HETEROTOPIC BRAIN TISSUE AS AN ISOLATED EMBRYONIC REST

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A single case history is worthy of report if it presents a previously unrecognized condition or if it sheds further light on unproven concepts of pathogenesis or treatment. The present case seems to fulfill both criteria.

Congenital cutaneous defects of the scalp are quite rare. When they occur they appear as variously discolored areas completely devoid of hair, usually over the vertex of the head. They range in size from circular lesions 1–2 cm. in diameter to large defects embracing both anterior and posterior fontanelles and a narrower area of the scalp between. Usually there is no associated defect in the underlying skull, but in a number of instances such a defect has been present and, in some, there may be a direct connection with a major blood vessel such as the sagittal sinus.

Though less common than those in the scalp, congenital cutaneous defects have been observed in many parts of the body including the trunk and extremities, and such lesions are usually neither bilaterally symmetrical nor localized to the midline. The etiology is unknown and may not be the same in all cases, but the previously accepted theory that they result from inflammatory adhesions between the amnion and body surface has been largely discarded. Whatever their origin they probably differ from the midline defects which occur anywhere from the cranium to the sacrum.

An associated lesion which has recently received wide recognition in British and American literature is the congenital dermal sinus which connects the body surface with the meninges or central nervous system. It is possible that many of the reported cases of isolated cutaneous midline defects may have had such a connection which was overlooked. Dermal sinuses are presumably related to the case under consideration.

In the present instance the lesion was believed, pre-operatively, and even at operation, to be an isolated congenital cutaneous defect without any transcranial connection. The correct diagnosis, derived from microscopic examination of the tissue, was unexpected. A reasonably comprehensive search of the literature revealed no identical case, but there can be no doubt that there is a direct correlation between this and certain previously recognized lesions. The case is therefore of interest with respect to differential diagnosis and management, and it is pertinent to some currently unsolved problems in the fields of embryology and neuropathology.

CASE REPORT

Children’s Hospital #88886. C.W., a 1-year-old white female infant, one of dizygotic twin children (male and female) of a physician, was born March 3, 1952. At birth she was observed to have a flat circular bluish-red lesion in the posterior midline of the scalp, approximately over the lambda. Neurosurgical consultation and detailed roentgenological study of the skull, shortly after birth, led to the conclusion that the lesion was not a meningocele or encephalocele and that there was no collateral defect in the skull or cerebral cortex. No immediate
treatment was advised and the infant's subsequent growth and development were normal in every way, paralleling those of her twin brother.

She was first seen on Feb. 24, 1953, when she was nearly a year old. Her physician-father reported that there had been no evidence of independent growth and that the lesion had increased in size only in proportion to the growth of the child.

Examination. The general appearance of the lesion was as shown in Fig. 1A. It was symmetrical and almost perfectly circular, about 3.25 cm. in diameter, and lay slightly anterior to the lambda. The color was dull red, with a slight pallor at the extreme periphery, and the entire lesion became somewhat cyanotic when the patient cried. It was not spongy or compressible, however, and though it blanched slightly under pressure, the color returned promptly and there was no clinical evidence of excessive vascularity. The growth of hair in the surrounding scalp was normal, and terminated abruptly at the periphery of the lesion, the surface of which was generally flat, with a small nodular elevation 0.5 cm. in diameter near the center. It was covered with a dry epithelium-like surface, without ulceration, tenderness, or evidence of existing or previous inflammation.

Fig. 1. (A) Gross appearance of the lesion prior to operation (see text for detailed description). (B) Appearance of wound on 11th postoperative day, when patient was discharged from hospital.

Aside from this single isolated lesion in the scalp, the physical examination and routine laboratory studies revealed no abnormalities and the child appeared to be perfectly healthy.

Operation was performed on March 20, 1953. The initial step in the procedure consisted of simple excision around the margin of the lesion, leaving a circular defect slightly over 4.0 cm. in diameter. The underlying skull was well exposed, and no demonstrable defect was recognized. Using parallel tangents to the circular wound, double sliding flaps were constructed, slightly wider at their bases, to provide maximum blood supply. After squaring off the central ends of the flaps, they were turned back and a series of parallel horizontal relaxing incisions was made through the galea aponeurotica, from its deep surface. The flaps were then laid back into place and were easily approximated to each other, obliterating the defect and producing an H-shaped scar (Fig. 1B).

Course. Healing was uneventful except that on the 4th postoperative day the child pulled off her dressing, with slight separation of the wound edges in two places. Neither of these re-