Maxillary Sinus Infraorbital Neurofibroma: A Case Report and Review of the Literature

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Abstract

Introduction: Neurofibromas (NF) are uncommon benign tumors of peripheral nerves. Sinonasal NFs remain rare with poorly described outcomes. We sought to describe a case managed surgically and review the literature to date.
Methods: Institutional review board exemption was obtained for retrospective review, Pubmed search was conducted.
Results: A recent case of infraorbital nerve NF treated with endoscopic resection is described. Literature review shows that the maxillary sinus is the most common sinus affected, with radiation exposure being a risk factor.
Discussion: NFs are rare sequelae of radiation treatment and must be differentiated from tumor recurrence. Surgical excision, stereotactic radiosurgery, and observation are reasonable treatment options.

Introduction

Neurofibromas (NF) are rare benign peripheral nerve sheath tumors most commonly found in the skin and subcutaneous tissues. Given the limited number of reported maxillary neurofibroma cases, we present a review of the relevant literature and report a unique case to further expand the skull base annals.

Methods and Materials

Institutional review board exemption was obtained for retrospective review. Relevant literature review was performed through PubMed search. Search terms included “neurofibroma,” “sinonasal,” and “sinus.”

Results

Case Report: A 19-year-old male with a remote past medical history of embryonal rhabdomyosarcoma of the skull base centered around the petrous apex presented to clinic for evaluation. He was treated with chemotherapy followed by radiation in childhood. The NF was incidentally noted in the right maxillary sinus on oncologic magnetic resonance imaging (MRI) surveillance. The lesion was noted to increase in size on a subsequent MRI, which prompted a biopsy consistent with neurofibroma. MRI revealed a T2 hyperintense enhancing lesion along the right inferior orbital fissure, measuring approximately 3.2 x 1.7 x 1.4 cm (Figure 1). Options for treatment of the NF were discussed including observation, gamma knife radiosurgery, or endoscopic resection. Resection was elected, so we proceeded with an endoscopic modified Denker’s approach, revealing the mass in the lateral maxillary sinus wall and inferior orbital fissure (Figure 2). Frozen section pathology demonstrated neurofibroma. Postoperatively, the patient had facial numbness as expected. His postoperative course was otherwise uncomplicated without diplopia or epistaxis. There has been no evidence of recurrence on endoscopic surveillance or MRI at 6 months post-operatively.

Literature Review:
-approximately 25-44% of neurofibromas arise in the head and neck, with less than 4% found in the paranasal sinuses.
-The most common location for a sinonasal NF is the nasal cavity, with potential symptoms of diplopia, facial pain/pressure, facial numbness, epistaxis or nasal obstruction depending on the size and location of the mass.
-Though rare, radiation exposure is a known risk factor for the development of solitary neurofibroma in patients without a history of neurofibromatosis type 1 (NF1).
-fewer than 100 cases have been reported worldwide, most (90%) are sporadic and the minority (10%) found in the setting of neurofibromatosis
-surgical excision is typically curative with no reported recurrences after complete resection

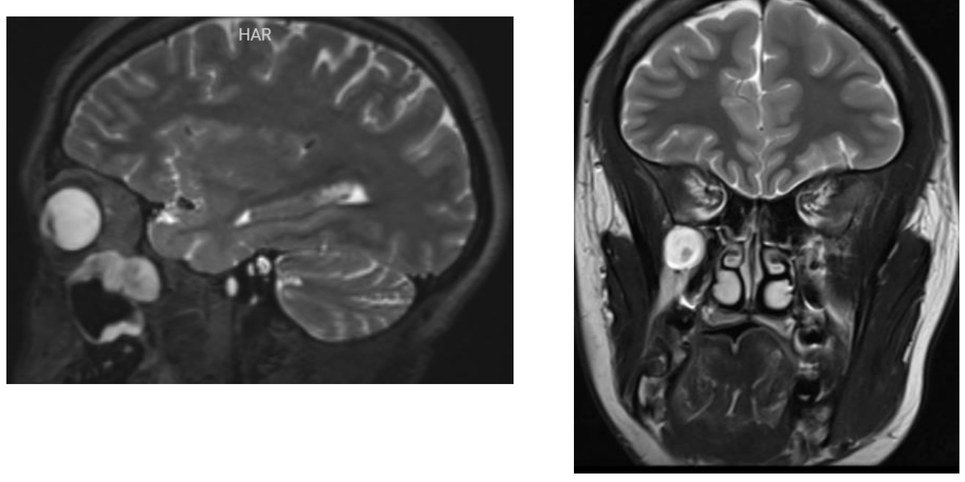

Figure 1. Imaging at the time of presentation. MRI exam demonstrating a T2 hyperintense, enhancing lesion along the right inferior orbital fissure.

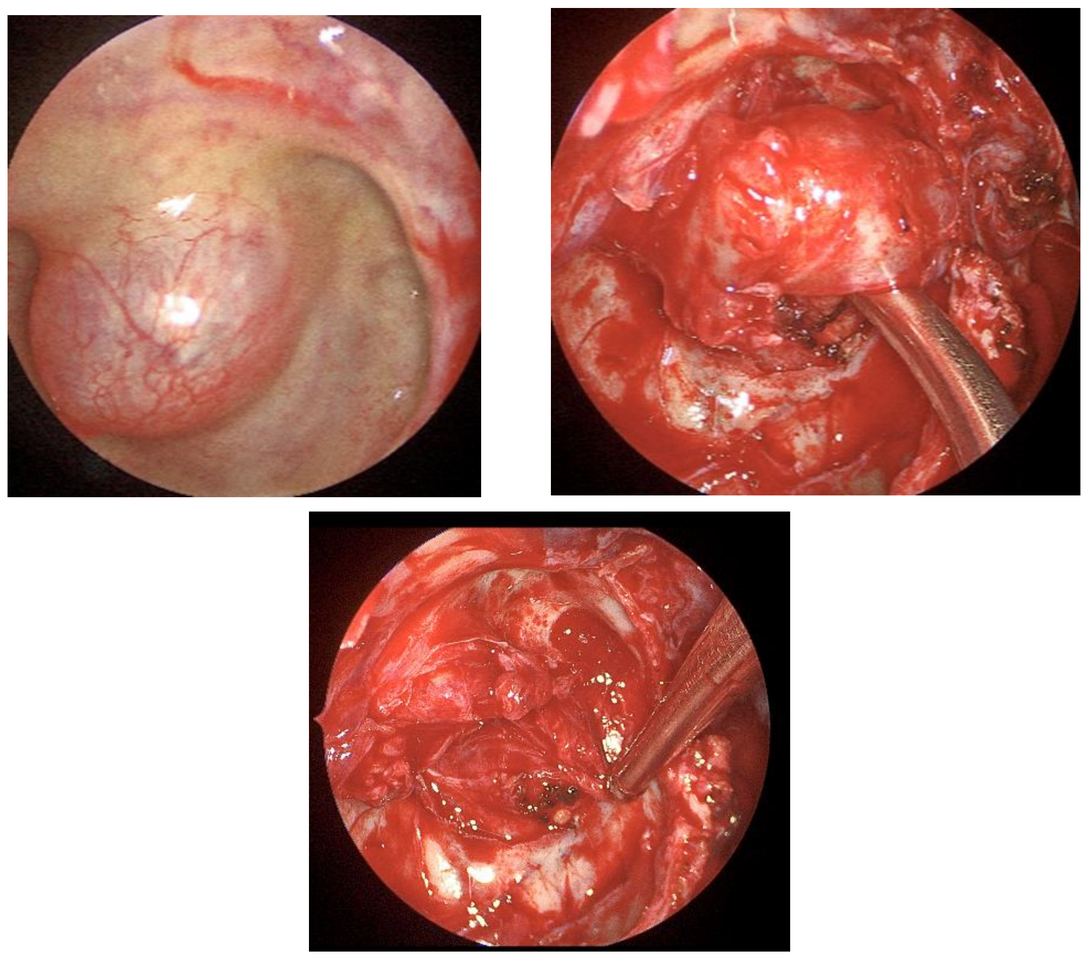

Figure 2. Endoscopic modified Denker’s approach, revealing adequate visualization of the lateral right maxillary wall and neurofibroma (C). Dissection was carefully performed to mobilize the mass away from the periorbita keeping the orbital fat and inferior rectus muscle intact. The infraorbital nerve was sacrificed to remove the mass

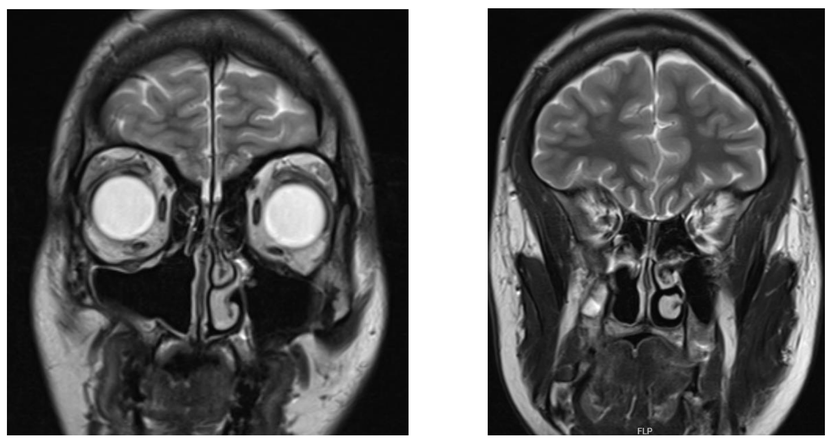

Figure 3. 6-month post-operative T2 weighted MRI showing no residual tumor

Discussion

-NF are a rare sequelae of skull base radiation and may require surgical excision.
-This lesion may be confused with tumor recurrence or development of a radiation-induced malignancy.
-Further research into risk factors for development of NF is needed. Further studies demonstrating the natural history of NFs will also help guide treatment strategies

Conclusions

-Sporadic NFs can arise as a long-term sequelae of radiation therapy
-Biopsy should be considered to rule out recurrence
-For confirmed NFs, surgical excision, stereotactic radiosurgery, and observation are reasonable treatment options

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