

Advanced Pediatric Sinonasal Rosai-Dorfman Disease: A Hybrid Endoscopic Endonasal Approach



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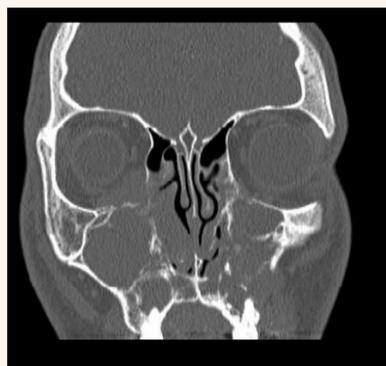
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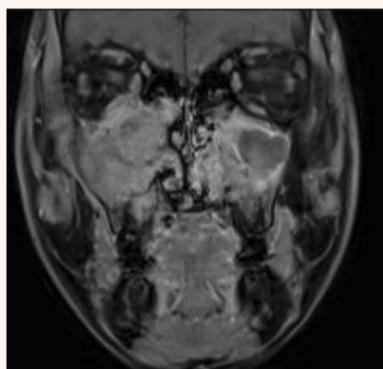
Abstract

Background

Rosai-Dorfman disease is a histiocytic disorder in the family of non-Langerhans cell histiocytosis. It was characterized by Rosai and Dorfman in 1969 after first being described by Destombes in 1965. The condition usually presents with bilateral, massive, painless cervical lymphadenopathy. Patients also have symptoms including fever, leukocytosis, elevated ESR, and polyclonal hypergammaglobulinemia. Extranodal presentations can include sinonasal cavities, GI tract, CNS, skin, breast, or soft tissue. The diagnosis ultimately relies on pathology. Though it is a benign disorder, it can mimic malignant pathologies and diagnosis can be challenging. Rosai-Dorfman disease usually develops in 1st or 2nd decade of life and it is estimated that extranodal disease occurs in 30-40% of patients, with 75% of these manifesting in the head and neck. Though it can spontaneously remit, the case presented required extensive surgical treatment.



Coronal CT without contrast showing extent of expansile soft tissue mass extending into right orbit, bilateral sinonasal cavities, and bilateral maxilla.



Coronal T1 weighted MRI showing soft tissue mass in the bilateral maxillary sinuses with expansion into the right orbit.

Case Description

Case Info

15y/o male with bilateral maxillary sinus masses who had biopsy proven Rosai-Dorfman disease presented in August 2020 for increased facial growth, nasal obstruction and snoring. He was started on Sirolimus in January 2021 and Trametinib was added to his treatment regimen in December 2021. There was subjective improvement in nasal obstructive symptoms; however, repeat imaging in June 2022 showed that the maxillary sinus masses were largely unchanged in size. Plans for repeat imaging and discussion of surgical debulking were started in March 2023. Shortly after this, patient was lost to follow-up until May 2024 when he returned with worsening nasal obstruction. He was found to have significant expansile growth into the right orbit without visual changes, growth into the bilateral maxilla, pterygopalatine fossa, and paranasal sinuses with extensive deformation. After tumor board discussion, he underwent multidisciplinary tumor resection and CMF reconstruction with ENT, neurosurgery, and oculoplastics.

Surgical Technique

- Intraoperative navigation done with Brainlab
- Tumor burden on the left was resected through and endoscopic endonasal approach
- Medial maxillary sinus mucosa was completely stripped to ensure complete removal of tumor burden

- Right sided tumor removal began through a maxillary vestibular incision - the tumor had partially eroded through the anterior maxillary wall
- Anterior maxillotomy was formalized and a SpiWay medium endonasal access guide was used to facilitate removal of the tumor



Eroded anterior maxillary wall through vestibular incision



Formalized anterior maxillotomy with arrow pointing to V2

- Oculoplastics then approached the tumor through a right orbitotomy approach
- The anterior maxillary wall was plated for complete CMF reconstruction
- Propel stents were placed into the bilateral nasal cavities with Nasopore spacers impregnated with Kenalog



Biopsy of the pterygopalatine fossa through the SpiWay endonasal access guide

Discussion

Conclusion

- Rosai-Dorfman disease is a histiocytic disorder in the family of non-Langerhans cell histiocytosis and steroids are the first line treatment
- In medically refractory cases with sinonasal manifestations, surgical intervention is sometimes required for symptom relief through removal of obstructing masses

Unique Case Points

- Combined endoscopic endonasal, open anterior maxillotomy, and orbitotomy approach provided access to ensure safe, maximal debulking in sinonasal Rosai-Dorfman disease refractory to medical therapy
- Propel stents and packing impregnated with Kenalog aid in high-dose topical delivery of steroids while avoiding need for systemic therapy of high-dose steroids - a particular concern for pediatric patients



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