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Rosai-Dorfman Disease with Nodal and Extranodal Involvements: A Case Report

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Abstract | **Case Report**

Rosai-Dorfman disease (RDD) is a rare histiocytic disorder characterized by the proliferation of non-Langerhans cells within the lymph nodes, and occasionally in extranodal sites. We report a case of RDD presenting as a left jugal swelling in a 43-year-old male. Imaging studies revealed a poorly defined lesion infiltrating the subcutaneous and muscular planes, as well as the maxillary bone. Biopsy confirmed the diagnosis of RDD. Treatment included surgical excision, followed by low-dose radiation therapy. The patient remained asymptomatic during a one-year follow-up.

Keywords: Rosai-Dorfman disease, MRI, CT.

INTRODUCTION

Rosai-Dorfman disease, or sinus histiocytosis with massive lymphadenopathy (SHML), is a rare but well-defined clinicopathologic entity first described in 1969 that affects predominantly children and adolescents. The most common presentation is bilateral painless cervical lymphadenopathy [1, 2]. Extranodal manifestations of the disease occur in up to 43% of patients; the most common sites affected are the soft tissues of the head and neck and the paranasal sinuses and nasal cavity [3–4]. Because of the rarity of the diagnosis, the extensive differential considerations, the nonspecific imaging findings, and the multiple sites of involvement, the diagnosis of SHML is often not considered, which leads to inappropriate treatment of this benign but often progressive disease [5].

CLINICAL CASE

A 43-year-old male presented with a left jugal swelling that had been progressively increasing in size over the course of a year. On examination, a hard and mobile mass was noted, with no clinically palpable cervical adenopathy. The CT scan (figure 1) showed a left jugal mass with poor delineation that measured approximately 31x27mm. The mass was isodense and heterogeneously enhanced on PDC. It infiltrated the maxillo-retro-zygomatic space and the masticatory space, coming into contact with the temporal and lateral pterygoid muscles without a separating line. Additionally, it contacted the maxillary bone with bone lysis and the masseter muscle externally, with loss of the separating line in places.

On MRI, the poorly defined left jugal lesion displayed T1 and T2 iso signal, with diffusion hypersignal and intense enhancement and ADC restriction after injection of Gadolinium. The lesion infiltrates the homolateral masseter muscle, medially comes into intimate contact with the left alveolar arches with bone lysis opposite, and mads contacts with the ascending ramus of the mandible without any anomaly of the bone signal in front of it. It also infiltrates the subcutaneous fatty plane.

Imaging studies, including CT and MRI, revealed a poorly defined lesion in the left jugal region. The lesion infiltrated the subcutaneous and muscular planes and also involved the maxillary bone. Biopsy of the lesion confirmed the diagnosis of RDD.

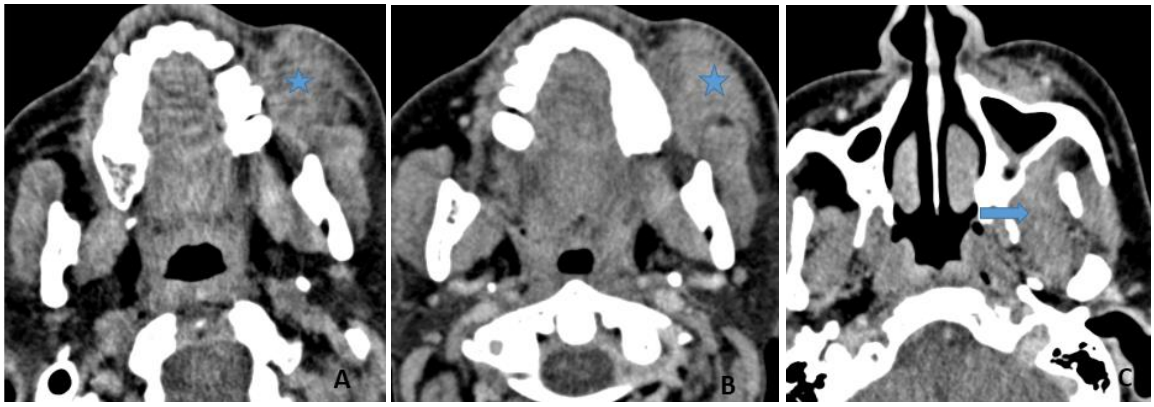

Figure 1: Axial acquisition of a facial scan without PDC injection (A) and with PDC injection at venous time: left jugal mass (asterix) with poor delineation. the mass was isodense (A) and heterogeneously enhanced on PDC (B and C). It infiltrated the maxillo-retro-zygomatic space (arrow) and the masticatory space, coming into contact with the temporal and lateral pterygoid muscles without a separating line

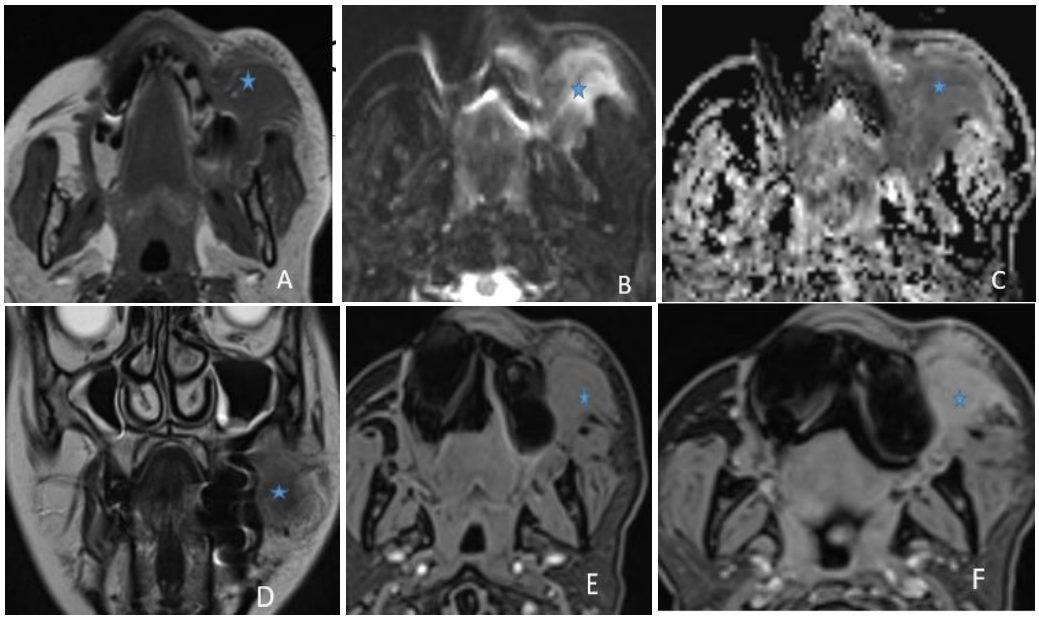

Figure 2: Axial T1-weighted (A) with diffusion-weighted sequence (B) and apparent diffusion coefficient (ADC) map (C), coronal T2-weighted (D), and Dixon water images before and after contrast enhancement. Poorly defined left jugal lesion (asterix) displayed T1 and T2 iso signal, with diffusion hypersignal and intense enhancement and ADC restriction after injection of Gadolinium. The lesion infiltrates the homolateral masseter muscle the subcutaneous fatty plane and also mads contact with the ascending ramus.

DISCUSSION

RDD is a rare disorder of histiocytic proliferation that can present with a wide range of clinical features, depending on the organs involved [6]. Although it usually presents with lymphadenopathy, extranodal involvement can occur, as seen in our case. RDD commonly affects children and young adults, with a slight male predominance [7]. The etiology of RDD remains unknown, but it is thought to be related to an immune dysregulation or a viral infection [8].

Imaging studies are essential in the diagnosis and management of RDD. CT and MRI scans can provide information about the extent of the lesion and its relationship to adjacent structures.

In our case, CT findings are also consistent with those reported in the literature for Rosai-Dorfman disease. A study by Chatal *et al.,* (2011) reported CT findings of Rosai-Dorfman disease, including a hypodense or iso-dense lesion with moderate or intense enhancement following contrast injection. Infiltration of surrounding tissues, including bone and muscle, was commonly observed [9, 10].

The MRI findings are consistent with those reported in the literature for Rosai-Dorfman disease. A study by McHugh *et al.,* (2001) reported MRI characteristics of Rosai-Dorfman disease, including iso or hypo-intense T1-weighted signal and variable T2-weighted signal, with heterogeneous or homogeneous enhancement following gadolinium injection [11].

Additionally, infiltration of surrounding tissues was commonly seen [12].

In our case, the lesion infiltrated the subcutaneous and muscular planes, and involved the maxillary bone, which required surgical intervention. Biopsy of the lesion confirmed the diagnosis of RDD, which was supported by the characteristic features of lymphoplasmacytic infiltrate and the presence of S-100 protein positive histiocytes.

Treatment options for RDD include surgical excision, corticosteroids, radiation therapy, and chemotherapy. Surgical excision is usually the first-line treatment for localized disease, as in our case. Radiation therapy can be considered in cases of incomplete resection or recurrent disease. Chemotherapy is reserved for cases of systemic disease or those that are refractory to other treatments.

CONCLUSION

RDD is a rare histiocytic disorder that can present with a wide range of clinical features, depending on the organs involved. Imaging studies, such as CT and MRI scans, are essential for diagnosis and management. Treatment options include surgical excision, radiation therapy, and chemotherapy. In our case, surgical excision followed by low-dose radiation therapy resulted in a favorable outcome with no evidence of disease during the one-year follow-up.

REFERENCES

1. Rosai, J. (1969). Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity. *Arch pathol*, *87*, 63-70.
2. Abla, O., Jacobsen, E., Picarsic, J., Krenova, Z., Jaffe, R., Emile, J. F., ... & Diamond, E. L. (2018). Consensus recommendations for the diagnosis and clinical management of Rosai-Dorfman-Destombes disease. *Blood, The Journal of the American Society of Hematology*, *131*(26), 2877-2890.
3. La Barge, D. V. (2008). rd, Salzman KL, Harnsberger HR, Ginsberg LE, Hamilton BE, Wiggins RH 3 rd, *et al.*, Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): Imaging manifestations in the head and neck. *AJR Am J Roentgenol*, *191*, W299-306.
4. Mar, W. A., Yu, J. H., Knuttinen, M. G., Horowitz, J. M., David, O., Wilbur, A., & Menias, C. O. (2017). Rosai-Dorfman disease: manifestations outside of the head and neck. *American Journal of Roentgenology*, *208*(4), 721-732.
5. Mantilla, J. G., Goldberg-Stein, S., & Wang, Y. (2016). Extranodal Rosai-Dorfman disease: clinicopathologic series of 10 patients with radiologic correlation and review of the literature. *American journal of clinical pathology*, *145*(2), 211-221.
6. Sodhi, K. S., Suri, S., Nijhawan, R., Kang, M., & Gautam, V. (2005). Rosai-Dorfman disease: unusual cause of diffuse and massive retroperitoneal lymphadenopathy. *The British journal of radiology*, *78*(933), 845-847.
7. McAlister, W. H., Herman, T., & Dehner, L. P. (1990). Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease). *Pediatric radiology*, *20*, 425-432.
8. Albano, D., Bosio, G., & Bertagna, F. (2015). 18F-FDG PET/CT follow-up of Rosai-Dorfman disease. *Clinical Nuclear Medicine*, *40*(8), e420-e422.
9. D'souza, M. M., Jaimini, A., Bansal, A., Tripathi, M., Sharma, R., Mondal, A., & Tripathi, R. P. (2013). Fdg-pet/ct in lymphoma. *The Indian journal of radiology & imaging*, *23*(4), 354.
10. Kwon, S. Y., Kim, T. K., Kim, Y. S., Lee, K. Y., Lee, N. J., & Seol, H. Y. (2004). CT findings in Kikuchi disease: analysis of 96 cases. *American Journal of Neuroradiology*, *25*(6), 1099-1102.
11. Bonekamp, D., Horton, K. M., Hruban, R. H., & Fishman, E. K. (2011). Castleman disease: the great mimic. *Radiographics*, *31*(6), 1793-1807.
12. Huang, B. Y., Zong, M., Zong, W. J., Sun, Y. H., Zhang, H., & Zhang, H. B. (2016). Intracranial Rosai–Dorfman disease. *Journal of Clinical Neuroscience*, *32*, 133-136.