STURGE-KALISCHER-WEBER SYNDROME OF BILATERAL DISTRIBUTION

BY

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Congenital naevi of the face (and body) may be associated with corresponding lesions of the leptomeninges, the latter giving rise to epilepsy—often Jacksonian in type—and in some cases hemiparesis. Sturge, in 1879, reported the case of a girl, aged 6½ years, with an extensive telangiectatic naevus, especially of the right side of the face and head, right-sided buphthalmos, and epileptic attacks starting in the left hand ; he concluded that the right side of the brain was also involved in the naevoid condition. Schirmer, in 1860, had already described the case of a man aged 36 with an extensive naevus, especially of the left side of the face, and left-sided buphthalmos, but he made no reference to epileptic attacks or a possible cerebral lesion.

Kalischer, in 1897, reported a case described as a diffuse evidently congenital telangiectasis of the scalp and brain, and in subsequent post-mortem study (Kalischer, 1901) the lesion is described as a blood-vessel tumour (angioma) of telangiectatic character with progressive alteration and participation of veins and capillaries in the growing lesion.

Parkes Weber, in 1922, reported the radiographic appearances of the brain in a typical case which showed a large telangiectatic naevus involving especially the left side of the face, right-sided hemiparesis, and hypotrophy of the affected limbs. *X*-ray examination of the skull revealed a more or less calcified and apparently " festooned " lesion on the surface of the left cerebral hemisphere. A year later Dimitri (1923), in the Argentine, described the radiographic findings in a similar case.

Other cases in which facial naevi—with or without similar lesions on other parts of the body—have been associated with proved leptomeningeal angiomata, or with symptoms such as epilepsy suggesting the presence of a similar cerebral lesion, have been reported by several observers. The condition may also be associated with buphthalmos (" ox-eye," congenital glaucoma) on the same side as the main cutaneous naevus, as in Parkes Weber's case (1922), three of Cushing's cases (1906 and 1928), and two of Brushfield and Wyatt's cases (1927). Mental deficiency may also be present, as in all Brushfield and Wyatt's cases (1927) and one of Cushing's (1928).

Unilateral intracranial haemorrhage from the abnormal blood vessels of the meningeal angiomatous condition may also occur in early life (or even intrauterine life) and may cause or increase the spastic hemiplegia. The history in one of Cushing's cases suggests this possibility, and in one of Brushfield and Wyatt's cases the left hemiplegia was first noticed after a severe left-sided convulsive attack at the age of 6 weeks.

The actual cerebral lesion would appear to be a capillary angioma of the leptomeninges, as judged from the few post-mortem examinations it has been possible to carry out —e.g., the cases of Kalischer (1901), Strominger (1905), Cushing (1906, 1928), and Brushfield and Wyatt (1927). Thus the meninges are described as " unduly vascular and in all probability representing a naevoid condition similar to that of the skin " (Cushing's first case) ; the cerebral meninges thickened and excessively vascular (Strominger's case) ; left cerebral hemisphere covered by a diffuse meningeal angioma (Brushfield and Wyatt). A very vascular condition of the dura mater with adhesions to the leptomeninges has been found at operation (Cushing). The affected cerebral hemisphere is sometimes smaller than that of the opposite side (e.g., Kalischer's case and one of Cushing's cases).

Parkes Weber (1946) concludes that the condition is due not to any genetic cause but to an " accidental local injury (mechanical, chemical, or physical) to the ovum at some period after fertilization—that is to say, to the embryo during early intrauterine life."

Should an extensive naevus involving mainly one side of the face be present in a patient with contralateral Jacksonian epilepsy or a hemiparesis, the presence of a pial angioma, even if not revealed by calcification, is sufficient to justify the diagnosis. All the cases hitherto recorded appear to have been unilateral. Oppenheim, however, in 1913 reported four cases, in two of which the apparently involved cerebral hemisphere was contralateral to the facial naevus.

The following case showed extensive cutaneous naevi on both sides of the face and body, although more extensive on the right side, which was also hemiparetic. *X*-ray examination of the skull showed evidence of bilateral intracranial calcification. The epileptic attacks were generalized, although clonic movements usually began and were more pronounced on the right side of the body ; also, the child was mentally deficient (amentia).

Case Report

The patient, a female child, was first seen in December, 1942, at the age of 7½ months. She was a first child, full-term, both the pregnancy and the birth (vertex) being normal ; weight at birth, 6 lb. 7 oz. (2.92 kg.) Convulsive attacks started at the age of 4 months and have continued at intervals ; the fits were mainly but not exclusively right-sided. She has had as many as five fits in one day and has seldom been free from attacks for longer than two or three days. On phenobarbitone, ¼ gr. (16 mg.) t.d.s., the attacks are somewhat reduced in frequency.

The family history showed that a cousin on her mother's side had a cutaneous naevus on the left side of the forehead but without fits. The father and mother were in good health and there had been no miscarriages. There was no consanguinity of the parents and the mother's blood Wassermann reaction was negative.

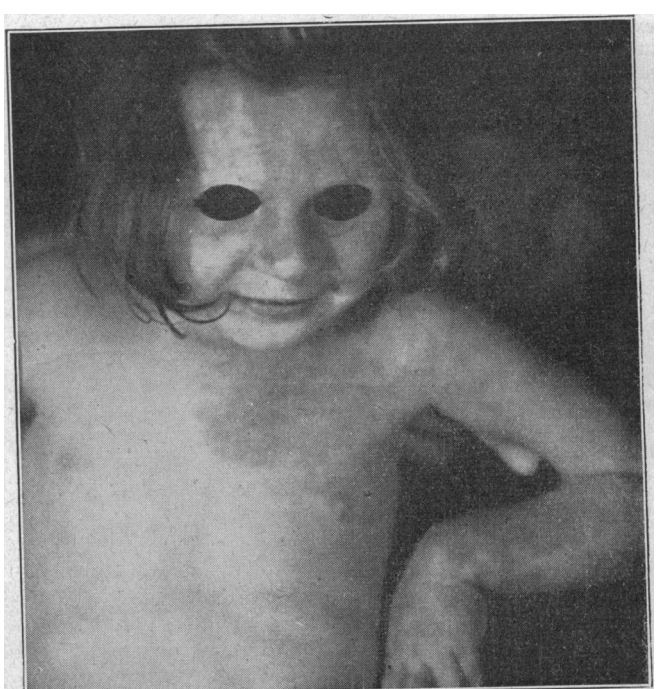

FIG. 1.—The patient at the age of 3½ years, showing extensive cutaneous naevi of left side of face and chest and left arm, with less pronounced naevus formation on right side of face.

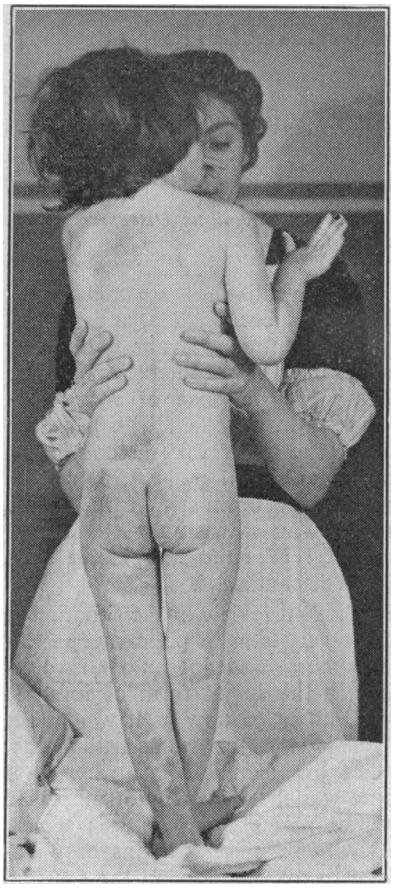

FIG. 2.—The patient at the age of 4½ years, showing the cutaneous naevus of the upper part of the back on the left side, both buttocks—mainly the left—and the left leg.

On examination (December, 1942), extensive and bright-red capillary naevi were present on the left side of the face, scalp, and upper portion of the trunk; the left arm and hand and the outer portions of the left leg were also involved. On the right side of the face and scalp similar but less extensive naevi were present; on neither side of the face did the lesions extend below the level of the mouth. Further, on the right side smaller naevoid patches were seen on the posterior aspect of the shoulder, scapula, gluteal region, and calf, and on the outer side of both dorsum and sole of the foot (Figs. 1 and 2, taken later). Her weight was 14 lb. 6 oz. (6.52 kg.) and she appeared to be mentally deficient. The cranial nerves and optic disks were normal. There was slight spasticity of the right arm and leg, with increased deep reflexes on the right side, but she moved the limbs fairly well. Heart and lungs were normal.

The cerebrospinal fluid was clear to the naked eye but showed 240 red cells, with 6 white cells per c.mm., mainly small lymphocytes but with an occasional small endothelial cell; total protein, 140 mg. per 100 ml., with globulin in slight excess. Lange 1211110000 and W.R. negative. Blood W.R. negative. *X*-ray examination of the skull at this stage showed no abnormality.

The fits observed in hospital started with twitching of the right side of the face—at the angle of the mouth and around the eye—and deviation of eyes to the right, the clonic movements spreading to the right arm and leg; there was loss of consciousness with spread of the twitching to the left arm and leg.

Subsequent Progress

The patient was seen again a year later (December, 1943), aged 1 year and 7 months. Epileptiform attacks had continued at intervals, usually beginning with twitching of the right side of the face, arm, and leg as before; occasionally, however, the attack would start with twitching of the left arm. On whichever side the attack started, however, both sides were eventually involved, but the right side usually more than the left. The child appeared unconscious for only a minute or so, and sometimes consciousness was not lost. The anterior fontanelle was closed. *X*-ray examination of the skull now showed extensive fine calcification on the left side of the cerebrum and also similar calcification but of lesser extent on the right side.

At the age of 3½ years she could sit up unsupported but was unable to stand. She made no attempt to speak, but at times uttered a monotonous cry. She followed a lighted electric torch with her eyes and turned her head at a sound. The cerebrospinal fluid now showed only two small lymphocytes per c.mm. and a total protein of 40 mg. per 100 ml.; both W.R. and Lange reactions were negative. A blood examination was

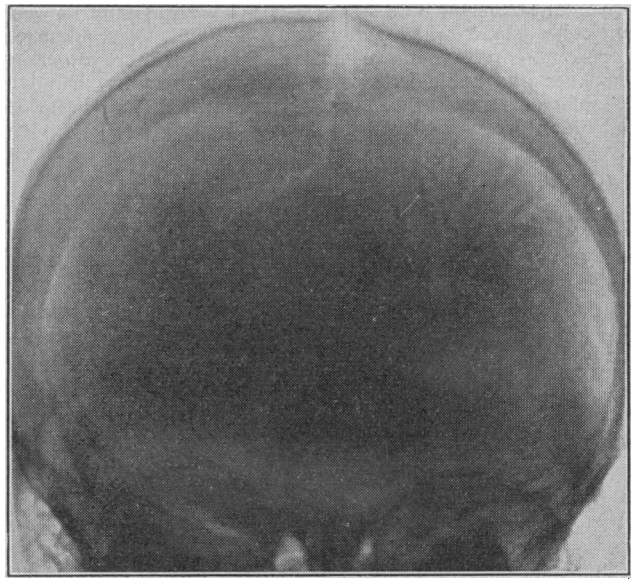

FIG. 3.—Skiagram of skull (antero-posterior view) showing bilateral intracranial calcification, more pronounced on the left side. (Patient at the age of 3½ years.)

normal, with 5,040,000 red corpuscles; Hb, 92%; colour index, 0.8; and white cells normal.

An *x*-ray examination of the skull again showed bilateral intracranial calcification, but more pronounced on the left side than the right. In the antero-posterior view (Fig. 3) the calcification is seen in fine linear markings radiating outwards towards the periphery. In the lateral view calcification extends from the cribriform plate to the occipital region, being more pronounced in the fronto-parietal areas (Fig. 4).

When seen at the age of 4½ years (January, 1947) she had remained free from fits for several months at a time. When they occurred she might have any number from 1 or 2 to 12 in one day. They remained more or less of the same type, mainly right-sided but spreading to the left side, and sometimes vomiting occurred.

On examination speech was still absent and she made only unintelligible noises. The right arm was spastic, held flexed at the elbow, wrist and fingers, but showed a fair degree of voluntary movement. The right arm jerks were all brisker than those on the left. The left arm was used more than the right. The abdominal reflexes were present, left brisker than right. There was slight adductor spasm of the legs, the right leg being more spastic than the left. Voluntary movements were present but were weaker and of less range in the right leg, which was ¼ in. (1.9 cm.) shorter than the left. There was a tendency to

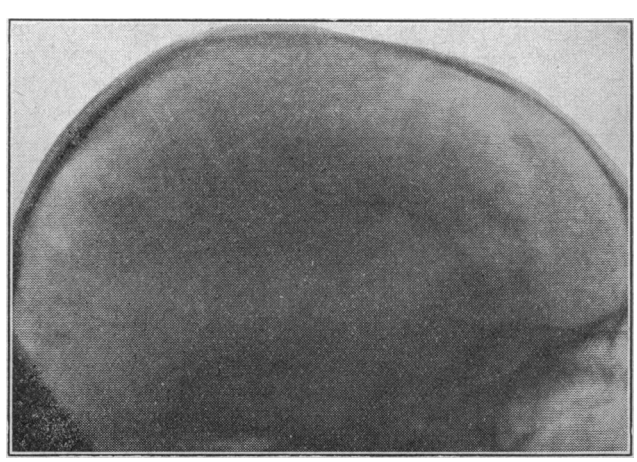

FIG. 4.—Skiagram of skull (lateral view), showing intracranial calcification extending from the supra-occipital to the frontal region. (Patient at the age of 3½ years.)

talipes calcaneus on both sides. She made no attempt to walk, but could stand when supported with both legs adducted. Occasionally restless movements of all limbs were observed. The knee and ankle jerks were brisk, right greater than left, and plantar reflexes indefinite. Heart normal, pulse rate 76, and blood pressure 105/70.

Commentary

I am not aware of any previous record of a case of Sturge–Kalischer–Weber syndrome showing bilateral intracranial calcification. Cases of extensive cutaneous naevi on both sides of the body, usually more continuous on one side and patchy on the other, in association with Jacksonian epilepsy are occasionally encountered. One such case under my care is described in an addendum to Parkes Weber's paper of 1929; in this case no intracranial calcification was demonstrable on *x*-ray examination. In the case now reported the calcification is more extensive and more intense on the one side (left cerebral hemisphere) than on the other, and this corresponds to definite hemiplegic signs in the spasticity of the right arm and leg and the increased deep reflexes. Similarly, the epileptiform attacks usually begin on this (right) side, and even when more generalized the right side is the more affected. The blood found in the cerebrospinal fluid at the first lumbar puncture at the age of 7½ months was almost certainly due to contamination at the time of lumbar puncture (no doubt resulting from intense local vascularity) rather than to a recent subarachnoid haemorrhage from the intracranial angioma, as the red cells were quite fresh and there was neither haemolysis nor xanthochromia. Then again, the cerebrospinal fluid examined at the age of 3½ years was quite normal.

The calcification of the meningeal angioma evidently began between the ages of 7½ months and 1½ years, since no *x*-ray abnormality was seen at the earlier age, but a year later intracranial calcification was demonstrated on both sides, the more profound lesion of the left cerebral hemisphere showing the more extensive and denser calcification.

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Presiding at a conference of the Tuberculosis Association in Belfast, Dr. Frederick Heaf, Director of the London County Council tuberculosis services, said the organization in Ulster for dealing with the disease was the envy of the profession in England. Dr. Brice Clarke, Director of the Northern Ireland Tuberculosis Authority, informed the conference that there were 1,200 beds in tuberculosis hospitals in Ulster, as well as a number of beds in general hospitals for tuberculosis patients. Seven hundred additional beds were planned and 250 of them would be ready within the next twelve or eighteen months. A children's orthopaedic hospital was being built and an infants' and children's hospital for pulmonary cases was being developed. It was hoped to have five or six clinics in Belfast and thirteen in other parts of Ulster. In addition, a contract had been placed for 100 rest chalets. Since June, 1945, 93,000 persons had been examined by the mass radiography unit in Belfast and a second unit had been ordered.

POLIO-ENCEPHALITIS

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The 1947 epidemic of poliomyelitis has again attracted attention to the question of polio-encephalitis, and there is no doubt that considerable confusion prevails concerning the definition of this condition. McAlpine (1947) stated that "during this epidemic the presence of nystagmus, photophobia, and neck rigidity in a patient who has been ill for a few days with headache and fever was considered sufficient evidence for the diagnosis of polio-encephalitis." Murray (1947) suggested that the brain-stem form of the disease should be regarded as an extension of the spinal type, and that the notification of polio-encephalitis as a separate condition should be discontinued.

Rothman (1931), in an excellent review of the situation at that time, stated that "the exact status of polio-encephalitis (using the term to mean cerebral and not bulbar involvement) and its relationship to the spinal form of infantile paralysis remains nearly as unsettled now as it was at the time of its description." This statement holds true to-day. It is hardly surprising that this should be so when it is remembered that the name was first given by Strümpell in 1885 to a form of cerebral paralysis in children which, although displaying many of the manifestations of poliomyelitis, was probably due to toxic action on the cerebral vessels causing thrombosis or haemorrhage without evidence of encephalitis (Ford and Schaffer, 1927).

It has been stated that increased tendon reflexes, absent abdominal reflexes, and extensor responses are evidence of cerebral involvement in poliomyelitis (Kiss and Fenyes, 1936). These signs, and the combination of spastic and flaccid paralyses, may be due to pressure on the pyramidal tracts from inflammation and oedema in the white matter of the cord.

Scheinker (1947), in a detailed study of six cases, found that no part of the central nervous system entirely escaped damage, and that the medulla, pons, mid-brain, and cerebellum were involved in every case, while the cerebral cortex was not affected at all. He also found that inflammatory changes were intense but neuronal destruction rare in the higher centres, in contradistinction to conditions in the anterior horns. Bodian (1947), reporting on 24 human cases, stated that to the pathologist all cases of poliomyelitis were encephalitic, that some areas of the brain were never affected but the main cerebral changes were from the brain-stem to the hypothalamus and thalamus. So far as the motor cortex was concerned, Bodian considered that only in rare cases were the lesions severe enough to cause clinical signs, and that spastic and psychic sequelae were not cortical in origin—nor were stupor, disorientation, and coma—but were produced by areas of focal softening in the basal centres.

The following case under our care appeared to show encephalitic manifestations and involvement of lower motor neurones.

Case Report

A girl aged 17½ had been well until Nov. 9, 1947, when she suddenly developed a cold and generalized headache. On Nov. 12 vomiting started, being repeated several times, and she was admitted to hospital as a case of suspected meningococcal meningitis.