

Neuroglial heterotopia of the scalp in an adult

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Sir,

We report a 50 year-old man who presented with a swelling of the occipital scalp which was noticed at birth and increased progressively in size. At physical examination the lesion was firm and alopecia (Fig. 1). Clinically, the diagnosis of benign adnexal tumor was suggested and a surgical resection of the lesion was performed. Preoperatively, the lesion did not adhere to the occipital bone and had no connection with the brain. Grossly, the tumor was nodular, white, ill-defined and measured 1.2 cm of diameter. Histological examination showed a well-circumscribed lesion of the deep dermis composed of mature glial tissue (Fig. 2). It consisted of a dense network of columns and clusters of neural cells within a fibrillar, fibrous and hyaline tissue (Fig. 3). Sections of nerves were also seen. The immunohistochemical study showed a diffuse and intense staining of cells with GFAP (glial fibrillary acidic protein) and S100 protein (Fig. 4). The cytokeratin was negative. These features confirmed the diagnosis of a neuroglial heterotopia. At 3 years of follow-up, the patient was asymptomatic and there was no recurrence.

The patient's informed consent was obtained.

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

Neuroglial heterotopia is a rare, non-hereditary malformative lesion, defined by the presence of an ectopic glial or neuroglial tissue outside the brain. It must be distinguished from meningeal heterotopias that are more frequent and derived from the brain and the spinal cord meninges [1].

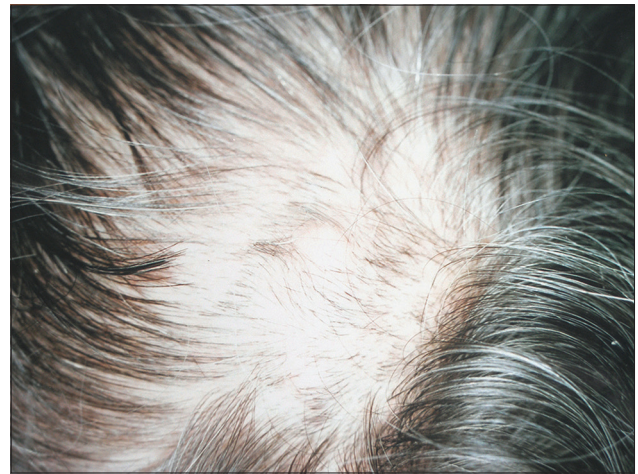


Figure 1: Occipital bald scalp plaque.

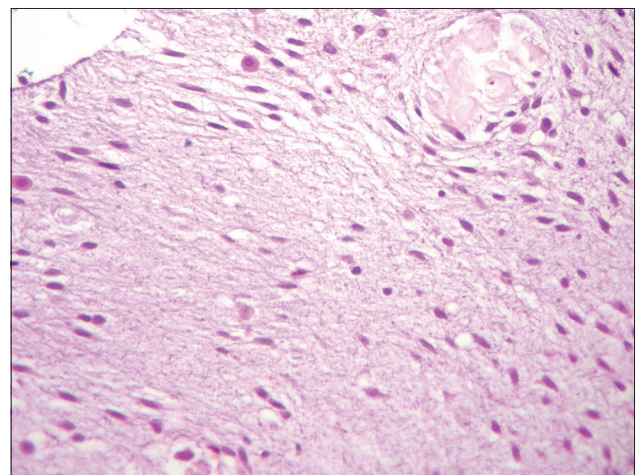


Figure 2: Range of neural and glial cells with eosinophilic fibrillar cytoplasm, and small, rounded nuclei, without atypia and mitosis (HEX40)

This congenital lesion is found mainly in the nose and less frequently on the palate, tongue, orbit, lung and chest wall [1]. The location at the scalp is extremely rare. Only 13 cases were described in the English medical literature.

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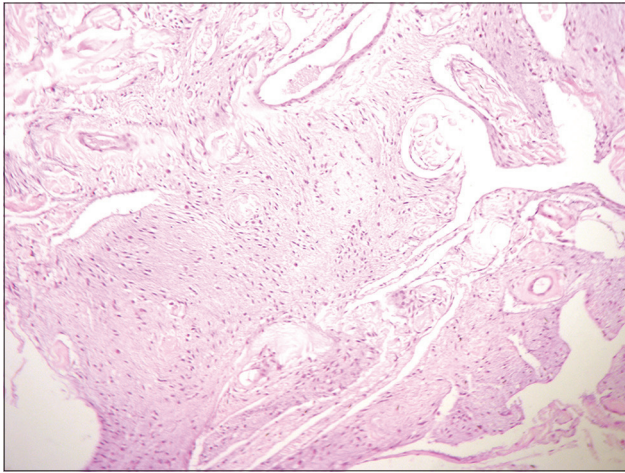


Figure 3: Cells arranged in layers within a hyalinized fibrous tissue (HEX20)

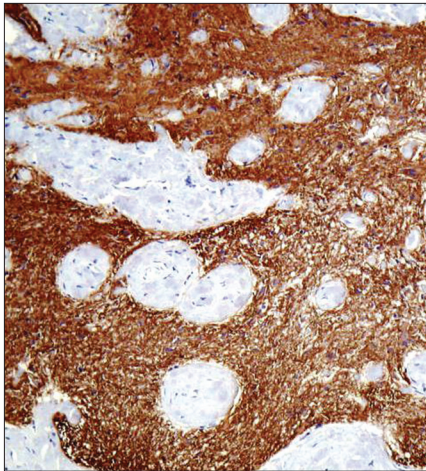


Figure 4: Diffuse and intense staining of neuroglial cells with GFAP

The exact pathogenesis of these lesions is unknown.

Neuroglial heterotopia is usually diagnosed in children, rarely in adults [1].

Clinically, it is a nodular lesion often discovered at birth, measuring 2 and 4 cm in diameter and increasing proportionately with the child's growth. It is solitary, circular, skin-colored, pink or bluish, mobile [1-3].

Grossly, the lesion is firm, nodular and has a grey-white cut surface. Histological examination showed a glial proliferation made of clusters of round or

oval cells, composed mainly of astrocytes sitting in a neurofibrillary and richly vascularized tissue. These cells have central nuclei, without atypia and nucleolei. The cytoplasm is fibrillar and finely granular. Some astrocytes are giant and multinucleated like ganglion cells. An associated neural component or sometimes ependymal or choroid plexus structures can be found. However, purely glial appearance is most frequently described in the literature [2].

The immunohistochemistry with shows positivity of glial component to GFAP (glial fibrillary acidic protein) and S100 protein [1,2].

The main differential diagnosis is with encephalocele which have the same clinical and histological appearance but encephalocele is connected to the sub-arachnoid space by a cavity [1].

The treatment consists of total excision of the tumor. Incomplete excision may cause recurrences. Furthermore, no malignant transformation has been reported in the literature.

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles. Written informed consent was obtained from the patient for publication of this article.

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