Surgical Management of Sclerosing Idiopathic Orbital Inflammation

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Abstract

Here we describe a case of sclerosing idiopathic orbital inflammation (SIOI) treated with endoscopic biopsy and resection and introduce SIOI into the otolaryngology literature as a rare subset of orbital inflammation and potential cause of orbital apex mass that should be recognized by otolaryngologists in order to provide appropriate management.

We present the case of a 59-year-old female who presented with recent history of left-sided complete visual loss, ophthalmoplegia, proptosis, ptosis, retroorbital pain, and decreased sensation in V1/V2 distributions. Imaging revealed a left orbital apex soft tissue mass involving the cavernous sinus and optic canal. She underwent endoscopic biopsy and orbital and optic nerve decompression. Pathology was consistent with SIOI.

Otolaryngologists should be aware of SIOI, a disease entity largely unaddressed in the otolaryngology literature, as a cause of orbital apex syndrome and orbital mass. Suspicion of SIOI should prompt a timely diagnostic biopsy and initiation of aggressive therapy.

Introduction

Sclerosing idiopathic orbital inflammation (SIOI), also referred to as sclerosing orbital pseudotumor, is a rare disease that falls under the larger category of idiopathic orbital inflammation causing replacement of orbital structures with fibrotic infiltrates. It makes up roughly five percent of cases of orbital inflammation.

It has a chronic course with gradual progression, and most often affects the superior and lateral aspects of the orbit, including the lacrimal gland. However, potential for a more aggressive course has been documented with spread into the infratemporal fossa, pterygopalatine fossa, cavernous sinus (as our case demonstrates), and intracranial in rare cases. It can be difficult to diagnose early on due to its insidious onset. Signs and symptoms include proptosis, ptosis, inflammation, pain, double vision, and restriction of extraocular movements. The site of inflammation largely dictates the associated clinical findings, with posterior inflammation causing visual disturbance due to optic nerve compromise and proptosis, and anterior inflammation causing more superficial pathology such as chemosis or lid edema.

Biopsy is critical for diagnosis, showing significant fibrosis with thinly distributed mixed chronic inflammatory cell infiltrate. There are currently no established treatment guidelines secondary to the rare nature of the disease and lack of larger numbers of consistent positive outcomes. It has been traditionally treated with steroids in the presence or absence of radiation therapy with mixed outcomes. Successful treatment has been reported with the use of immunomodulators, such as azathioprine or cyclophosphamide, but only in small case series. Surgical debulking has also established a place in treatment regimens with multiple successful cases reported in the literature.

Methods and Materials

We present a case report of a 59-year-old female who presented with recent history of left-sided complete visual loss, ophthalmoplegia, proptosis, ptosis, retroorbital pain, and decreased sensation in V1/V2 distributions. Imaging revealed a left orbital apex soft tissue mass involving the cavernous sinus and optic canal. She subsequently underwent endoscopic biopsy and orbital and optic nerve decompression.

Results

The patient’s pathology was consistent with SIOI. Postoperatively the patient’s pain and proptosis significantly improved and she could perceive indeterminate light. However, her ophthalmoplegia remained unchanged.

Discussion

While orbital disease is typically managed by ophthalmologists, orbital apex and retroorbital space pathology often require collaboration from neurosurgical and otolaryngologic specialists. With the development of endoscopy and advanced imaging, skull base surgeons can manage orbital apex and periorbital skull base lesions with minimal morbidity. Otolaryngologists should be aware of SIOI, a disease entity largely unaddressed in the otolaryngology literature, as a cause of orbital apex syndrome and orbital mass.

Suspicion of SIOI should prompt a timely diagnostic biopsy and initiation of aggressive therapy. As the disease is still poorly understood, treatment guidelines are lacking. Therapy includes a combination of steroids, radiation, immunomodulators, and surgery, all with limited success. These patients should be managed in a multidisciplinary manner. Surgically, biopsy and therapeutic resection/decompression may play valuable roles in treatment of SIOI.

Conclusions

Sclerosing idiopathic orbital inflammation is a rare cause of orbital apex syndrome and orbital mass. It is currently largely unaddressed in the otolaryngology literature, but something otolaryngologists should be aware of as successful treatment requires prompt a timely diagnostic biopsy and initiation of aggressive therapy with a potential role of surgical debulking.

References


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