Cholesterol granuloma of the petrous apex

Case report

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The authors report a cholesterol granuloma that originated from the sphenoid sinus, eroded through the petrous apex, and presented as a tumor of the cerebellopontine angle. The differences between these rare tumors and cholesteatomas are discussed.

KEY WORDS  epidermoid tumor  cholesteatoma  cholesterol granuloma  cerebellopontine angle tumor

A true cholesterol granuloma of the head is rare and usually presents in the middle ear or mastoid sinus. Our review of the literature suggests that such tumors rarely exhibit neurological signs. We are reporting such a case.

Case Report

The patient was a 57-year-old woman who, 2 years prior to admission, had 2 to 3 days of transient episodes of tinnitus in the right ear associated with a gait that veered to the right. The symptoms recurred 1 year and again 3 months before admission, but this time they were accompanied by intermittent right hemifacial spasm, diplopia on right lateral gaze, right ocular pain, and intermittent "shooting" pains in the right forehead.

The patient was admitted to another hospital where the following evaluation was made. Skull films with laminograms revealed erosion of the right petrous apex and a soft tissue mass in the sphenoid sinus. Angiography demonstrated that the right carotid artery was circumferentially compressed at the foramen lacerum, and that the right posterior cerebral and superior cerebellar arteries were slightly elevated. She was placed on Decadron. A transnasal biopsy of the sphenoid sinus was undertaken, which showed only normal sinus mucosa; following this, the pain diminished dramatically. The patient's serum cholesterol was noted to have changed from 276 mg% in 1971 to 370 mg% at the time of the biopsy 2 years later.

Examination. On admission to the University of Washington Hospital, neurological examination revealed only a right sixth nerve paresis. Complete blood count, electrolytes, blood urea nitrogen, and VDRL were normal. A pneumoencephalogram combined with upright polytomography showed a mass extending from the petrous apex posteriorly and surrounded by an extremely thin rim of calcium.
The fourth ventricle was displaced slightly posteriorly and to the left. Cerebrospinal fluid (CSF) pressure, chemistry, and electrophoresis were normal. A brain scan with $^{99}$Tc pertechnetate showed increased uptake in the right posterior fossa. Audiometrics revealed normal thresholds and adaptation with a slight bilateral high frequency sensorineural loss greater on the right. Electronystagmography showed that response to $30^\circ$ C irrigation on the right was 37% less than that on the left.

**Operation.** Using a right subtemporal transtentorial approach we exposed an extradural tumor. The tumor capsule was entered between its encroachments onto the fifth, seventh, and eighth cranial nerves. The total intracapsular contents of yellowish avascular "cheesy" material were apparently completely removed. Postoperative recovery was uneventful. The diplopia subsided in a few weeks.

**Pathology.** Most of the tumor consisted of amorphous necrotic debris. There was no evidence of squamous epithelium or keratin, but one section showed a small amount of well-differentiated columnar epithelium. Numerous fragments of fibrous tissue included cholesterol clefts surrounded by multinucleated giant cells (Fig. 2). Sections stained for bacteria, acid-fast bacilli, spirochetes, and keratin were negative. The presence of giant cells, fibroblasts, macrophages, chronic inflammatory cells and copious cholesterol crystal clefts was characteristic of a cholesterol granuloma.

**Discussion**

This case is a typical cholesterol granuloma, although in an unusual location. Friedmann produced cholesterol granulomas experimentally in the middle ear of guinea pigs by installation of a sterile suspension of cholesterol. Cholesterol granulomas would not form if the suspension became infected. Paparella and Lim suggested that sterile collections of blood within the middle ear were the naturally-occurring correlate of Friedmann's experimental model. The mechanism for the spontaneous development of...
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granulomas within the sinus was suggested by Beaumont's demonstration\(^2\) that obstruction of the airways in the pneumatized head of the humerus in chickens caused submucosal edema (possibly as a result of negative pressure) with subsequent inflammation, lipid deposition from degenerating cells, and, in 2 or 3 months, a cholesterol granuloma. These studies have emphasized the role of cholesterol in the production of granulomatous tumors, but this may not be the only factor since cholesterol also is found in a variety of other tumors (craniopharyngioma, epidermoid, desmoid) without the additional progressive granulomatous reaction. Thus the crucial factors for the production of a cholesterol granuloma seem to be the obstruction of an air cell or sinus, without secondary infection.

Cholesterol granulomas appear to share some common pathogenic features with the “acquired” epidermoid tumors of the middle ear, although they are histologically distinct. Acquired epidermoids appear to arise secondary to migration of stratified squamous epithelium through a ruptured tympanic membrane. The predisposing factors for both acquired epidermoids and cholesterol granulomas are similar, namely, chronic upper respiratory infection with inflammatory block to the Eustachian tube and secondary otitis media.\(^4\),\(^5\),\(^6\),\(^7\) The resulting histological appearance seems to be largely dependent on whether the tympanic membrane remains intact. Rupture of the drum allows migration of the stratified epithelium into the middle ear producing an “acquired cholesteatoma.” When the drum remains intact, the usual outcome is a cholesterol granuloma.

Indeed the term “cholesteatoma” has been used to describe four pathologically distinct entities: 1) cholesterol granulomas, 2) acquired epidermoids, 3) intradural epidermoids, and 4) craniopharyngiomas. The distinction between cholesterol granulomas and epidermoids has already been mentioned. Craniopharyngiomas, as is well known, are thought to arise from rests of squamous epithelium derived from embryonic endodermal lining of the stomodeum, the precursor of mucosal epithelium. By contrast, the intracranial epidermoids\(^1\),\(^6\),\(^7\) seemed to arise from embryonic ectodermal rests of squamous epithelium as precursors of epidermis. Thus, these tumors can be distinguished histologically. In the epidermoid, epithelium is characterized by dry keratin, which has a wiry appearance unless the thin flat hexagonal cells are cut tangentially, plus a stratum granulosum as in skin. Craniopharyngiomas demonstrate “wet keratin;” keratohyalin-containing cells appear globoid in any cut section, and, as in oral mucosa, there is no stratum granulosum. This confusion of four pathologically distinct entities under the title of “cholesteatoma” suggests that this is a term to avoid.

In our patient, the cholesterol granuloma was probably the result of temporary obstruction of a portion of the sphenoid sinus or air cells in the medial petrous bone. Enlargement of the granuloma to destroy the petrous apex was most likely due to pressure, although production of osteolytic material by these granulomas has also been suggested.\(^3\) There was little or no evidence of bleeding or inflammatory origin. This patient’s prognosis can only be extrapolated from what is known about the more common granulomas within the attic or the middle ear where slow recurrence is inevitable unless the entire mass of cholesterol is removed.\(^5\)

The avascular nature of this granuloma simplifies total removal even in locations as in our tumor where exposure is limited. Indications for removal include correction of existing neurological deficits and prevention of further enlargement. The prompt reversal of these deficits following removal of this cholesterol granuloma suggests that they are secondary to pressure from the enlarging granuloma.

References


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