ANTHOGANULOMA is a disorder of histiocytes characterized by solitary or multiple yellow-red nodules on the skin. It is predominantly a disease of infancy or early childhood, but may also occur in adults. Various organs can be affected, and the central nervous system is occasionally involved in systemic xanthogranulomatosis or histiocytosis-related conditions. Isolated lesions limited to the intracranial compartment commonly appear in the ventricular system (intra-ventricular type). These lesions arise from the choroid plexus, and sometimes cause signs of increased intracranial pressure due to obstructive hydrocephalus. However, a solitary nonsystemic xanthogranuloma located in another intracranial region is extremely rare; only six reports have been found in the literature, including xanthogranuloma in the middle fossa, sellar region, cerebellopontine angle, Meckel cave, and intraparenchymal temporoparietal lobes. We report the first case in which a symptomatic solitary xanthogranuloma has been documented to occupy the interdural space of the CS with no systemic manifestation.

Case Report

History. This 42-year-old woman presented to an ophthalmology clinic on August 28, 1999 with decreased visual acuity on the right side. On examination, her visual acuity was found to be 10/20 on the right and 16/20 on the left side. Mild exophthalmos and papilledema were observed in the right eye. Goldmann and Hufnry visual field perimeters disclosed an inferomedial isoptor depression of the right side. She was referred to our clinic to rule out the presence of intracranial lesions.

Examination. The patient was alert and did not report double vision, abnormal facial sensations, or headache. Her extraocular movements were full bilaterally and her pupils were isoric and reactive. The corneal reflex was prompt on both sides. Thus, no neurological deficit was noted except for the visual disturbance on the right side. Skull x-ray films demonstrated an enlarged right superior orbital fissure, and CT scanning revealed a low-density oval mass in the right cavernous region. This mass was...
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2 × 3 cm in diameter and extended into the right orbit through a dilated superior orbital fissure, compressing the right optic nerve in the superolateral to inferomedial direction (Fig. 1A). Marked contrast enhancement was noted in the margin (Fig. 1B). The size, location, and extension pattern of the lesion were clearly visible on a reconstructive three-dimensional CT scan (Fig. 1C). The tumor exhibited isointensity on T1-weighted (Fig. 2A) and mixed-pattern hyperintensity on T2-weighted (Fig. 2B) MR images, with typical smooth contours. Contrast enhancement with gadolinium–diethylenetriamine pentaacetic acid was homogeneous, but most prominent in the margin of the lesion (Fig. 2C and D). The intracavernous portion of the right ICA was mildly compressed toward the medial direction without encasement or narrowing.

Operation. To decompress the right optic nerve and to obtain a sample for histopathological confirmation, on October 4, 1999, the lesion was partially removed using both extra- and intradural approaches through a right-sided frontotemporal craniotomy with zygomatic osteotomy. The exposed soft, yellow mass, which was confined to the dura propria, was easily removed using an ultrasonic aspirator. The oculomotor nerve and the first and second divisions of the trigeminal nerve were involved in the lesion. The ICA was not exposed during the operative procedures. To avoid neurological deterioration, the cranial nerves were carefully preserved and most of the lesion was left in place. Histopathological examination of tumor samples revealed the characteristic features of xanthogranuloma: homogeneous foamy cells containing Sudan III–positive granules in conjunction with slight infiltration of inflammatory cells (Fig. 3A and B). For the immunohistochemical study, antibodies to human lysozyme (1:200), human KP1 (1:50), and bovine S-100 protein (1:300) (Dako Corp., Glostrup, Denmark) were used. The foamy xanthomatous cells were immunopositive for lysozyme (not shown) and KP1 (Fig. 3C), but negative for S-100 protein (Fig. 3D).

Postoperative Course. After surgery the patient was alert, but suffered from transient mild deficits in the right oculomotor nerve and the frontal branch of the facial nerve. No radiation therapy or chemotherapy was administered. Two months postoperatively, her visual acuity on the right side had improved to 20/20, accompanied by a
full recovery of the visual field. Postoperative MR images demonstrated that the anterior part of the mass had been partially removed, resolving the distortion of the right optic nerve.

Although a moderate degree of hypercholesterolemia (299 mg/dl) was seen on admission, the patient had no history indicative of familial hypercholesterolemia. Her serum cholesterol level spontaneously decreased to the normal range (171 mg/dl) during 3 weeks of hospitalization. No other xanthomatous lesions were detected anywhere in her body.

**Discussion**

Xanthogranuloma represents an accumulation of histiocytes that lack Birbeck granules. It is still controversial whether xanthogranulomas should be considered true neoplasms or reactive granulomatous proliferations that form in response to unknown stimuli. Currently, xanthogranuloma is categorized as histiocytosis Class II, which is defined as histiocytosis of mononuclear phagocytes other than Langerhans cells. Recent immunohistochemical studies have indicated that xanthogranulomas have macrophagic differentiation based on their expression of variable histiocytic markers, including CD68 (KP1), HAM56, cathepsin B, vimentin, KiM1p, and factor XIIIa, although they are negative for S-100 protein. These findings are consistent with the immunohistochemical results in the present case.

Xanthogranulomas have been further divided into two categories, that is, AXG and JXG, but significant histological discrimination between these two subgroups is complicated. A recent study in which JXG and AXG were compared using light microscopy and immunohistochemical studies demonstrated that giant cells were more prominent in AXG than in JXG; oncocytic cells, characterized by some eosinophilic cells and mostly periodic acid–Schiff–negative giant cells, were not observed in classic JXG. Within this context, the CS xanthogranuloma in our case may correspond to JXG based on its histological features, including the absence of giant cells and lower inflammatory cell infiltration.

Whereas xanthogranuloma is classified as a normolipemic histiocytosis, cases of several patients with multicentric intracranial lesions related to familial hyperlipoproteinemia IIa have been reported. Although hy-
percholesterolemia was observed in our patient, her relatives had never been affected by familial hyperlipidemia. Moreover, her high level of serum cholesterol on admission decreased to within the normal range during her hospital stay, strongly suggesting that our patient’s hypercholesterolemia may merely have been due to her inadequate diet.

Neuroimaging and intraoperative findings indicated that the xanthogranuloma in this case was confined to the interdural space of the lateral wall of the CS. Results of preoperative CT, MR, and angiography studies of the lesion closely mimicked those associated with trigeminal neuroma. El-Kalliny, et al.,3 have classified tumors of the CS into three types and defined interdural CS lesions as those remaining between the outer dura propria and the inner membranous layer. These tumors have commonly included trigeminal neuromas, and, less frequently, oculomotor and trochlear neuromas, epidermoids, melanomas, and cavernous angiomas. Various combinations of cranial nerve signs have been present in these interdural tumors.3 In contrast, the neurological symptoms in our patient were unexpectedly mild. The less serious neurological deficits in the present case may be partly accounted for by the extreme softness of the xanthogranuloma. Nevertheless, two patients previously reported to have similar isolated xanthogranulomas in the Meckel cave15,24 suffered from neuroma. El-Kalliny, et al.,5 have classified tumors of the CS xanthogranuloma15,24 suffered from neuroma. El-Kalliny, et al.,5 have classified tumors of the CS xanthogranuloma in Hand-Schüller-Christian disease. Case report. J Neurosurg 77:508–514, 1992


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