Rosai-Dorfman Disease of the Head and Neck – Diagnosis and Management in an Elderly Patient

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Abstract

Rosai-Dorfman disease (RDD) is a rare disorder of histiocyte proliferation of unknown etiology. It typically affects children and young adults, and usually presents as painless bilateral lymphadenopathy of the head and neck. The course of the disease can range from spontaneous remission to progressive disease requiring treatment with surgery, steroids, chemotherapy and other therapeutic modalities.

Introduction

- RDD typically presents with massive cervical lymphadenopathy in younger patients[1]
- Due to the low incidence of the disease, there is scant literature describing optimal management
- We review radiographic and pathologic findings as well as response to a treatment course of low-dose steroids

Case Report

- 69 year-old woman with progressively enlarging bilateral cervical lymphadenopathy
- Occasional flare-ups with cervical tenderness. Otherwise no pain or constitutional symptoms, including fatigue, fevers, night sweats, or weight loss
- Onset of symptoms nine months prior to presentation in the setting of upper respiratory infection
- Initial FNA ordered showed indeterminate pathology
- Treated with multiple rounds of oral antibiotics without improvement
- Surgical pathology from excisional biopsy consistent with RDD
- Initially managed conservatively with observation; serial CT scans showed increasing size and number of enlarged nodes
- Neck CT scan with contrast (Figure 1) showed bilateral lymphadenopathy, most pronounced in levels Ia and Ib of the neck, with nodes measuring up to 3.3cm, as well as multiple enlarged bilateral intraparotid lymph nodes.
- Started on low-dose Prednisone 20mg daily with a slow taper over several months.
- She responded well with decreased size and number of cervical and intraparotid lymph nodes.

Pathology

- Partially preserved lymph node architecture with follicles and marked sinusoidal proliferation of large histiocytes with foamy cytoplasm, round vesicular nuclei, and prominent nucleoli (Figure 2)
- Large histiocytes containing intact lymphocytes (emperipolesis) (Figure 3)
- CD20, CD79a, BCL-6, and CD10 positivity, along with negative staining for BCL-2 confirmed the reactive nature of the observed follicles
- Histiocytes strongly S100 positive
- CD1a, AE1/AE3, and special microorganism stains (AFB, PAS, Warthin-Starry) negative

Figure 1

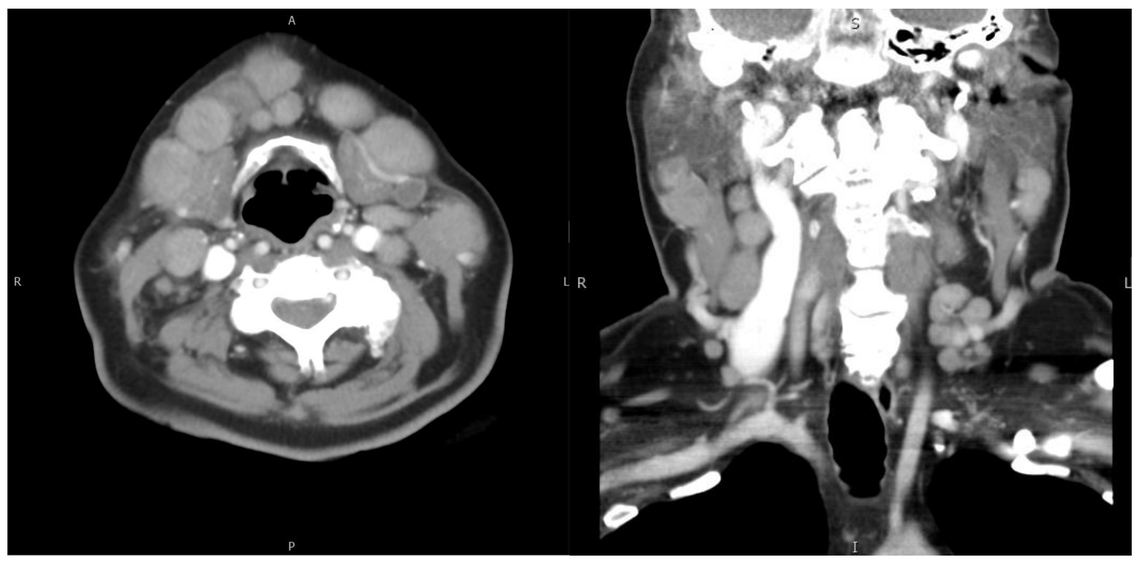


Figure 2

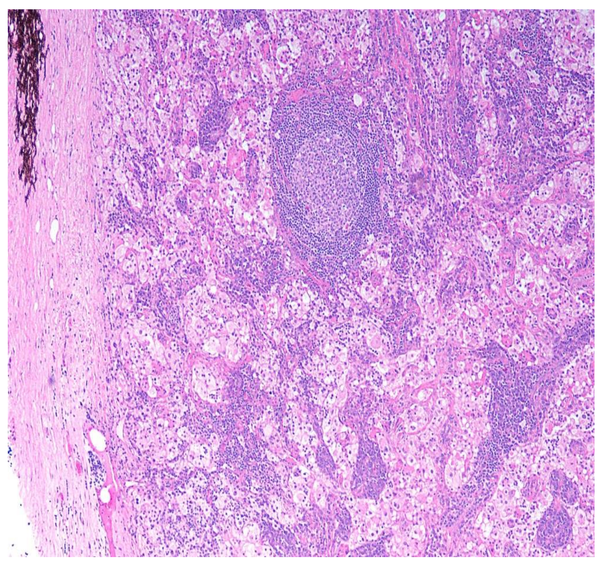
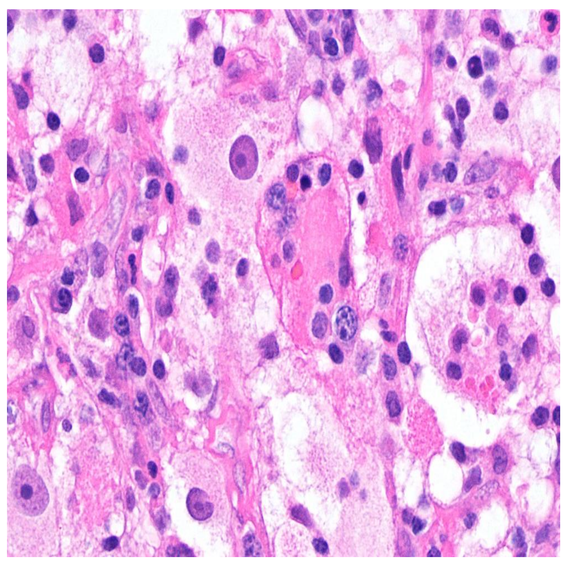


Figure 3



Discussion

RDD is a rare disease that most commonly affects younger patients. There is limited literature describing its natural history and treatment in older adults. The mean age of onset is 20.6 years, although cases have been documented in individuals up to 74 years of age.[1] The disease usually presents as massive cervical lymphadenopathy, but extranodal involvement as in our patient has been described in 30-43% of cases, with 75% of these involving the head and neck.[1,2,3] Within the head and neck, RDD typically affects the upper aero-digestive tract, especially the nasal cavity and paranasal sinuses.[3,4] Upper airway involvement resulting in obstruction and respiratory distress is also possible.[2]

Pathologic examination is important for the diagnosis of RDD, as this disease in the head and neck can be non-specific. Histopathologically, the diagnosis of RDD may be confused with Langerhans cell histiocytosis, melanoma, granulomatosis with polyangiitis, Hodgkin's disease, rhinoscleroma, and other fibroinflammatory lesions.[4] The hallmark of RDD is proliferation of large histiocytes containing intact lymphocytes (emperipolesis) and expanding the lymphoid sinuses. Histiocytes typically stain strongly positive with S-100 and CD-68, as was the case in our patient.[1,4,5]

The natural history and severity of RDD is highly variable and dependent on the affected sites. The disease is usually self-limited, with most patients undergoing complete remission after a relapsing-remitting course.[1] In a review focused on the treatment of RDD, Pulsoni et al. recommended conservative management with observation if lesions are not massive and there is no vital organ involvement. Steroids were recommended in case of fever and/or sudden lymph node enlargement.[6] In certain cases where the disease affects vital structures (e.g. subglottic lesion causing airway obstruction), surgical debridement may be necessary.[2]

Conclusions

In the diagnosis of RDD, tissue diagnosis is critical to rule out any lymphoproliferative process, lymphoma, or malignancy. Imaging is useful to evaluate burden of disease and the affected anatomic areas of the body. Children and young adults are most commonly affected but this disease may also affect elderly adults. Treatment of Rosai-Dorfman Disease is variable and dependent on patient symptoms and the anatomic areas affected.

References

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