Fig. 2. Range of cells with eosinophilic fibrillar cytoplasm, and small, rounded nuclei, without atypia and mitosis (HEX40).

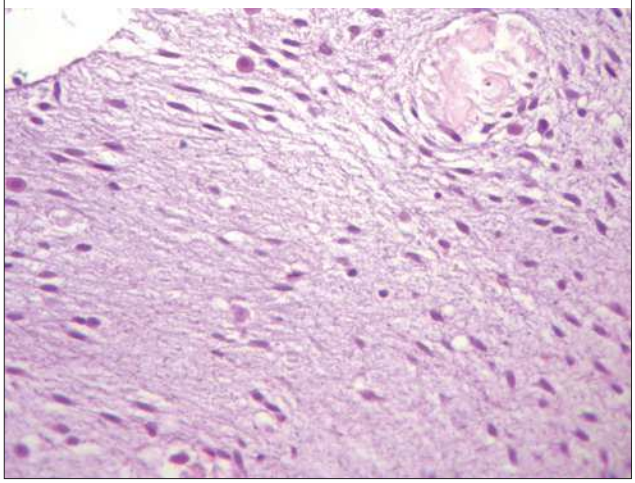
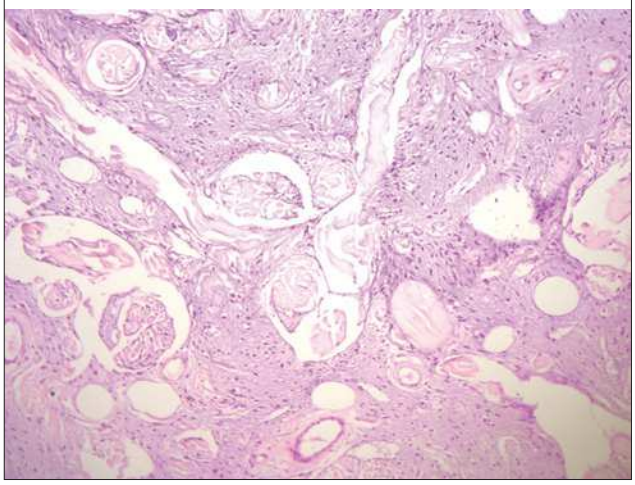


Fig. 3. Cells arranged in layers within a hyalinized fibrous tissue (HEX20).



(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.

Discussion

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

Several other names have been brought to describe this entity: brain heterotopias, astrocytoma, glioma, teratoma and choristoma [2].

The term cerebral heterotopia was first introduced by Lee and Mac Laurin in 1955 to describe the nasal glioma [1].

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. Gray et al reported 2 cases of glial heterotopia of the scalp, in a series of 11 children with heterotopic neural tissue [1].

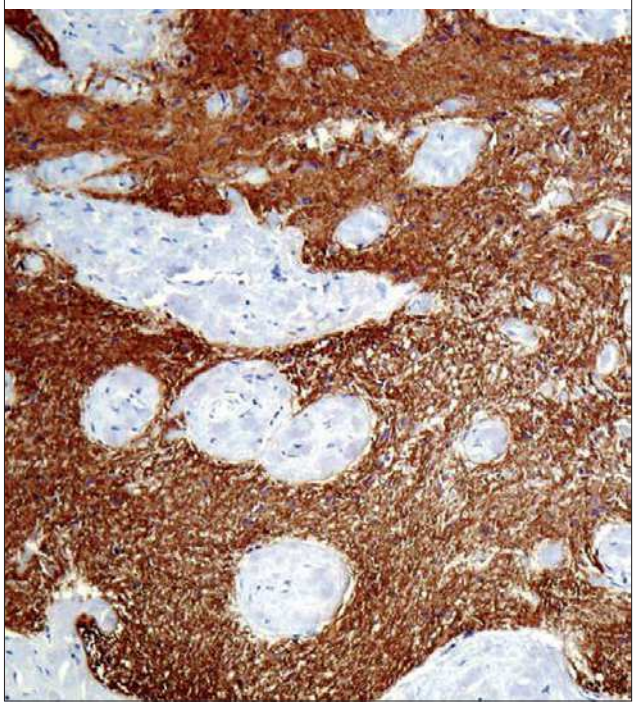
The exact pathogenesis of these lesions is unknown, although hypothetical etiopathogenesis mechanisms have been postulated. First, heteretopic brain may derive from an encephalocele that subsequently loses its communication with the brain [3].An alternative theory suggests that ectopic brain in remote locations may result from sequestration of extracranial embryonic neural tissue [4]. The third hypothesis suggests that heterotopic brain derives from isolated rests of displaced pluripotent neuroectodermal cells [1 5 6].

This lesion is usually diagnosed in children, rarely in adults [1]. It has a preferentially occipital, parietal or sometimes median location. It is a nodular lesion often discovered at birth and increasing proportionately with the child's growth [6]. It is solitary, circular, skin-colored, pink or bluish and mobile. It is most often bald scalp plaque sometimes surrounded by a crown of hair. It measures 2 to 4 cm in diameter [1 5].

The CT scan is a complementary study that is necessary in preoperative planning to determine the extent and location of a mass. On CT scan, a heterogeneous hyperattenuated mass with or without a cyst is a common feature.

Grossly, the lesion is firm, nodular and has a grey-white cut surface. Histological examination showed a glial

Fig. 4. Diffuse and intense staining of neuroglial cells with GFAP.



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Case report

Neuroglial heterotopia of the scalp

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Key words

Glial heterotopia · Scalp

Summary

Heterotopic glial nodules of the scalp are non hereditary congenital malformations composed of mature brain tissue isolated from the cranial cavity. The majority of these lesions are found in the nasal region and occur rarely on the scalp. They are frequently diagnosed in newborn infants. However, they may rarely be found in adults. The pathogenesis of these lesions remains unknown. We describe the case of a temporal scalp nodule in a 50 year-old man. At the time of the excision, the mass was not associated with intracranial connection. Histological examination revealed neural tissue staining with S100-protein and the glial fibrillary acidic protein (GFAP).

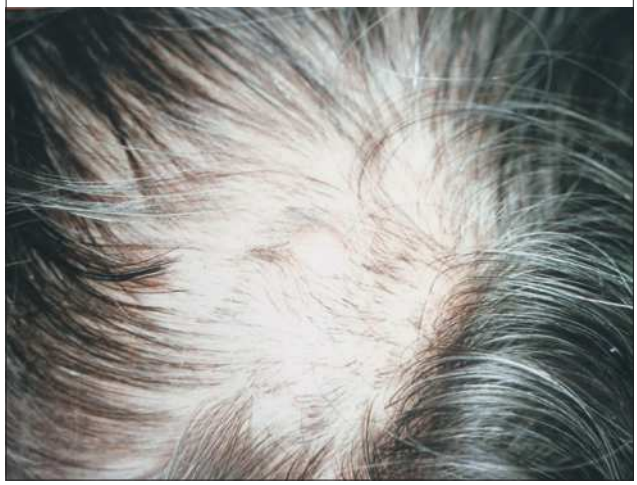
Background

Glial heterotopias are rare, benign congenital lesions, corresponding to the presence of extra cranial ectopic brain tissue without connection to the central nervous system. His pathogenesis is still unclear. They result from abnormal extra cranial sequestration of neural tissue during the embryogenesis. These malformative lesions mostly occur in childhood, before the first year and are often located at the nose. The location at the scalp is rare. Only 13 cases of scalp localized lesions are reported in the English medical literature. We report a rare case of glial heterotopia of the scalp, discovered incidentally in a 50 year-old man.

Case report

A 50 year-old man 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 alopecic (Fig. 1). Clinically, the diagnosis of benign adnexal tumor was made 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, 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. 1. Occipital swelling with a bald scalp plaque.



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epithelial cells suggests the endodermal component is also alterated [14]. In our case, the abnormal interaction between endodermal and mesodermal components of lung buds has happened at a specific time point, presumably around the thirteenth week of embryonal life, which corresponds to the pseudo glandular stage just before the establishment of connections between mesodermal plexus and pulmonary circulation. So, according also to the recently proposals for the classification of pulmonary malformations [1], CPAM would not be an hamartomatous lesion and the classification proposed by Stocker [3] based on the apparent "site of maldevelopment" might be incorrect. The right classification criteria may consider the time rather than the site of malformation. This new type of approach based on the temporal aspects of the endoderm/mesoderm interaction, suggested also by Clements et al [15], could lead to a more modern and correct classification of all types of CTMs.

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