Xanthogranuloma of the Anterior Skull Base: A Case Report

ABSTRACT—A case of a xanthogranuloma arising in the anterior skull base involving the bone of the left orbital roof, the dura mater, and the periorbit is presented. The lesion was completely removed by a supraorbital “eyebrow” skin incision using frameless stereotactic image guidance. A reconstruction of the orbital roof was performed using a titanium mesh graft. The neuropathological investigation revealed all the characteristics of a xanthogranuloma. Intracranially, xanthogranulomas appear as rare tumors of the dura or choroid plexus, usually arising in association with histiocytosis X or familial hyperlipoproteinemia. One case not associated with these diseases is discussed.

Xanthogranulomas involving the central nervous system are rare and may be the manifestation of a variety of pathologic processes. The exact etiology is not fully understood, whether neoplastic, inflammatory, or metabolic remains still a matter of debate. Reports concerning symptomatic xanthogranulomas usually involve the choroid plexus, presenting with hydrocephalus or involving the dura mater, showing neurological deficits due to mass effect. Although such lesions usually occur in association with histiocytosis X or familial hyperproteinemias, this case was not associated with these diseases.

CASE REPORT

History

A 39-year-old man presented with a 6-month history of headache and progressive enlargement of the left supraorbital rim. During the last month the patient had experienced intermittent diplopia and impaired vision in the left eye. His family medical history was negative.

Examination

Physical examination revealed a protrusion of the left bulbus and a reduction of vision down to 40% in the affected eye. Computed tomography (CT) and magnetic resonance imaging (MRI) scans of the head showed a 2 × 2 × 1 cm non-contrast-enhancing mass, enlarging the left supraorbital rim and compressing the orbital gyurses and the ocular globe (Fig. 1). The periorbital perioisteum appeared to be intact, and the dura mater was not clearly identifiable. An infiltration of the brain was not seen, but the orbital roof and the zygomatic process of the frontal bone were destroyed, apparently due to prolonged pressure. The lateral wall of the frontal sinus was intact. A T2-weighted MRI demonstrated a high liquid content in this mass (Fig. 1C, D).

Operation

A preoperative CT using skin fiducials was performed the day before surgery, and the approach was planned on a computer workstation of the Zeiss SMN Skull Base Surgery, Volume 8, Number 4, 1998 Department of Neurosurgery (K.R., K.N., C.M.), Institute of Neurology (C.J.), and Department of Neuroradiology (H.G.), University of Vienna Medical School, Vienna, Austria. Reprint requests: Dr. Karl Roessler, Department of Neurosurgery, University Hospital of Vienna, Waehringer Guertel 18–20, A-1090 Vienna, Austria. Karl.roessler@telecom.at Copyright © 1998 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel.: +1 (212) 760-0888. All rights reserved.
frameless stereotactic navigating microscope (Zeiss, Oberkochen, Germany). The margins of the lesion were defined on the images and projected as a contour onto the arrested head in the operating theater immediately before surgery, with the help of the contour-planning mode of the microscope. Using this system for operation planning, a small eyebrow incision was deemed appropriate to expose the involved portions of the frontal skull base and orbit on the left side (Fig. 3A). After the thinned supraorbital rim with 2 cm of frontobasal bone had been removed, a lobular, firm yellow mass was encountered, encapsulated within a layer of smooth, hard fibrous tissue in a bony bed (Fig. 3B). The bone of the orbital roof was totally replaced by the lesion, and one layer of the frontobasal dura mater as well as the periorbit were within the capsule. By gentle dissection, the lesion could be removed from both the dura as well as the periorbit and from its bony bed in the os frontale, which was not infiltrated but showed marginal hyperostosis due to the pressure of the lesion. A reconstruction of the orbital roof was performed by using a titanium mesh graft and a galea-periosteal flap (Fig. 3C).

Postoperative Course

The postoperative course was uneventful. The protrusion of the left bulbus as well as vision normalized within the next 4 weeks. By this time postoperative swelling had resolved and the patient was able to return to work. No evidence of regrowth of the tumor was found 5 months after surgery (Fig. 2A–E).

Pathologic Examination

Histopathologic examination of the tumor showed a proliferation of foamy histiocytes, multinucleated giant cells, and cholesterol clefts. These were interspersed with hemosiderin deposits within histiocytes and in the intercellular space. Additionally, an infiltration of inflammatory cells throughout the lesion was seen. There was no evidence of proliferation of meningothelial cells (Fig. 3D). These characteristics were entirely consistent with those of xanthogranulomas.
Intracranial xanthogranulomas are rare and occur in a variety of pathological conditions, most commonly as choroid plexus xanthogranulomas or as tumors in the dura mater with similar histological features.

Choroid plexus xanthogranulomas normally present with symptoms of obstructive hydrocephalus and are thought to arise from desquamated epithelial cells migrating into the choroid plexus, which release lipids, provoking the infiltration of inflammatory cells. In contrast, dural xanthogranulomas present with signs and symptoms of a mass lesion and are usually very large at the time of diagnosis. In contrast to reported cases of purely intracranial xanthogranulomas, in our patient the fronto-orbital location lead to an early manifestation of ocular protrusion, which permitted an early diagnosis of the small lesion (2 × 2 × 1 cm).

In our patient the lesion involved the frontobasal dura, leading to chronic pressure and thinning of the orbital roof and the adjacent frontal bone. In contrast to plexus xanthogranulomas, dural xanthogranulomas are...
usually associated with histiocytosis X or familial hyperlipoproteinemia. Lipotrophic factors within the putative autoimmune process of histiocytosis X are thought to stimulate mesenchymal cells in the dura, causing them to undergo xanthomatous transformation. The reported case occurred without a background of histiocytosis X or hyperlipoproteinemia, and we found no clinical, radiological, or laboratory signs of these diseases.

Another differential diagnosis includes Erdheim-Chester’s disease, a systemic disease with lipogranulomatous lesions of the viscera and bones, characterized by foamy lipid-containing histiocytes. However, a careful internal investigation including ultrasound and skeletal radiographs of the long bones failed to demonstrate additional lesions characteristic for this disease.

An alternative pathogenesis described is the posttraumatic formation of xanthogranulomas within the involved tissue components. In our patient there was no history of trauma, although the histological investigation revealed hemosiderin deposits within the foamy histiocytes, not typical for xanthogranulomas of other than traumatic origin.

In summary, the reported case of a xanthogranuloma in the frontal skull base seems to represent a rare case of a intracranial xanthogranuloma without association with a systemic disease, such as histiocytosis X, hyperlipoproteinemia, or Erdheim-Chester’s disease, as previously reported. Although the described histology would support a posttraumatic origin, there is no clinical or radiological evidence of previous trauma.
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REFERENCES