Introduction

The term pseudotumor refers to an idiopathic, inflammatory lesion which most commonly presents in the orbit. This non-specific collection of inflammation may represent 5 to 16% of orbital masses in adults. In rare instances, idiopathic pseudotumor (IP) may be found outside of the orbit and may present as a skull base lesion. Reported sites of involvement include multiple areas of the skull base, parapharyngeal space, nasal cavities, and paranasal sinuses. Diagnosis and treatment of pseudotumor of the skull base can be challenging, especially when these lesions involve atypical anatomic locations. We describe the clinical manifestations of an extraorbital pseudotumor of the petrous apex. Diagnosis of this condition often requires tissue biopsy, and recent advances in endoscopic techniques allow surgeons to obtain tissue samples with less morbidity than with open surgical techniques.

Case Report

A 50 year old previously healthy female presented with worsening left sided headaches and double vision. Physical exam revealed significant vision loss involving the left eye and limitation of left lateral gaze. No other focal neurologic deficits were found. A contrast-enhanced CT scan was performed showing a lytic lesion of the left petrous apex. A follow up MRI scan further delineated the approximately 2x2 cm enhancing lesion of the left petrous apex and cavernous sinus without orbital involvement (Fig. 1 and 2). Laboratory studies showed only nonspecific inflammatory markers and a negative autoimmune workup. Because of the findings of a lesion on imaging studies, a biopsy was planned for diagnostic purposes.

Consideration was given to various surgical techniques that could be used to access the petrous apex. Ultimately, a transnasal endoscopic approach was used. A wide sphenoidotomy was performed (Fig 3). The petrous apex was accessed by exposing the area just posterior to the anterior segment of the left internal carotid artery (Fig. 5). The bone in this area was removed using a drill, and dissection proceeded posterior and lateral to the internal carotid artery. As the petrous apex was entered, friable soft tissue was appreciated (Fig. 4 and 6). Biopsies were taken and cultures were sent for possible infectious etiologies.

Pathology revealed “atypical lymphoid infiltrate consistent with idiopathic orbital inflammation” (Fig 7 and 8). These findings were consistent with pseudotumor, and additional tapering steroids were used with further improvement with vision. After a Rheumatology evaluation, the patient was placed on a steroid sparing regimen that included Mycophenolate mofetil (Cellcept™). Her visual symptoms have resolved; she has normal vision and has no limitation in her extraocular movements.

Discussion

This report describes a case of pseudotumor involving the petrous apex and cavernous sinus. The most common lesions of the petrous apex include cholesterol granuloma, followed by cholesteatoma. A variety of other pathologic entities are also possible, but pseudotumor involving the petrous apex has not previously been described.

Clinical manifestations of pseudotumor of the skull base are variable. In the case presented here, the patient had vision loss and ophthalmoplegia due to the involvement of the left optic nerve and abducens nerve. However, a variety of cranial neuropathies can exist based on the anatomic sites involved. Skull base pseudotumor should be a part of the differential diagnosis of patients with cranial neuropathies and imaging findings suggesting a skull base mass.

The imaging findings for skull base pseudotumor are not pathognomonic. Therefore, surgical biopsy is usually indicated in order to differentiate pseudotumor from entities such as neoplasms. In areas such as the petrous apex, performing surgical biopsy can be difficult. Access to the petrous apex has typically been obtained by way of open craniotomy or transmastoïd approaches. As this report illustrates, a transnasal endoscopic approach may allow access to the petrous apex in appropriate cases. The endoscopic approach provides excellent visualization and may be associated with less morbidity than the traditional surgical approaches.

Conclusion

Pseudotumor is an idiopathic, inflammatory lesion rarely found in extraorbital locations. This entity is a poorly understood inflammatory process. Extraorbital involvement may extend into the skull base, temporal bone and intracranial areas. Endoscopic advances allow management of lesions in areas such as the petrous apex with improved visualization and decreased morbidity.

References