A CASE OF INFANTILE HEMIPLEGIA ASSOCIATED WITH FACIAL NÆVUS AND MENTAL DEFECT.

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The following example of the now familiar syndrome which is characterised clinically by infantile cerebral hemiplegia in association with a contralateral capillary nævus of the skin appears to differ from all previously reported cases in that, notwithstanding the marked degree of cutaneous involvement, the pathological changes found after death were hardly visible to the naked eye.

REPORT OF CASE.

The patient, M. B., a girl, age 17, was admitted to hospital on September 22, 1926. Her mental state was one of low grade imbecility; she was faulty in her habits, destructive, and capable of saying only a few words.

An extensive capillary nævus of the 'port-wine' type was present over the greater part of the left side of the face and scalp, on the left scapular region, and on both legs.

On the face the nævus was sharply defined, reaching slightly beyond the middle line in the region of the nose (fig. 1). On the scalp it extended backwards as far as the vertex. The chin and submental region showed only irregular blotching of a colour somewhat lighter than that elsewhere. The scleral vessels at the inner and outer canthus of the left eye were slightly varicose and markedly injected. The mucous membrane of the left cheek and of the left side of the palate was nævoid; on the hard palate the nævus reached accurately to the median raphe.

The left side of the face was also slightly larger than the right, the left eyebrow on a higher horizontal plane than the right, the tissues of the left cheek were thicker and the left half of the upper lip more prominent than the corresponding parts on the opposite side.

Central nervous system.—Pupils equal; reactions to light and accommodation normal; other cranial nerves normal.

The right limbs were almost completely paralysed and spastic. The right upper limb was in close juxtaposition to the trunk, flexed at the elbow with the hand in a position of extreme flexion; the lower limb was extended with the foot in the position of talipes equinovarus.
Abdominal reflexes present. Plantars: right extensor response; left normal. Right knee jerk brisker than left.

The patient was subject to epileptic convulsions of the Jacksonian type commencing near the angle of the mouth. At the onset of an attack the head and eyes turned to the right followed by tonic and clonic movements of muscles on the right side.

In October 1927 the patient commenced to have fits in large numbers and finally died convulsed on November 1, 1927.

PATHOLOGICAL FINDINGS.

Post-mortem examination.—No changes of importance were found in the visceral organs. The calvarium and dura mater were normal. The brain was small and showed marked asymmetry, the left cerebral hemisphere being considerably smaller than the right; its weight was 40 oz. The pia mater was slightly congested and thickened, especially in the neighbourhood of the great longitudinal fissure. Over the frontal and parietal convolutions of the left hemisphere there was a noticeable fulness of the veins, but careful scrutiny failed to reveal anything in the nature of an angioma (fig. 2). A decided roughness, however, could be felt beneath the pia arachnoid covering the upper end of the gyrus centralis, and a vertical coronal section showed that the cortical grey matter in this situation was infiltrated with minute gritty particles (fig. 3).
Fig. 2.—Cerebral hemispheres viewed from above. The scale lies over the frontal lobes. No nevus can be seen.

An additional point of interest was the smallness of the left optic nerve and tract in comparison with those of the opposite hemisphere.

Microscopical Examination.—In the superficial layers of grey matter at the upper end of the gyrus centralis and to a lesser extent in the posterior end of the superior frontal gyrus, there was a rich deposit of spherical, oval and mulberry-shaped masses of inorganic substance (fig 4). In areas a little

Fig. 3.—Vertical coronal section to display infiltration of upper end of precentral gyrus with calcarceous deposit (arrow). Note inequality of hemispheres.
further away numerous capillaries outlined by granules could be seen. Chemical analysis of the affected brain cortex showed that the deposit consisted almost entirely of calcium in the proportion of 123 mgm. of metal per 100 grm. cortex. In areas where the deposit was particularly dense the grey matter showed no details of structure and contained minute foam-like spaces. Individual nerve-cells were well preserved, but there was considerable disturbance of cell lamination in the affected area and poverty of nerve-cells was obvious in most laminae; in the precentral gyrus the Betz cells showed a marked numerical deficiency.

Fig. 4.—Low power view of upper end of gyrus centralis showing masses of calcium salts lying in the superficial layers. Lying in the pia-arachnoid is a small arteriole with uneven development of its wall. This is better seen in the next figure.

No angiomatous vessels could be found in either the grey or white matter, but in the pia mater vessels showing definite abnormality were noted. In this situation the minute arterioles displayed great variations in the thickness of their walls (fig. 5). Apart from this there was no indication whatever of abnormal vascular changes.

DISCUSSION.

In its clinical aspects the case conformed closely to those reported by other writers. The patient was mentally defective, and from an early age had been subject to epileptic fits of the Jacksonian type. An extensive capillary nævus was present on the greater part of the left side of the face and on certain parts of the trunk and limbs. On the right side there was an incomplete hemiplegia. No skiagrams were taken of the patient's head, but in view of the similarity of the case to that reported by Kalischer¹ and others
it was confidently anticipated that the post-mortem examination would reveal an extensive leptomeningeal angioma on the left side of the brain and its failure to do so gave to the case its chief interest. Apart from a moderate degree of thickening and congestion of the pia-arachnoid, inspection of the convex surface of the brain revealed nothing of importance and it was only by careful palpation that evidence of an underlying pathological condition was revealed. In an area limited to the upper end of the precentral gyrus and the grey matter of the adjacent superior frontal gyrus, a deposit of gritty material was discovered which on microscopic examination proved to be composed of minute particles of calcium salts. In the pia-arachnoid covering this area there were present several small vessels with faulty development of their walls. *These constituted the sole evidence of angioma.*

![Image](http://jnnp.bmj.com/first-published-as-10.1136/jnnp.s1-12.45.47-on-1-july-1931/downloaded-from-http://jnnp.bmj.com/)

**Fig. 5.—Higher magnification, showing the abnormal pial vessel seen in fig. 4.**

As already mentioned the left cerebral hemisphere was considerably smaller than the right. This abnormality, which was present in Kalischer's case and in the first case reported by Cushing, occurs on the same side as the facial and leptomeningeal angioma—an association which suggests that the latter may in some way be responsible for both the cerebral hypoplasia and the contralateral hemiplegia.

In the case reported above, however, where no angioma visible to the naked eye could be found, such an explanation is clearly inadmissible. Moreover, in the writer's experience inequality of the two cerebral hemispheres is not an uncommon finding in the brains of persons who exhibit mental defect without cutaneous nævus; the condition seems to be the result of a simple developmental arrest, and when associated with congenital infantile hemiplegia, as is
often the case, examination of the cortex reveals little else than paucity of the cortical neurones. It seems likely, therefore, that the meningeal angioma is an associated developmental anomaly which has nothing to do with either the mental defect or the infantile hemiplegia.

SUMMARY.

Examination of the brain of an imbecile girl, who during life exhibited an extensive capillary nævus of the skin with contralateral hemiplegia, revealed no macroscopic evidence of angioma. The pathological changes were limited to a small area of the cortex and consisted of a few minute vessels with imperfect walls and a rich deposit of calcium salts in the grey matter.

REFERENCES.

1 KALISCHER, Arch. f. Psychiat., 1901, xxiv, 171.