Nasal Plexiform Schwannoma with Intracranial Deformities Successfully treated with Skull Based Endonasal Resection: A Case Report

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Introduction

Plexiform schwannoma is a rare benign type of Schwann cell tumor typically be associated with NF-2 or schwannomatosis. Similar to other schwannoma types, they originate from any nerve with Schwann cells, including cranial nerves. Tumors may involve multiple nerve fascicles which can make resection challenging. These tumors are characterized by a plexiform pattern of interneural growth and commonly multinodular Most cases, 23%, are found in the head and neck though cases within the sinonasal tract are less common, with an incidence of less than 4.3%. Surgical resection is first-line treatment, although the nerve origin is indeterminate in 50% of cases. We present a case of a nasal plexiform schwannoma with cranial deformity treated with endoscopic endonasal resection.

Case Report

A 44-year-old man with no past medical history presented complaining of one year of congestion with right nasal deformity and an intranasal mass. The patient sought evaluation by otolaryngology. CT sinus showed a large, up to 5.8cm, isodense mass in the right nasal cavity extending towards the cribriform plate and right frontal sinus, with bony remodeling and dehiscence of the anterior cranial fossa. Subsequent MRI revealed an avidly enhancing, T2 isointense mass with mild diffusion restriction and intracranial extension, without clear intradural invasion. In office, endonasal biopsy was positive for neurofibroma.

Results

Given the size and symptomatic nature of the tumor, surgical resection was pursued via a multidisciplinary endoscopic endonasal approach. The inferior portion of the tumor was centrally debulked using the ultrasonic aspirator and dissected free from the surrounding nasal structures. Of note, the inferior portion of the tumor was tan-pink and firm, in nature, grossly more consistent with a neurofibroma. During dissection, the tumor was found to be densely adherent and inseparable to the septal mucosa, which was the presumed origin. As dissection was carried superiorly towards the skull base, the tumor became fatty and yellow in appearance, more consistent with a schwannoma. Tumor was resected from the eggshell, thin cribriform plate, superiorly, and a gross total resection was achieved. There was no intradural invasion of the tumor. Pathology was positive for plexiform schwannoma with ancient characteristics. Three specimens consistent with multiple nodules of spindle cell neoplasm lined with nasal mucosa. Verocray bodies seen throughout. Immunohistory chemistry revealed lesional cells positive for S-100, SOX 10 and Negative for CD 34 and AE1/E3 and with low KI67 proliferation.

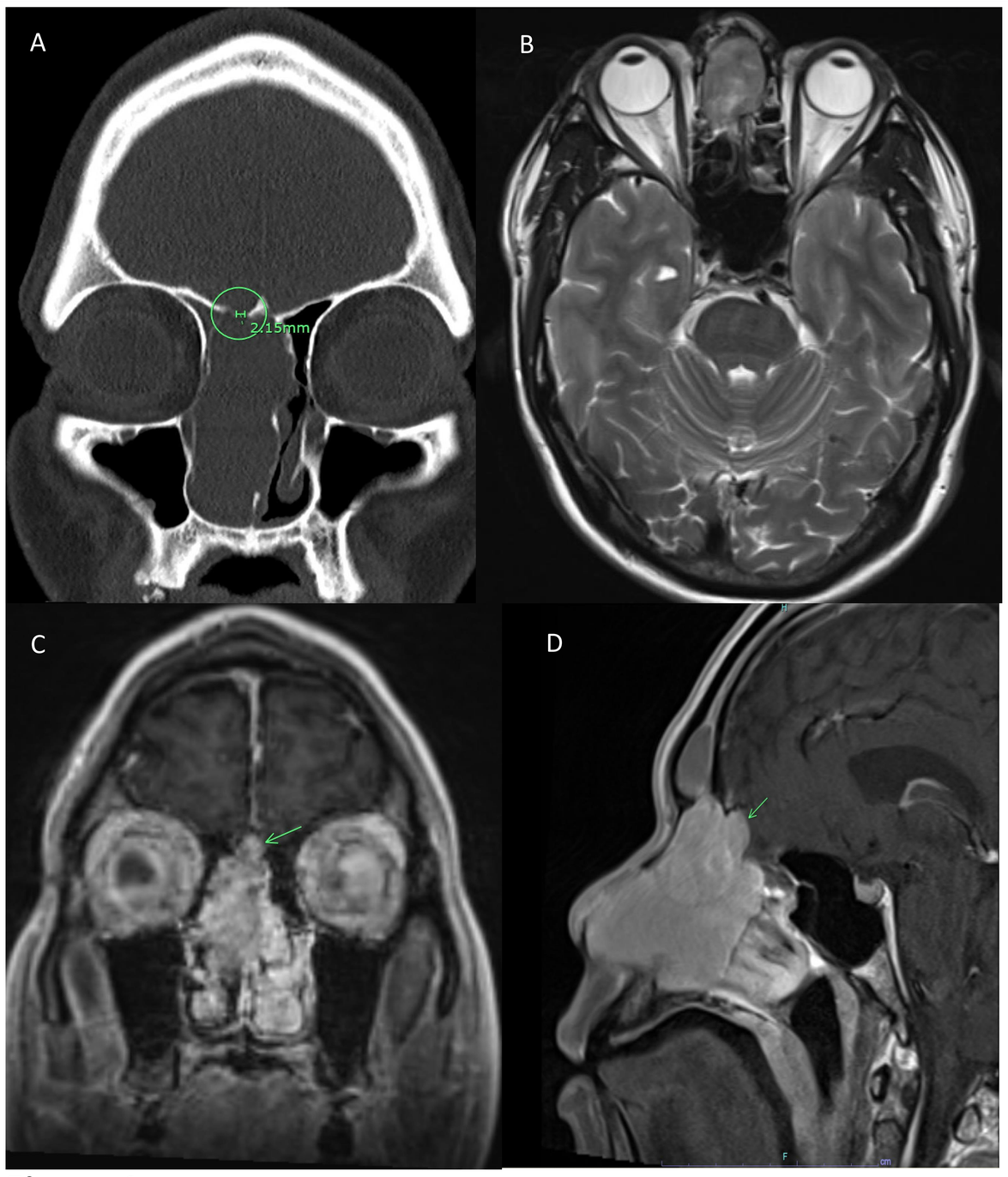

Figure 1. A. CT sinus isodense right nasal mass with multiple boney defects involving the vomer, right frontal bone, right nasal bone and right cribiform plate. B. MRI Pituitary T2 -weighted axial view showing an isointense mass with intracranial extension through the cribiform plate. C&D. T1 -weighted imaging post contrast coronal and sagittal views showing an avidly enhancing right nasal mass.

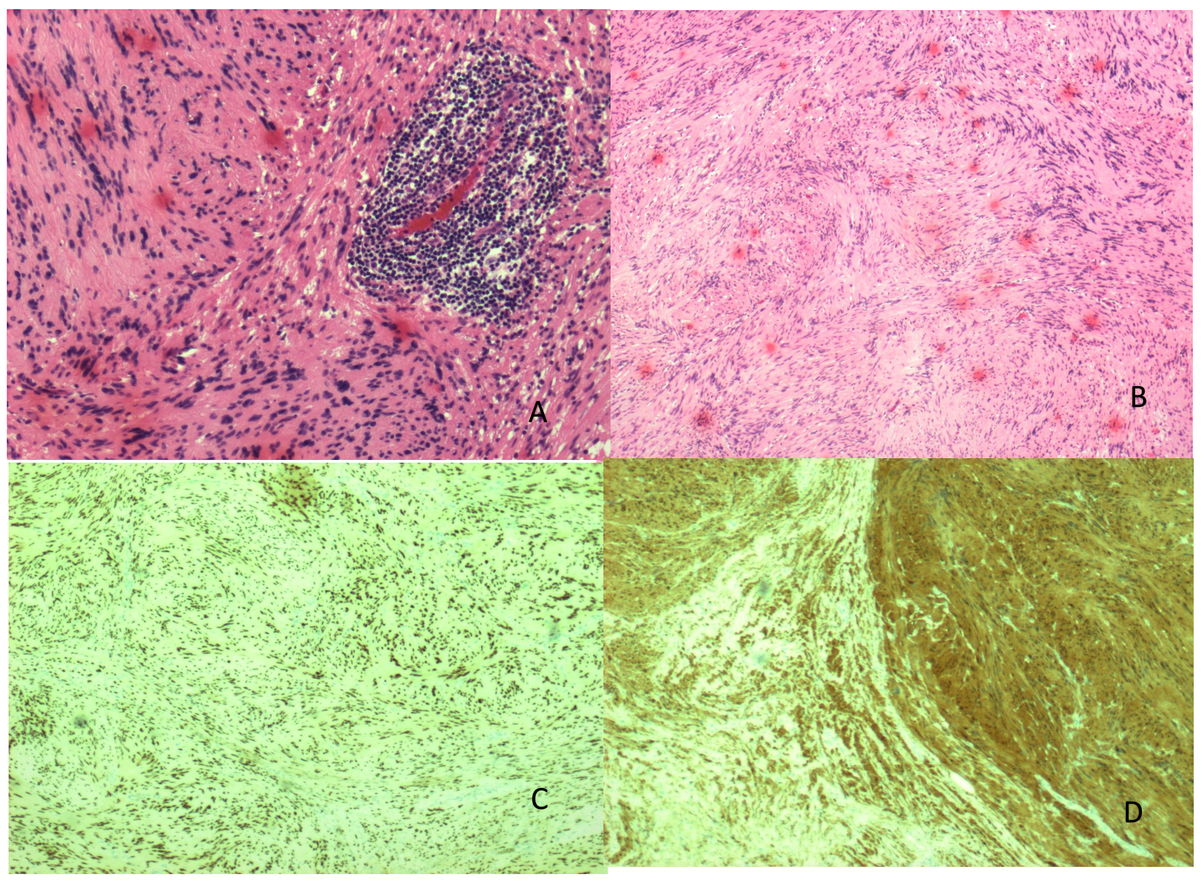

Figure 2. A and B. H &E staining with verocray bodies and multiple nodules of spindle neoplasms seen throughout. C. Sox-10 positive D. S-100 positive.

Conclusions

This case serves to demonstrate an uncommon presentation of a rare pathology, managed safely and effectively via a multidisciplinary endoscopic, endonasal approach. Gross total resection was achieved without complication and in a minimally invasive fashion.

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