Rosai-Dorfman Disease: A Case Report

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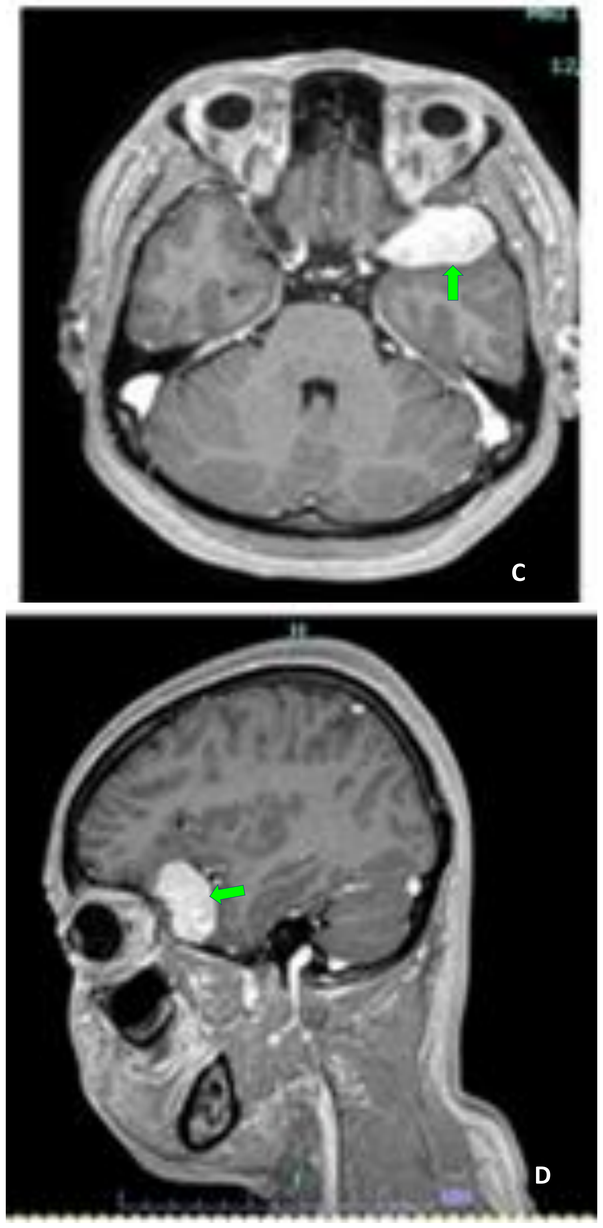
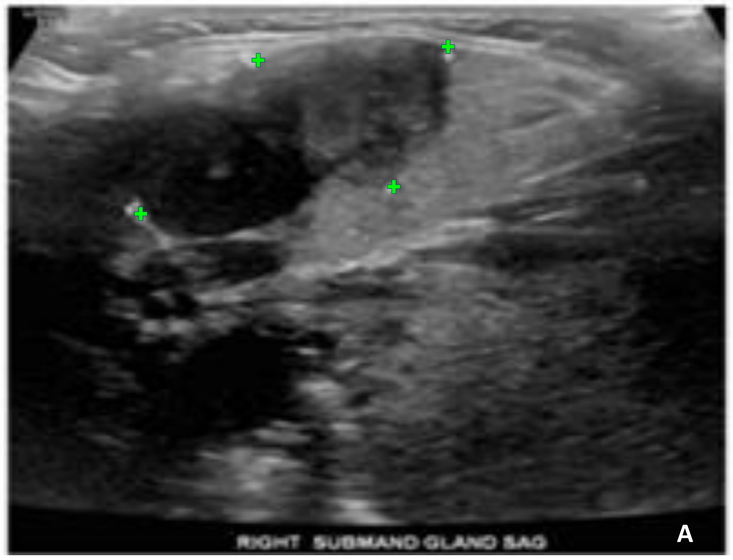
Background

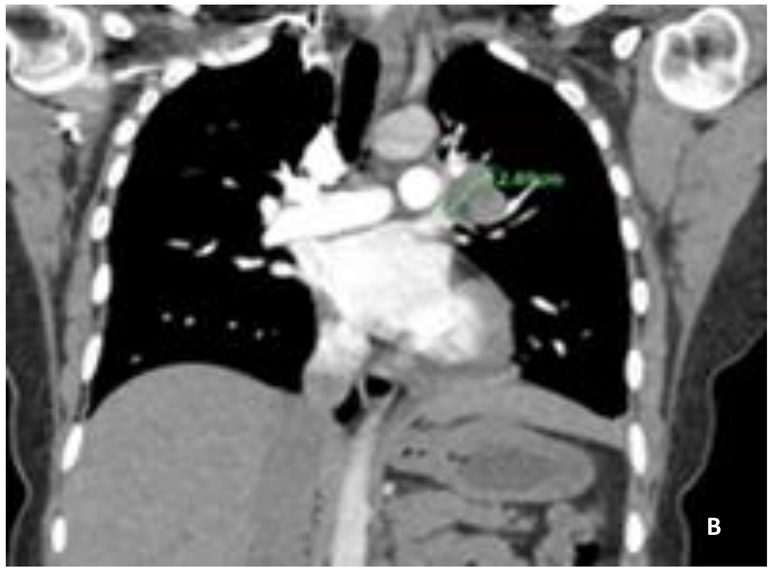
Rosai-Dorfman Disease (RDD) is a rare subtype of non-Langerhans cell histiocytosis characterized by the overproduction of activated histiocyte immune cells in affected tissues. The classic nodal type presents as cervical lymphadenopathy, with or without symptoms of night sweats, fevers, or chills. The extra nodal disease presentation, which is about 43% of RDD cases, can have multisystem involvement. Prognosis usually correlates with the number of systems involved. Diagnosing this disease can be difficult and requires high clinical suspicion as well as collaboration between multiple specialties as seen in our case review. As a primary care provider, at times being the first physician a patient encounters, one has the responsibility such as in this case, to coordinate the proper care required for the good of the patient.



Case

27 year old female presented with a tender submandibular mass worsening over a six month period. She denied fevers, chills, night sweats, or weight loss. She also denied any family history of lymphoma, leukemia, or sarcoidosis. On physical exam, a submandibular mass was palpated and found to be fixed, firm, and measuring approximately 3 cm x 2 cm.





Neck US (Image A) showed a hypoechoic right submandibular mass measuring 2.8 cm x 1.6 cm x 2.1 cm. FNA of the mass was negative for malignant cells, but showed an abundance of lymphoid cells, mixed inflammatory cells, and histiocytes. Flow cytometry was negative for monoclonal B cell population. Further imaging studies were significant for additional systemic lesions including an extra-axial dural based mass along the left sphenoid ridge (Images C and D), 2.6 cm hilar mass (Image B), and enhancing edema of the left parotid gland (not pictured).

Case Presentation

The patient subsequently underwent a complete excision of the submandibular mass for biopsy. Pathology revealed findings consistent with Rosai-Dorfman Disease. Immunohistochemistry revealed histiocytes that were positive for S100, CD68, cyclin D1 and OCT2 and negative for CD1a. These findings altered the treatment plan as patient initially was slated to undergo sphenoid mass removal by neurosurgery. However, following the discovery of the diagnosis, patient was initiated on IV steroids with an oral steroid taper by oncology. Thus far, patient has been responsive to the steroid regimen. Patient is planned to undergo repeat surveillance imaging following completion of treatment to assess overall response.

Conclusion/Discussion

Rosai-Dorfman Disease (RDD) is a condition that typically affects children and young adults with an average age of onset of 20 years with a slight predisposition for males over females. Common extranodal sites include orbital tissue, skin and central nervous system (CNS). Diagnosis of this condition requires imaging, clinical suspicion, immunohistochemistry, and multiple disciplines for treatment and management. After the provider's physical exam led to proper imaging, the case unraveled. Immediately when the location and pathology findings for the neck mass were discovered, the patient was set up with ENT specialists who then coordinated with Oncology and Neurosurgery. In our case above, extra nodal findings included intracranial involvement, submandibular, parotid glands and intrathoracic manifestations. CNS involvement can mimic a meningioma, which was seen in this case. Prior to the biopsy findings, the first step in treatment would have likely been excision and biopsy of the cranial mass. However, with the collaborative effort of ENT, oncology, and neurosurgery, steroids were thought to be the best appropriate step in treatment. Systemic treatment was sought out first, and thus far has seen response to IV steroids. If there was no response, chemotherapy especially with CNS involvement could have also been in the discussion. RDD in general is a benign disease. Prognosis tends to be good especially in nodal-involvement only cases where almost complete resolution can be seen. Overall, these cases will need close observation and surveillance imaging, especially given the unpredictable multisystem presentations.

References

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