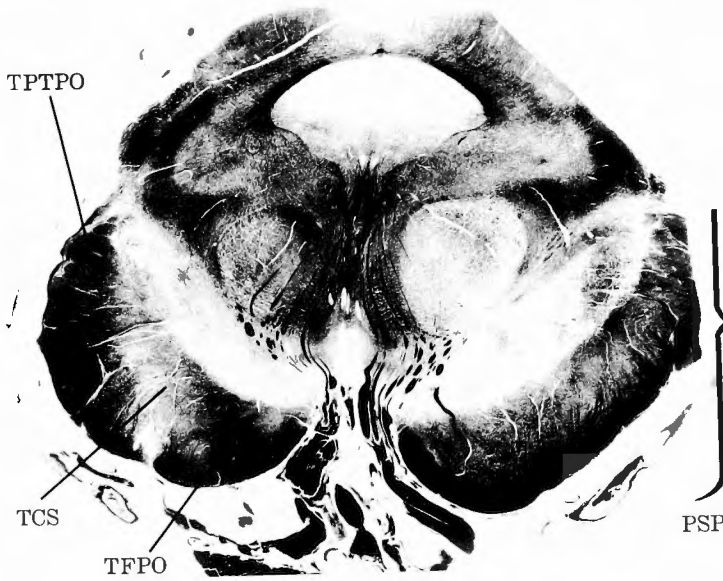


Fig. 21

TCS:
Tractus cortico-spinalis

TPTPO:
Tractus parieto-temporo-
pontinus

TFPO:
Tractus fronto-pontinus

PSP:
Pes pedunculi

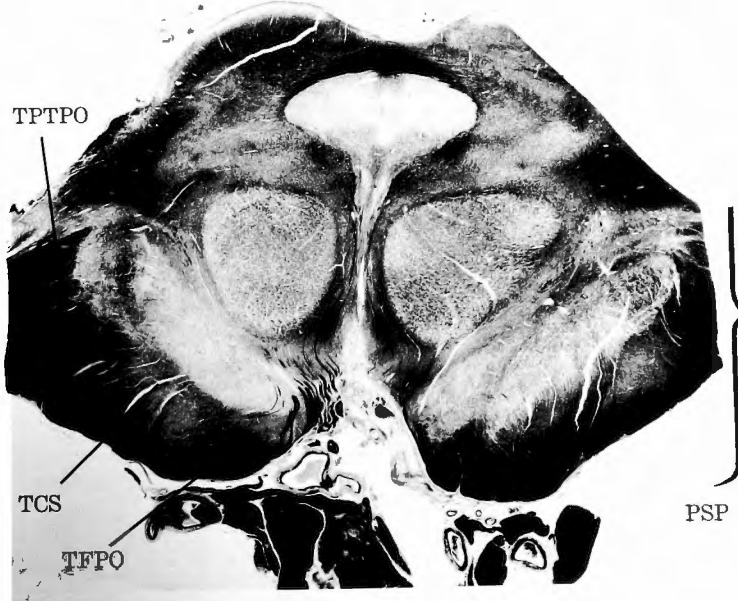


Fig. 22

oclastic trocusses. Arch. Path., 59; 529, 1955. 8) Sunderland S.: The projections of the cerebral cortex on the pons and cerebellum in the macaque monkey. J. Anat., 74; 201, 1940. 9) Takabayashi H. and Kanki S.: Cortico-fugal connections of frontal lobe in man. Medical Journal of Osaka University, 3; 201, 1952. 10) Ogawa T.: The anatomy of the brain stem. Igakusoho 2; Sogensya. Tokyo, 1948. 11) Horimi T.: The frontal subcorticotomy and the brain funktion. Nokenkyu, 1; 54, 1948. 12) Hirazawa K.: The anatomy IV. Nippon-Isyo-Syuppan Co. Tokyo, 1950. 13) Hayashi T. and Okinaka S.: The pyramidal tracts, the extra pyramidal tracts, and the autonomic nervous system. The physiological lectures, 10; Japan Physiolog. Society Press. 14) Hayashi T. and Suda I.: Brain stem, spinal, cerebellum. 15) Hayashi T.: Cerebral cortex, The physiological lectures, 10; Japan Physiolog. Society Press. 16) Katayama Y. and Ishihara S.: The study of the nervous fibre in the operated human brains. The Japan Anatomical Society Press., 30; 98, 1955. 17) Takase K.: The study for development of the neural sheath in the central-nervous system of human foetus. (The third month) Nagasaki-Igakukai-

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附図説明 (1/3 縮小, 凸板)

- Fig. 1. Case 1. Right lateral view
 Fig. 2. Case 1. Left lateral view
 Fig. 3. Case 1. Dorsal view. The line indicates the level of frontal section shown in Fig. 5.
 Fig. 4. Case 1. Ventral view
 Fig. 5. Case 1. Frontal section of the frontal lobes at the level of the interruptal line in Fig. 3.
 Fig. 6. Case 2. Right lateral view
 Fig. 7. Case 2. Left lateral view.
 Fig. 8. Case 2. Dorsal view
 Fig. 9. Case 2. Ventral view
 Fig. 6~Fig. 9. に於ける
 A: Lacerated at operation
 B: Tumor rest

和文抄録

前頭葉の先天性欠損及び後天性損傷の際に於ける 前頭葉から下部脳幹に至る下行路の変性に就て

(京都大学医学部外科学教室第1講座 荒木千里教授指導)

米子博愛病院外科部長 黒 田 秀 夫

両側前頭葉の奇形性脳水腫性囊腫(臨床症状は非定型 Little 氏病)及び右前頭葉底面に生じた脳膜腫の人脳各1例に就て、その脳幹部の連続切片を作り、Weigert-Pal 染色 Schnitzler 黒田変法を用いて、髄鞘の染色を行い、主として前頭葉からの下行路を追求し次の知見を得た。

(1) 症例1. 両側前頭葉の奇形性脳水腫性囊腫

胎生期の後半に於て両側前頭葉に囊腫の発生があり、生後更にそれが増大したと思われるもので、前頭葉は両側共発達せず欠除に等しい状態である。組織標本で前頭橋路は欠除している。皮質脊髓路は右側は著明に変化しているが、左側の変化は軽い。頭頂側頭橋

路も右側で変化を認める。

症例1は、Little 氏病型でこの運動障害は、前頭葉皮質に由来したものと謂うよりも、皮質下の変化特に内包の変化に由来したものであろう。

(2) 症例2. 右嗅神経溝脳膜腫

剖検脳右前頭葉底面及び前極に著明な変化が認められるが、組織標本で前頭橋路、皮質脊髓路及び頭頂側頭橋路に変化は認められない。この所見から考えると、前頭橋路は、C. V. Monakow の謂う如く、前頭極より発しているのではなく、Sunderland, Riley等の謂う如く前中心回から発しているのではなかろうか。