A NOTE ON CUTANEOUS AND MENINGEAL HÆMANGIECTATIC NÆVI ASSOCIATED WITH CONTRALATERAL EPILEPSY AND/OR HEMIPLEGIA (SYNDROME NEURO-CUTANÉ)*

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PERSONAL CASE

The patient is a girl, age 10, an only child, who was brought to hospital with the complaint of 'attacks during which she becomes paralysed all down the right side.'

The history is that the birth was normal, and the birthmark on the left side of the face was more vivid then than now. She was quite healthy until the age of six months,

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when she had a generalized epileptic attack involving the whole body. No paralysis was noticed, but the attacks recurred until she was 12 months old and then ceased.

In the intervals between she was quite normal. She walked at 12 months, and weakness of the right side was then first noticed, the arm being held limp from the elbow and the leg dragged slightly. With massage the paresis gradually improved.

At the age of two years, the present Jacksonian attacks commenced, but she has

been able to attend school regularly and has been quite active. The child knows when the attacks are coming; she says 'I'm bad, mama.' Then jerking of the right side of the body sets in, commencing in the face and spreading to the right limbs. She does not lose consciousness, knows her mother and can reply to questions, though in a

mumble. The turns last only a few seconds, and she sleeps afterwards and is generally limp and quiet for a few days. The hemiparesis is accentuated during this time—she cannot pick anything up and drags the leg in walking—but it then gradually improves. The frequency of the fits is about once every 3–4 weeks. In the intervals between, she is quite well. The maternal grandmother had fits, but there is no history of skin nævus.

The patient is a bright intelligent child of normal physical development. There is a 'port-wine stain' nævus involving chiefly the skin area of the first and second divisions of the left trigeminal nerve, the inner margin ending quite sharply in the midline. The right limbs are shorter, smaller and weaker than the left. She is a left-handed child. There is no noticeable abnormality in the gait. There is weakness of the right side of the face, but the remaining cranial nerves are normal. Deep reflexes in the right limbs are all increased; the right plantar response is extensor, and the right abdominal reflexes are weak.

Ophthalmic Report.—Vision, full. Pupil reactions—direct, normal. Wernicke's hemianopic reaction not present. Fundi and retinal vessels normal. Slight conjunctival injection of left eye, of a type allied to the facial nævus. No vascular abnormality of left iris on slit-lamp examination. No buphthalmos in left eye. Tension of both eyes normal (right 12, left 15). Fields of vision show a well-defined homonymous hemianopia with loss of the right half of each field. No hemianopic scotomata.

X-ray of Skull.—Left half smaller than right. A sinuous calcified shadow visible in occipital region above the level of the tentorium, extending upwards and forwards to central region of hemisphere: (?) a similar shadow visible also in anteroposterior view.

Wassermann negative. Remaining systems normal.

DISCUSSION

Most of the recorded cases of the angiomatous malformations of the brain have been unexpectedly met with at exploratory craniotomy for Jacksonian epilepsy, hemiplegia or tumour and have not been associated with cutaneous nævus, and in consequence there was nothing to help in the preoperative pathological diagnosis. All the series described by Cushing in his book, *Blood-vessel Tumours of the Brain*, and Sargent's six cases were of this type. On the other hand, in a minority of cases of intracranial angiomatosis it has been possible to make such preoperative pathological diagnosis from the presence of cutaneous and especially facial nævus in the patient, an association which has long been known and has recently been described in a few cases as familial. In neither of the groups has the histopathology of the brain lesion been carefully worked out, but it seems to be established that when intracranial lesions are to be expected in a case of cutaneous nævus they will be found to be of the group of angiomatous malformations and not of that of the true tumour hæmangioblastomata. Even here, however, an occasional exception has been recorded (quoted by Lindau) in which two cases of cerebellar hæmangioblastomata occurred in association with skin nævus (type of latter not stated). The angiomatous malformation in such cases with cutaneous nævus is usually of the venous type, although Soutter has met with one of arterial origin. As, however, according to Cushing, venous angiomata tend later in their course to become increasingly aneurysmal, with secondary

arterialisation of the veins, arteriovenous angiomas may be expected to be found occasionally in the patients of more adult years.

Such lesions are said to be always situated above the tentorium. However, cases of both venous and arterial angiomas have been found in the hindbrain, although in these instances no skin naevus was present. The venous malformations may be simple varices or may be widespread racemose congeries of vessels, many of which extend far beyond their focal localization, and while apparently superficial, really penetrate deeply into the brain in a wedge-shaped prolongation, base to the surface and apex terminating near the ventricle, into which a terminal hæmorrhage may occur. This type would be better termed 'cerebromeningeal angioma.' The process may extend also on to the mesial and basal surfaces of the hemisphere. In certain cases—more especially those associated with cutaneous naevus—calcification may occur either in the walls of the vessels, in their thrombosed channels, or in the membranes surrounding them, and may then become visible in an X-ray film. In such instances it may be possible to distinguish the deep penetrating racemose type, in which surgical treatment is out of the question, from the truly superficial variety, where, however, the operative results have been very little better owing to the great vascularity not only of the pial angioma but also of the dura and bone. In some of the cases of the latter type in which operation has been performed, an external hæmorrhage has occurred later through the dura and trephine aperture with the production of a large hæmatoma, as in Bathe Rawling's case, in which the epilepsy was relieved by the operation. Similar angiomas may occur in connection with the spinal meninges, associated in isolated instances with skin naevus of the corresponding metameric surface.

The cutaneous lesion present in this syndrome may be the ordinary 'port-wine stain' or naevus flammeus, or it may also have a more deep-seated cavernous component, when actual hypertrophy of the bones or limb may be seen; it may be pigmented or it may be a naevus anæmicus. The process may involve the buccal or nasal mucous membranes. It may be sharply demarcated to the trigeminal skin area, even more sharply than in zoster, and when it is, according to Cushing's cases, the associated intracranial (meningeal) naevus may topographically correspond, so that in a naevus involving the first trigeminal skin area the corresponding portion of the dura innervated by that branch may be chiefly involved. But it may not be confined to the face, and very extensive skin areas of the entire body may be involved, but in that event one side of the body or face may be predominantly covered; the intracranial naevus is then to be expected on that side. In three cases, however, the meningeal angioma was found at operation or post mortem to be contralateral to the facial cutaneous lesion. In another typical instance with skin naevus and contralateral epilepsy, at autopsy no intracranial vascular malformation was found. Grieg's patient, explored for focal epilepsy, with an angioma venosum extending into the

cortex found at operation, had adenoma sebaceum of the face, a condition which he states to be a cutaneous tumour of bloodvessel origin.

Associated with the cutaneous nævus may be present certain ocular abnormalities in the eye of the affected side. There may be buphthalmos from congenital glaucoma with enlargement of the pupil. Abnormal vascularity may be found in the iris by slit-lamp examination. The retinal veins may be unduly tortuous, the retinal background may be a darker red, while externally the sclera may have a bluish 'china-doll eye' appearance. In a few cases these ocular findings have been associated with intracranial angioma but without any skin nævus. Capillary hæmangioblastoma of the retina (v. Hippel's disease) is not found accompanying cutaneous or intracranial angioma, but is instead a part of Lindau's syndrome, where similar cystic hæmangioblastomata are found usually in the cerebellum, though in Barnard's case it was in the forebrain.

Visual defects of peripheral origin do not occur in the venous angiomata as, at first at any rate, the lesion is not an expanding one, though if by pathological anastomosis it becomes transformed into an arteriovenous (aneurysmal) angioma, then increased intracranial pressure and papilloedema follow. Visual defect of cortical origin has occurred with the present case, in four other instances, and in every one a facial nævus was found and the intracranial lesion shown to be calcified by X-ray examination.

Lastly, mental defects varying from irritability of temper to idiocy have been a marked feature of many of the cases, as is instanced by the number reported from asylums, especially in Brushfield and Wyatt's series. Such mental changes have been ascribed to a gliosis of the cortex or to hæmorrhage from the intracranial nævus, but another possibility seems to be lack of cerebral development, as the X-ray films of many show the affected side to be considerably smaller than its fellow. The latter may also be the explanation of the hemiplegia or epilepsy.