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Cerebellar symptoms as sequelae of traumatic lesions of upper brain stem and cerebellum.

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Cerebellar Symptoms as Sequelae of Traumatic Lesions of Upper Brain Stem and Cerebellum

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INTRODUCTION

An acute midbrain syndrome arising after a head injury is in nearly all cases a consequence of a compression of the upper brain stem due to a transtentorial herniation. The acute midbrain syndrome can disappear or a chronic stage with the symptoms of the apallic syndrome may develop. Pathophysiologically the apallic syndrome is characterized by a lack of all higher cerebral functions and the reduction of the higher central nervous functions to the midbrain level. The morphological findings may show diffuse lesions in the white and grey matter of cerebrum and cerebellum and in the upper brain stem. During the remission stage of the apallic syndrome a gradual reintegration of the cerebral functions takes place.

In about 65 % of the patients with a traumatic apallic syndrome, a defect stage with severe symptoms of cerebral damage and of lesions in the upper brain stem is to be found. The symptomatology of the defect stage of the traumatic apallic syndrome varies according to the multiplicity of the lesion pattern. The diffuse damage of the central white matter causes a dementia. Superposed lesions of primary and secondary traumatic origin cause typical local cerebral symptoms. The lesions in the upper brain stem cause symptoms on the part of the oculomotor system, the substantia nigra, the descending motor pathways and the cerebellar connections. Depending on the severity of the lesions, the various groups of symptoms differ in the their intensity.

MATERIAL

The investigation was conducted from two different viewpoints: Firstly, the clinical symptomatology was studied in a group of 42 patients. In all of these cases morphological studies could not be done, because all the patients survived. For this reason we studied the morphological alterations in a second group of 13 cases.

According to our experience gained from about 130 patients suffering from traumatic apallic syndrome definitive cerebellar symptoms are only to be observed in the advanced stage of recovery. In this state the prognosis quo ad vitam is always good. Patients who died in the early remission stage or in the full blown stage of traumatic apallic syndrome, in general showed no cerebellar symptoms. Therefore, we are unable to correlate the clinical symptomatology with the morphological findings in the single case.

Fig. 1. —Pat. R. H., 5o a, Remission stage of traumatic apallic syndrome. Gait ataxia.

A. Clinical observations

In the first part of the investigation we studied 52 patients in the late remission and defect stage of traumatic apallic syndrome; 42 of them showed cerebellar symptoms. The cerebellar symptomatology is classified in 3 groups. This classification is based upon generally agreed somatotopic considerations of cerebellar disturbances.

1. A wing beating phenomenon was found in 7 patients, in 4 of which there was unilateral accentuation, in 3 it was only unilateral. In 6 of these patients the phenomenon disappeared after 2 to 8 weeks; in 1 this symptom is still existing after five years. A stereotactic operation in the nucleus ventro-oralis thalami of the left side could be the reason for the long persistence of this symptom. The wing beating in this one case was predominant on the right side. This symptom is first observed in the remission stage at the phase of the grasping reflex, more marked in the phase of the Klüver - Bucy - syndrome, and it disappears with further recovery. One of the 6 patients showed a palatal myoclonus after the disappearance of the symptom.

The wing beating phenomenon can be interpreted as a sign of a lesion in the superior cerebellar peduncle.

2. Gait and truncal ataxia, more or less severe, were found in 32 cases (see fig. 1). In 9 patients the ataxia remained rather severe; in 3 cases it even led to abasia and astasia persisting over years. Six patients showed a transitory tremor of the head. The first signs of the gait and truncal ataxia became noticeable in the late remission stage, when the patients were able to get out of bed. At the end

Fig. 2. — Pat. T. L., 18 a. Remission stage of traumatic apallic syndrome. Pneumoencephalogram: Dilatation of the aqueduct and of the fourth ventricle; dilatation of the cella media and the posterior horn.

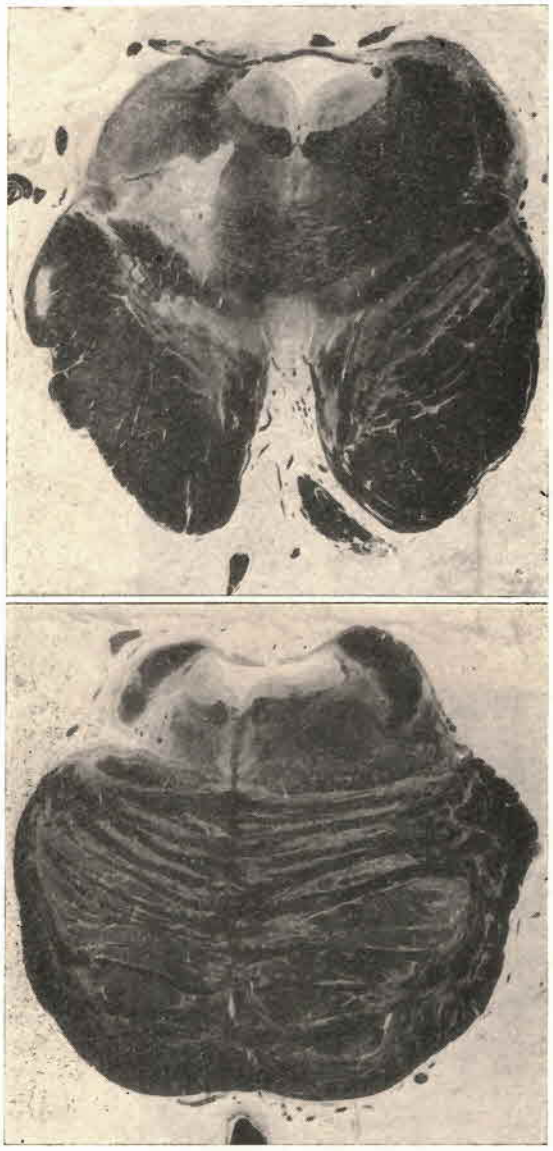

Fig. 3. — Unilateral lesion of the superior cerebellar peduncle and partial destruction of the decussation. (Myelin stain) (Case SN 67/67).

of the remission stage, most of the patients showed an improvement of this symptom. The ataxia of gait and trunk is thought to be due to a lesion of the vermis, especially of its posterior part. Animal Experiments indicate that a lesion of the superior cerebellar peduncle should also be considered as a possible cause of gait and trunk ataxia.

From the patients of the second group, 21 cases showed a combination with cerebellar hemispherical symptoms, 4 to a higher degree, where as all the 7 cases showing the wing beating phenomenon also had gait and truncal ataxia.

3. Symptoms of ataxia of the hemispherical type such as dysmetria, the past-pointing phenomenon, coarse tremor, dysdiadochokinesis, asynergia, megalographia, dysarthria etc. were found in 27 patients. In 6 of them these symptoms were isolated, in 21 cases there was a combination with the symptomatology of the second group; and in 7 cases out of these 21, the wing beating phenomenon was also observed. Hemispherical symptoms start being observed at the end of the remission stage and show a rapid progression. The tendency to improve sets in after a relatively short time. Permanent symptoms were slight or of moderate severity. Only in the 7 cases showing the combination of vermis-symptoms and the wing beating sign did the permanent symptoms remain severe.

As for the hemispherical symptoms in cases of traumatic apallic syndrome, it is difficult to find a somatotopical correlation. Lesions of the cerebellar pathways as well as cortical or subcortical lesions in the cerebellar hemispheres may be considered.

In all 42 patients with cerebellar symptoms, the other typical permanent symptoms of the apallic syndrome were to be found at a more or less severe degree, i.e. spastic and pseudobulbar paralytic symptoms, signs of Parkinsonism (in 30 patients only), focal symptoms of the oculomotor system (in nearly all cases) as well as symptoms of diffuse and focal cerebral lesions.

The pneumoencephalogram was performed in 30 % of the cases. Patients with severe and moderately severe upper brain stem symptoms showed dilatation of the 4 th ventricle and the aqueduct (fig. 2). In some cases a dilatation of the basal cisterns could be demonstrated. This find-

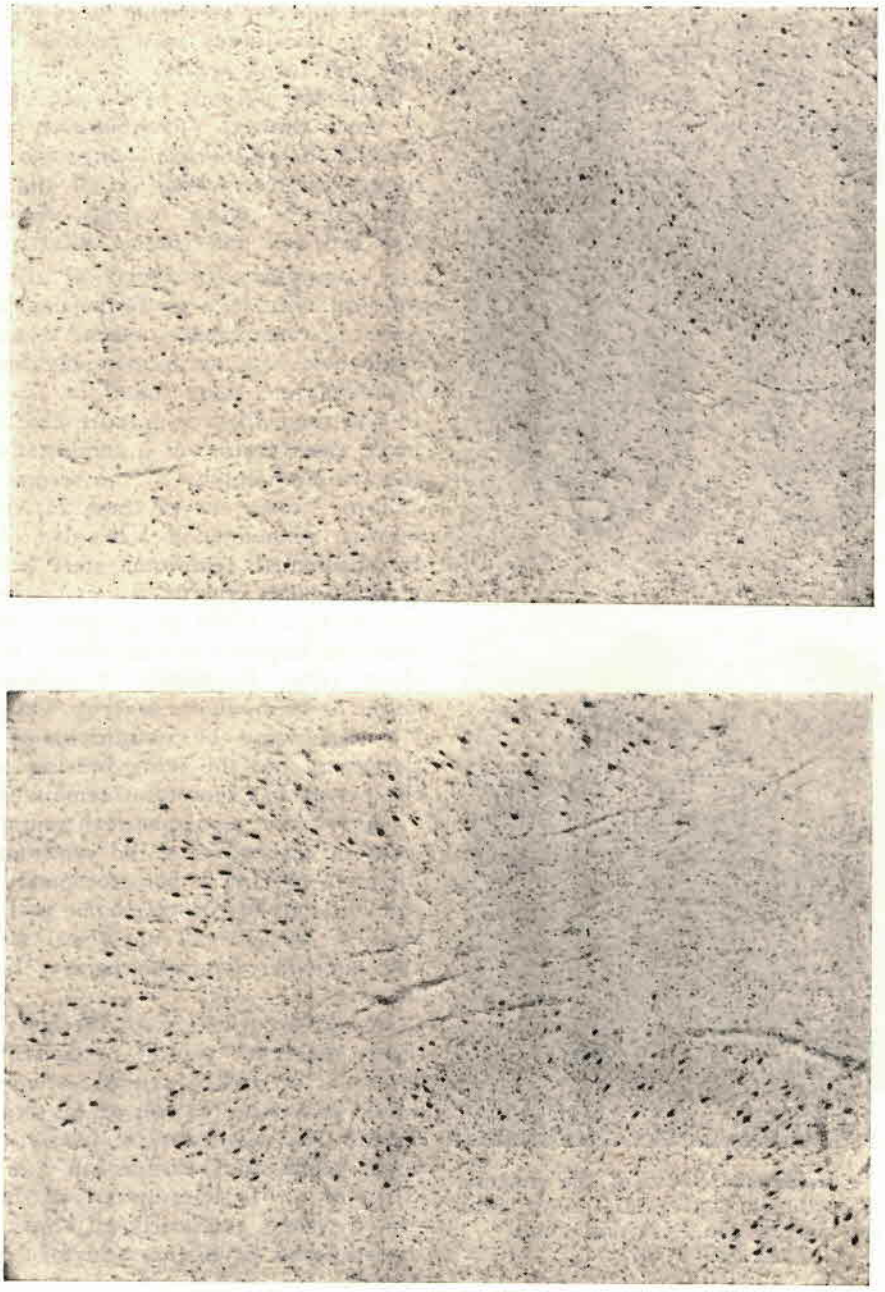

Fig. 4. — Same case as fig. 3. Neuronal population of the dentate nucleus from the intact side and of the dentate from the damaged side. (Nissl stain) (Case SN 67/67).

ing indicates a loss of substance in the upper brain stem region.

For comparison, we studied another group of 41 patients in which the acute

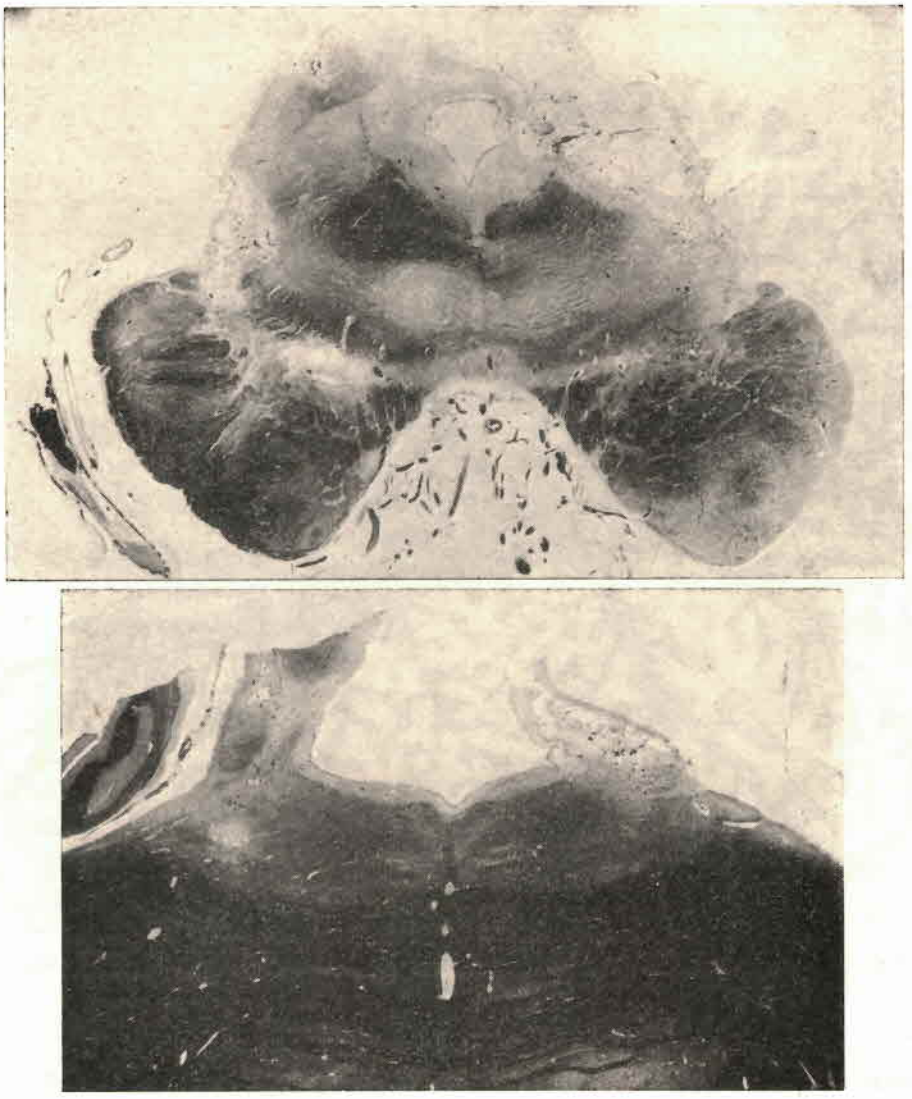

Fig. 5. — Bilateral lesions of the superior cerebellar peduncle. Advanced demyelination of the decussation. (Myelin stain the picture at the top, Elastica van Gieson stain for the picture at the bottom) (Case SN 255/64).

midbrain syndrome improved without developing an acute traumatic apallic syndrome. Only three of these patients showed cerebellar symptoms. The symptomatology was of the hemispherical type, in all cases transitory and of slight degree, in one of them unilateral exclusively.

B. Morphological observations

As remarked above, the patho-anatomical study has been done in a different group because the 42 patients of the clinical study survived.

13 cases of traumatic apallic syndrome

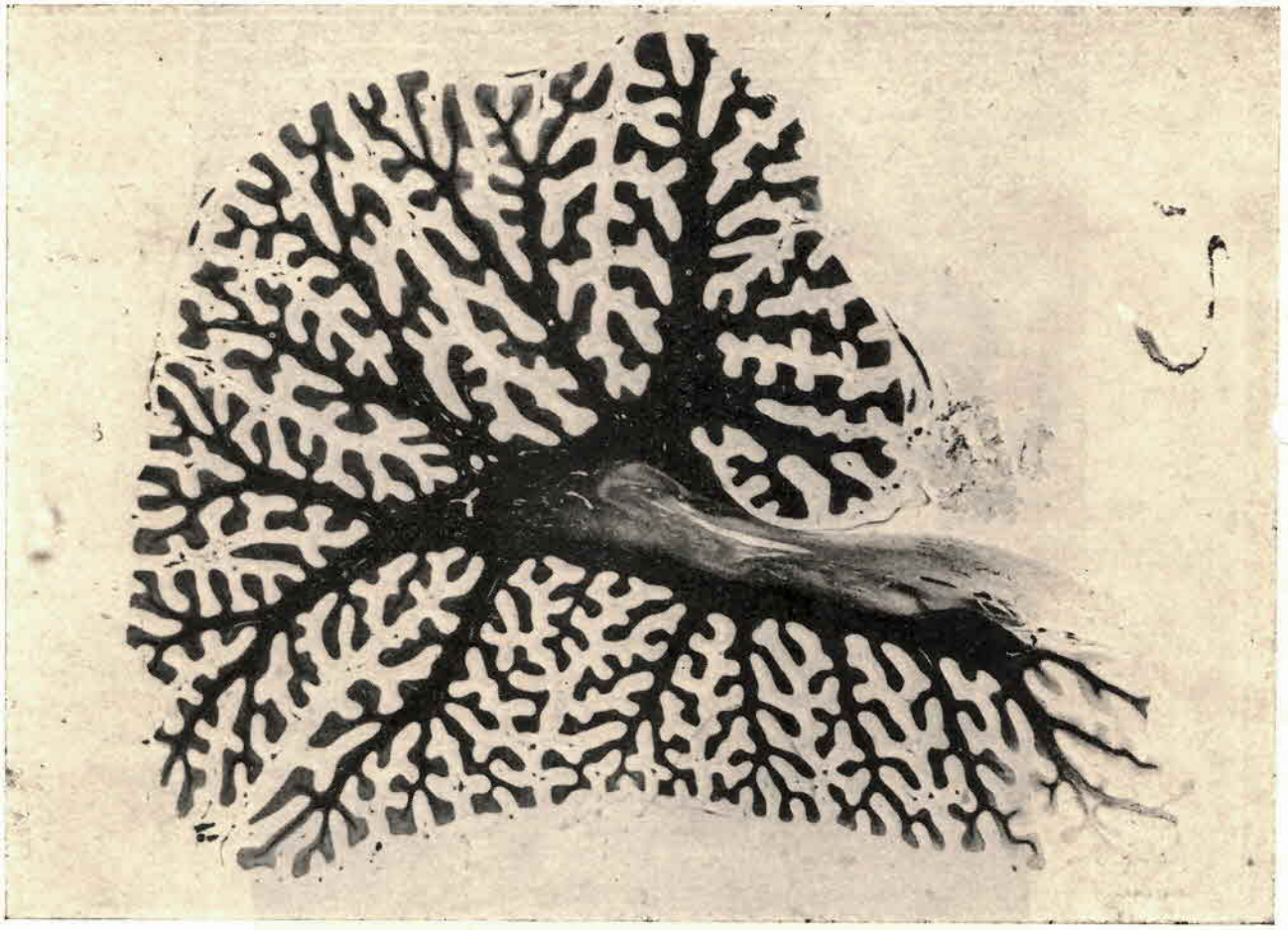

Fig. 6. — Same case as fig. 5. Secondary degeneration of the efferent fibres of the dentate nucleus. (Myelin stain) (Case SN 255/64).

have been studied. 6 patients died in the full stage of the apallic symptomatology, the other seven in the remission state.

According to the anatomical localization, the observed lesions could be divided in 4 groups:

1. Lesions in the superior cerebellar peduncle.
2. Lesions in the vermis.
3. Lesions in the cerebellar hemispheres.
4. Lesions in the pons.

All cases are listed in table 1.

Group 1: Lesions in the brachia conjunctiva were demonstrated in 6 cases. In 3 cases the lesions were minor; in two of these there was a small circumscribed area of incomplete edema necrosis, in the third case a small area of complete stage 2 necrosis in the upper part of one superior cerebellar peduncle with edematous loosening up of the adjoining tissue was found. Secondary changes in the dentate nucleus could not be ascertained. In the remaining 3 cases extensive areas of necrosis were found in the superior cerebellar peduncle (one unilaterally [fig. 3], two bilaterally [fig. 5]) and in one case also

TABLE 1

Localization and intensity of morphological changes (y = year; m = months; d = day; rem = remission stage; s.c.p. = superior cerebellar peduncle)

Number of cases	Autopsy number	Age at time of death	Survival time	Stage of p. ap. Sy. at time of death	Localization and intensity of morphological changes							
					Brain stem			Cerebellar hemispheres		Vermis		
					Pons	Sup. cerebellar peduncle	Decussation of s.c.p.	Dentate nucleus	White substance	Dorsal vermis	Ventral vermis	Vermis in toto
F_1	255/64	29 y	11 m	full stage	—	+++	++	+++	+	++	—	+
F_2	67/67	25 y	4 ½ m	Rem I	+	+++ 1 side	+++ 1 side	+++ 1 side	+	—	—	+
F_3	292/66	20 y	24 d	full stage	+	++	+	—	+	+	—	+
F_4	71/69	29 y	3 ½ y	Rem I	—	++	+	++	+	+	—	—
F_5	409/66	17 y	33 d	full stage	—	+	(+)	—	+	—	—	+
F_6	37/62	11 y	9 m	Rem I	—	+	(+)	(+)	+++	—	+	—
F_7	367/61	42 y	6 m	Rem II	+++	—	—	—	+++	++	—	+
F_8	13/64	51 y	23 d	full stage	+++	—	—	—	+++	+	—	—
F_9	39/64	44 y	7 m	Rem II	+++	—	—	—	+++	—	+	(+)
F_{10}	313/64	53 y	7 m	Rem II	++	—	—	—	+++	—	—	+
F_{11}	271/63	43 y	5 m	full stage	++	—	—	—	++	—	—	(+)
F_{12}	111/61	45 y	22 m	Rem III	++	—	—	—	++	—	+	—
F_{13}	398/68	9 y	3 m	full stage	+	—	—	(+)	+	—	—	(+)

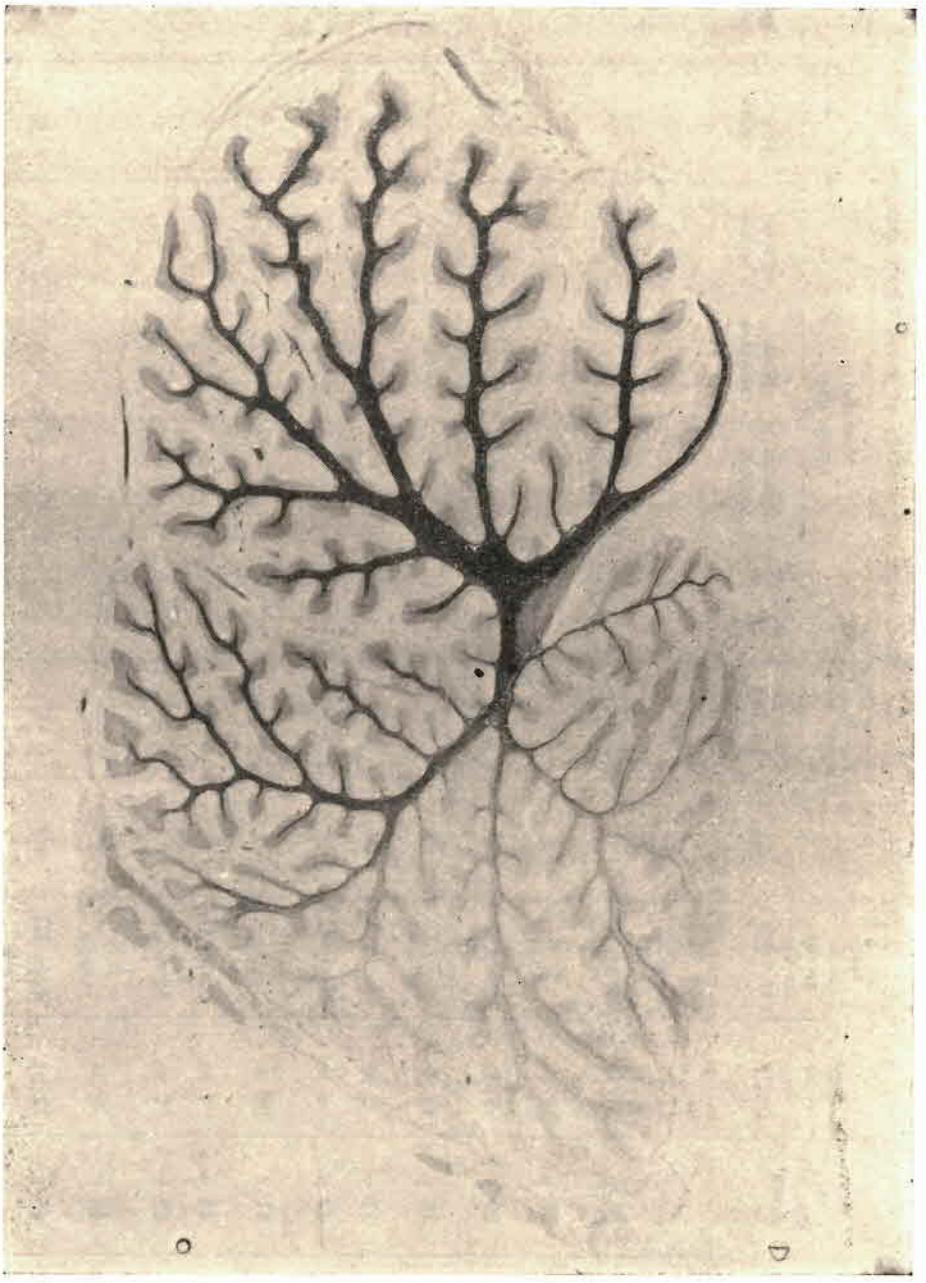

Fig. 7. — Sagittal section trrough the vermis. Demyelination of the ventral vermis. (Myelin stain) (Case SN 37/62).

in their decussation. In these 3 cases secondary lesions in the dentate nucleus were already evident and consisted of severe nerve cell and myelin loss (fig. 6). In the case with unilateral lesions in one superior cerebellar peduncle only the ipsilateral dentate showed some loss of nerve cells (fig. 4). One case with bilateral

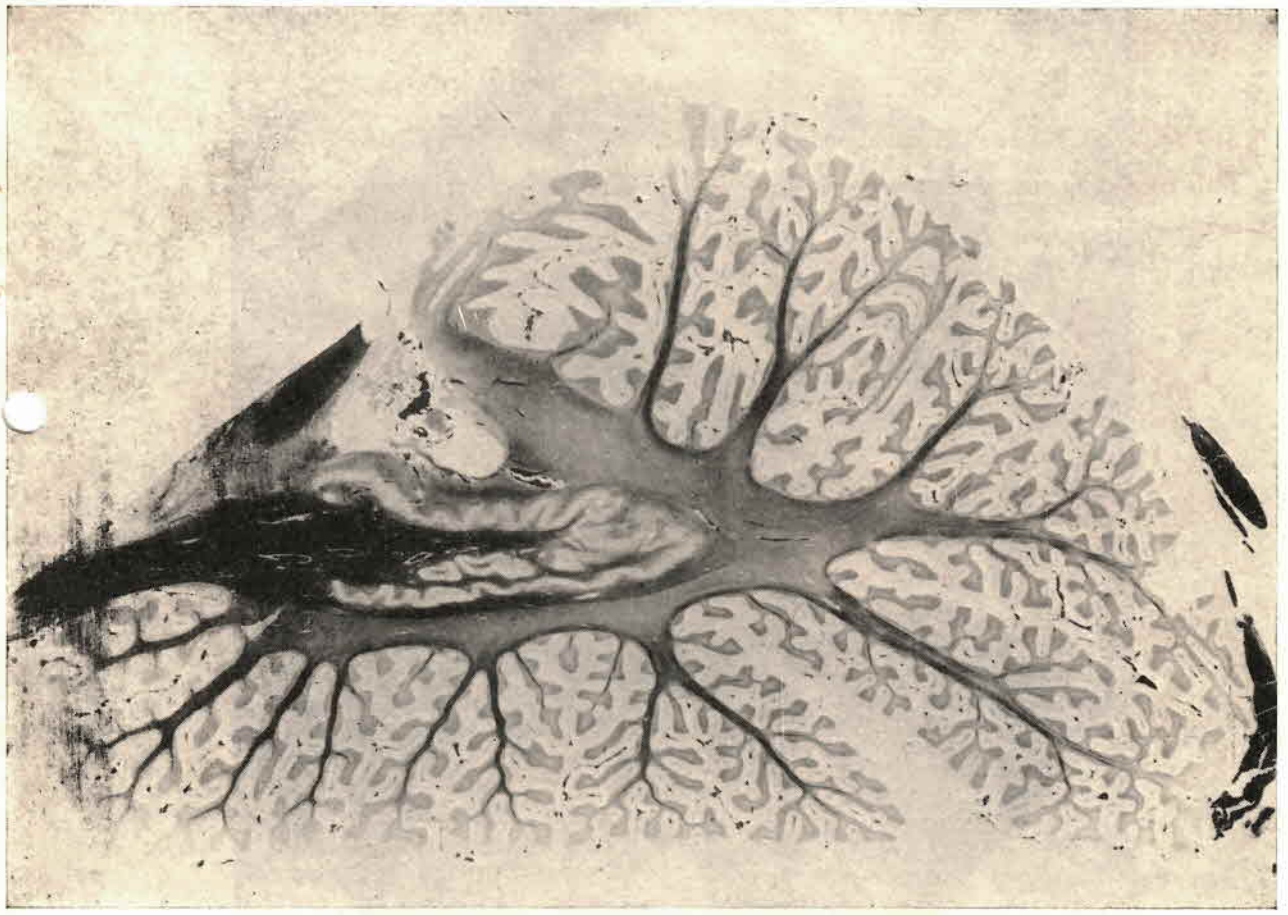

Fig. 8. — Severe demyelination of the cerebellar hemispheres with sparing of the nerve fibres in and around the dentate nucleus. (Myelin stain) (Case SN 367/61).

lesions (survival time 11 months) displayed a marked fibrillary gliosis in both dentates.

Group 2: The lesions frequently observed in the vermis were a mild or severe loss of Purkinje cells, an unevenly reduced nuclear density of the granular layer and a more or less pronounced pallor of the corresponding foliar white matter. In 6 cases, these findings were more prominent in the dorsal vermis, whereas in 3 cases they were more evident in the ventral vermis (fig. 7) and in 4 cases they were equally distributed throughout the vermian cortex. Furthermore, in 4 cases a diffuse, slight to moderate paleness as well as a moderate glial reaction of the central white matter were encountered.

Group 3: The lesions found in the cerebellar hemispheres consisted of an edematous alteration of the central white matter, corresponding to the lesions observed in the cerebrum in cases of traumatic apallic syndrome (fig. 8). Thus, in 5 cases a mild, diffuse pallor of the myelin preparations was found along with a slight to moderate glial reaction. In 7 cases the loss of myelin ranged from moderate to severe with a corresponding degree of glial reaction, which, in some isolated cases, was made conspicuous by a very distinct fibrillary gliosis. With the

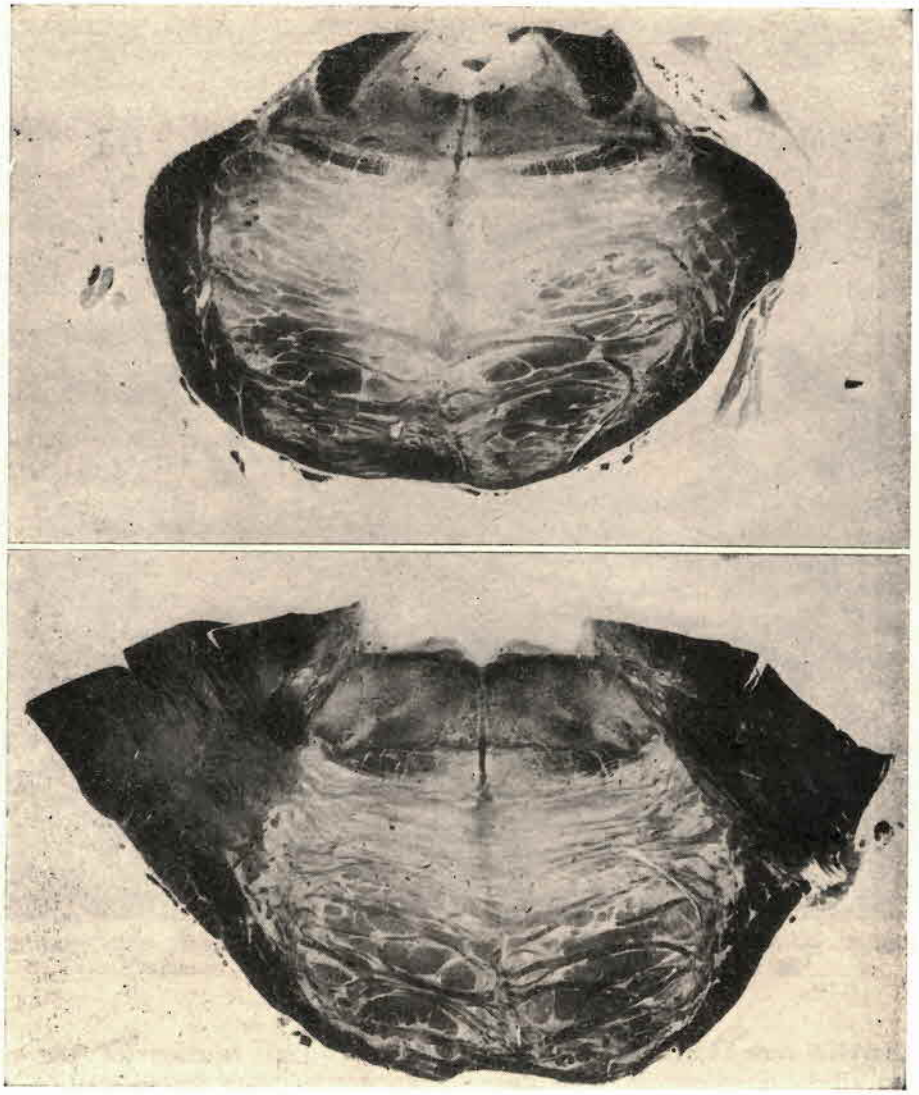

Fig. 9. — Extensive central demyelination of the rostral and middle portion of the pons (Myelin stain) (Case SN 13/64).

appropriate staining, axonal rarefaction as well as fragmentation and beading could be detected in the severely demyelinated areas. Widespread and marked axonal changes were not encountered.

No mention of the cerebellar cortex changes was made in the description of the pathological findings because they could not be clearly separated from the non specific and terminal changes, often found in this structure.

Group 4: To these findings in the hemispheres very frequently corresponded similar findings in the pons. And indeed the degree of severity of the edematous lesions in both regions matched very closely, with one single exception. Thus in 6 cases a marked pallor of the myelin

sheets was found, in three of which moderate, in three severe. The pallor was always accentuated in the central pons, the brachium pontis remained almost unaffected (fig. 9).

In none of the 13 cases studied, the changes found at the level of the inferior cerebellar peduncles were ever worth mentioning.

DISCUSSION

In the material available we could not demonstrate a direct correlation between the clinical cerebellar symptomatology and the patho-anatomical findings in each single case of posttraumatic apallic syndrome. As mentioned above, the cerebellar symptoms are not encountered before the advanced remission stage. In this stage the prognosis normally is favourable, so that it is rarely possible to confirm the clinical symptomatology by demonstrating patho-anatomical findings. In view of this difficulty we considered it necessary to compare two different groups of patients, one having been studied only clinically, the other one only morphologically. Furthermore this comparison seemed to be justified by the selection of identical groups of patients as far as the history is concerned: in both groups the patients had suffered a severe head injury and showed the picture of a traumatic apallic syndrome after going through an acute midbrain syndrome.

Morphological changes of the cerebellum and its tracts as sequelae of a closed head injury have already been described by other authors (Strich [6, 7], Jellinger [3] and others). Strich found lesions of the superior cerebellar peduncles in 12 of her 20 cases, Jellinger in 6 out of 25 cases. Secondary degenerative changes in the tracts and nuclei concerned have been described repeatedly. Diffuse oedematous alteration of the cerebellar white matter often is encountered. Chronic oedema-caused lesions of the central pons are reported by Jellinger [3] in 6 of his 25 cases. In our material the occurrence of morphological changes is about of the same order. In spite of the variety and number of possible combinations of clinical symptoms as well as of patho-anatomical lesions a distinct accentuation of a clinical feature (e.g. the wing beating phenomenon) and also of an outstanding morphological lesion (extensive necroses in the brachia conjunctiva) may be found in certain cases. This observation does not permit definite conclusions, but it fits well into the generally agreed concept of structure-function correlation. The same applies to the correlation of other cerebellar symptoms such as gait and trunk ataxia, dysmetria, the past-pointing phenomenon, coarse tremor, dysdiadochokinesis and others with the morphological findings in the vermis and the cerebellar hemispheres. As to the pathogenesis of the morphological changes we would like to refer to the results of Peters [5], Jellinger [3], Mayer [4] and Strich [7]. As a rule, they probably represent secondary traumatic changes (although differentiation may be difficult, especially if necroses and haemorrhages are of long standing). From the patho-anatomical point of view it further seems to be worth mentioning that there is a remarkably frequent coincidence of advanced demyelinisation of the cerebellar hemisphere with lesions in the central pons while pronounced lesions in the brachia conjunctiva frequently lack substancial changes in the hemispheres and pons. For the changes in the cerebellar white matter a traumatic cerebral oedema generally is regarded as the essential pathogenetic factor, but opinions are uncertain regarding the pathogenesis of the pontine lesions. The remarkably frequent combination of lesions of the cerebellar white matter and pons which we observed would rather suggest a common pathogenetic mechanism. Necroses in the cerebellar peduncle could most easily be explained as being secondary changes following an obstruction of the venous drainage after midbrain herniation. The accentuation of the lesion in either the upper or the lower part of the vermis is a result of an ascending and a descending herniation respectively.

SUMMARY

Patients suffering from a traumatic apallic syndrome often show a cerebellar symptomatology during the remission phase. This corresponds well to the partly severe morphological changes in the cerebellum and its tracts. The difficulty of direct correlations in the single case has been pointed out.

RESUMEN

Los pacientes con síndrome traumático apálico a menudo presentan una sintomatología cerebelosa durante el período de su recuperación. Esto corresponde a los cambios morfológicos del cerebelo y sus vías, que son en buena parte considerables. Se señala la dificultad de establecer correlaciones directas en casos aislados.

RÉSUMÉ

Les patients ayant un syndrome apallique traumatique souvent présentent une symptomatologie cérébelleuse pendant la période de recouvrement. Ceci correspond aux changements morphologiques du cervelet et ses voies, qui sont en partie considérables. On signale la difficulté pour établir des correlations directes dans des cas particuliers.

ZUSAMMENFASSUNG

Patienten mit einem traumatischen apallischen Syndrom haben oft eine zerebellaere Symptomatologie in der Phase der Remission. Dieses entspricht vollkommen den teilweise schweren morphologischen Veraenderungen im Kleinhirn und seinen Bahnen. Die Schwierigkeit direkter Beziehungen im einzelnen Falle werden hervorgehoben.

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