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Subcutaneous Pediatric Nasal Glial Heterotopia

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Dear Editor,

The space-occupying masses of the nose, nasal cavity, and nasopharynx in childhood still pose a diagnostic challenge and, consequently, a challenge to the appropriate type of treatment. These topographical regions are the sites of widely different pathologies, resulting in very different therapeutic approaches: congenital and developmental disorders (congenital nasolacrimal duct mucocele, dermoid cyst, encephalocele, and nasal glial heterotopia (NGH)); inflammatory/infectious processes (mucocele, polyp, and pyogenic granuloma); benign neoplasms (infantile hemangioma and juvenile nasopharyngeal angiofibroma); malignancies (rhabdomyosarcoma and nasopharyngeal carcinoma); and masses related to previous trauma.[1] If we focus more specifically on the study of congenital developmental midline nasal masses, the differential diagnosis certainly narrows but remains open to several entities, including nasal dermoids, encephaloceles, and NGH.[2] The following case we diagnosed and treated fits into this context.

A 5-year-old child presented with a subcutaneous mass on the bridge of the nose, since birth. Physical examination revealed a firm, non-tender, non-pulsatile mass. Brain magnetic resonance imaging (MRI) ruled out central nervous system involvement (Figure 1A), and surgical intervention was planned.

Gross examination revealed intact skin covering the lesion (Figure 1B). Microscopy revealed diffuse dermal-hypodermal fibrosis interspersed with fascicles of loose, mildly hypercellular neural tissue consisting of mature astrocytes and neurofibrillary processes. Importantly, there was no significant atypia or mitotic activity (Figure 1C). Immunohistochemistry (IHC) was performed to further characterize the lesion. Neural fascicles were positive for glial fibrillary acidic protein (GFAP, Figure 1D), S100, cytokeratins AE1/AE3, and beta-catenin, and negative for smooth muscle actin, desmin, epithelial membrane antigen (EMA), CD31, and CD34. The Ki67 proliferation index was less than 5%, supporting the diagnosis of NGH. Additional IHC for isocitrate dehydrogenase (IDH1-R132), p53, and BRAF were also negative, consistent with a wild-type profile typical of non-neoplastic glial tissue. Indeed, NGH is a congenital entity (choristoma) characterized by ectopic mature glial tissue outside the neurocranium, the pathogenesis of which is not fully understood.

To date, just over 150 cases of NGH have been described,[3] although not all have been histologically documented,[4] and only 8% have been diagnosed in adults. In fact, NGH occurs primarily in pediatric patients and can be classified by location: 60% are extranasal (often subcutaneous), 30% intranasal (in the upper nasal cavity), and 10% mixed.[5] Subcutaneous NGH may radiologically or clinically resemble a dermal neoplasm, as in the case presented here, necessitating histology to determine the glial nature of the lesion. In addition to its diagnostic purpose, surgery is the therapeutic treatment of choice for NGH, although lesions that are not completely excised may recur.

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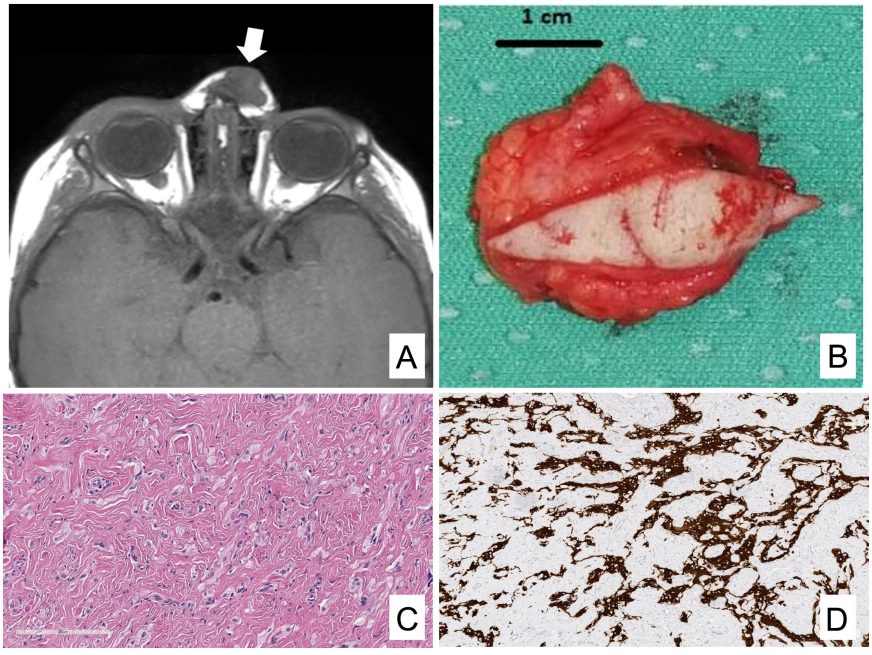

Figure 1. (A) Axial T1-weighted MR image shows a smoothly marginated mass (arrow) at the root of the nasal pyramid, without any connection to the central nervous system; (B) macroscopy, showing a bulging mass but with an intact skin surface; (C) photomicrograph showing a dense fibrous dermal layer and revealing the presence, in the context of fibrosis, of neural/glial fascicles without significant cellular atypia and/or mitosis (H&E, magnification ×40); (D) immunohistochemistry showing clear positivity for GFAP, supporting the glial nature of the lesion and, together with other findings, the diagnosis of NGH (magnification ×20).

This case highlights the importance of considering NGH in the differential diagnosis of nasal masses in pediatric patients and emphasizes the need for a thorough diagnostic approach, including neuroimaging and histopathologic/IHC evaluation.

Availability of Data and Materials: The data that support the findings of this study are available on request from the corresponding author.

Informed Consent: Verbal informed consent was obtained from patient's parents who agreed to take part in the study.

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