Inflammatory pseudotumor of the paranasal sinuses and anterior skull base: A unique case of resolution with radiotherapy

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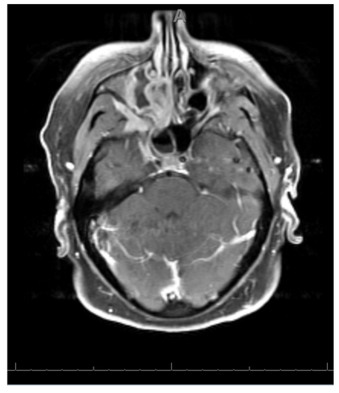
Background

Inflammatory pseudotumor is a rare, benign inflammatory process of uncertain etiology. While it typically presents in the 5th decade, inflammatory pseudotumor has been described in both infants and octogenarians. Inflammatory pseudotumor most commonly affects males (3:2). Pseudotumor of the skull base tends to be relatively aggressive and most commonly involves the anterior cranial base (70%). The diagnosis of inflammatory pseudotumor requires tissue biopsy, as the clinical and radiologic presentation can often be misinterpreted as malignancy or serious infection. Treatment commonly involves combinations of various therapies including corticosteroids, surgery, radiotherapy, immuno-modulators, and antibiotics.

Case Presentation

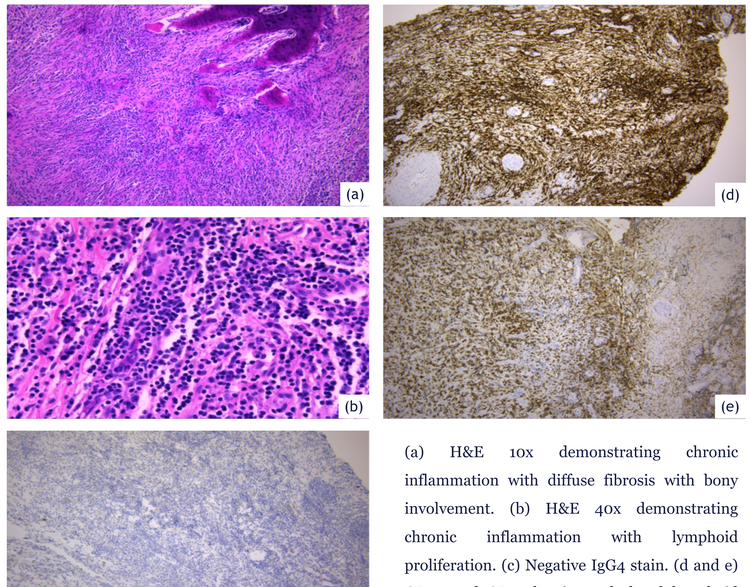
48 year old female with worsening visual acuity, retro-orbital pain, and increased intra-ocular pressure. MRI showed a lesion originating from the posterior ethmoid extending into the pterygopalatine fossa and inferior orbital fissure.



Initial Management

The patient underwent endoscopic sinus surgery with biopsy. Staining to rule out lymphoproliferative disorders and IgG4-related disease was performed. Pathology was consistent with lymphoid predominant inflammatory pseudotumor.

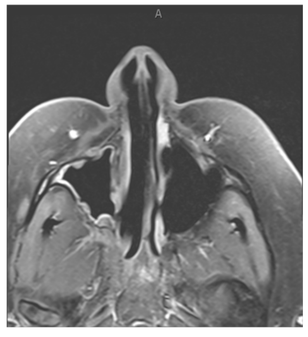
Histopathology



(a) H&E 10x demonstrating chronic inflammation with diffuse fibrosis with bony involvement. (b) H&E 40x demonstrating chronic inflammation with lymphoid proliferation. (c) Negative IgG4 stain. (d and e) CD20 and CD3 showing polyclonal lymphoid proliferation.

Initial Management

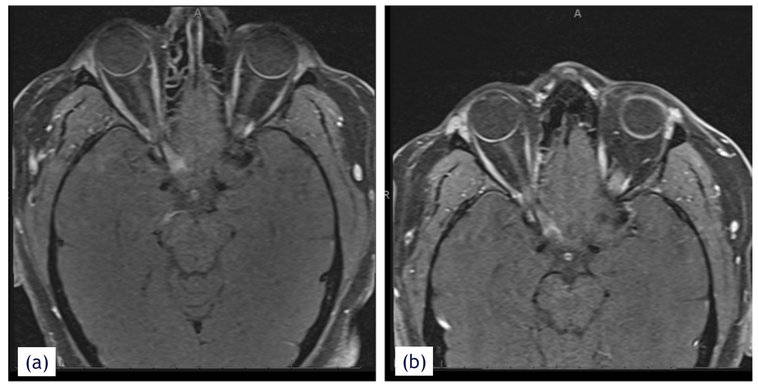
The patient was initiated on an extended course of high dose corticosteroids. Near complete resolution of disease – both radiographically and symptomatically – is appreciated at 1 month.



Follow-up

3-month post-treatment MRI showed a new lesion of the optic nerve and shortly thereafter, developed visual loss. The patient did not respond – either symptomatically or radiographically – to high dose corticosteroids.

Follow-up Imaging



(a) Routine 3-month post treatment MRI demonstrating a new lesion of the right optic nerve. (b) 6 weeks following the initiation of repeat corticosteroid treatment, repeat MRI demonstrates persistence of the right optic nerve lesion.

Additional Workup and Treatment

Referrals to rheumatology and neurosurgery were made. Further IgG and autoimmune workups were negative. Trials of methotrexate, azathioprine, and cyclosporine were unsuccessful. The decision was made to pursue definitive radiotherapy – 20Gy over 10 sessions.

Conclusions

The patient returned to baseline vision following completion of radiotherapy. 3-month post treatment MRI shows resolution of the optic nerve lesion. To our knowledge, this is the first case of successful treatment of optic nerve pseudotumor managed by radiotherapy alone.