Rosai-Dorfman Disease in the Parapharyngeal Space: The First Reported Case

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Introduction

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, represents a rare and benign proliferation of histiocytes. Although first described in 1969, the pathogenesis remains unknown. Rosai-Dorfman disease primarily affects male children, adolescents, and young adults. The disease usually manifests as painless bilateral cervical lymphadenopathy; however, extranodal involvement has been reported.

We present a case of a 53-year-old woman with RDD of the parapharyngeal space and a brief review of the literature. To our knowledge, this is the first reported case of RDD involving the parapharyngeal space.

Case Report

A 53-year-old African American woman with no significant past medical history presented to the otolaryngology clinic complaining of chronic and intermittent right-sided otalgia, neck pain, neck swelling, and hearing loss. Examination revealed a right-sided serous otitis media but was unremarkable for cervical lymphadenopathy. The patient underwent contrasted magnetic resonance imaging of her neck to further evaluate her neck complaints. Imaging revealed a 2.8 cm × 1.3 cm solid enhancing mass deep to the right parotid gland, partially effacing the parapharyngeal space (Figures 1 and 2). The mass appeared concerning for primary parotid malignancy due to its atypical characteristics. An enlarged right level II lymph node was also identified. Surgical excision was planned.

Excisional biopsy of the level II lymph node was performed. The parapharyngeal mass was adherent to the medial pterygoid muscle. Frozen histopathology favored pleomorphic adenoma. Given the difficult exposure and a provisional benign diagnosis, a gross total resection was taken without oncologic margins. The procedure was completed without apparent complications.

Permanent histopathology of both the lymph node and parapharyngeal mass revealed a diagnosis of RDD. The patient recovered without any complications and is disease-free 6 months postoperatively.

Figure 1. Coronal T1-weighted magnetic resonance imaging showing a 2.6-cm hyperintense right-sided parapharyngeal space mass.

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Received: April 5, 2020; revised: April 27, 2020; accepted: April 28, 2020

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Discussion

The National Institutes of Health characterizes RDD as an abnormal benign proliferation of histiocytes, a phagocytic white blood cell that manifests primarily in the cervical lymph nodes.\(^3\)

Epidemiology

Rosai-Dorfman disease primarily affects males who are in their second to third decade of life, with African Americans being most commonly affected.\(^4\) The incidence and prevalence of RDD are difficult to calculate due to its rarity.

Pathogenesis

No consensus exists regarding the pathogenesis of RDD; however, limited evidence suggests that viral infection with either human herpesvirus-6\(^5,6\) or parvovirus B-19\(^7\) may play a role. Others propose there exists a genetic predisposition to RDD, since up to 33\% of cases have mutations in the KRAS and MAP2k1 genes.\(^8\)

Presentation

The hallmark of RDD is painless cervical lymphadenopathy, which is present in up to 90\% of cases. Forty percent of cases may have extranodal involvement, such as those involving the central nervous system (CNS) and aerodigestive tract.\(^9\) Patients affected with RDD typically present with signs and symptoms secondary to mass effect of the areas involved (eg, neurological deficits with CNS involvement). Up to 30\% of cases involve fever. Some patients may present with complaints related to autoimmune disorders (eg, arthralgias or myalgias).

Rarely, RDD affects only the skin, manifesting as multifocal discolored papules. These patients tend to present at an older age (5th decade) than classically described patients with RDD.\(^2\)

Diagnosis

If RDD is suspected preoperatively, laboratory findings of an elevated erythrocyte sedimentation rate (present in 90\% of cases),\(^8\) anemia, and thrombocytopenia may help narrow the diagnosis. And while case reports suggest that a diagnosis of RDD can be made by fine needle aspiration\(^9\); ultimately, however, diagnosis of RDD requires pathological confirmation.

Classic pathology findings of RDD include dilated sinuses with pericapsular fibrosis,\(^1\) with or without foamy eosinophilic histiocytes. The hallmark microscopic finding of RDD is emperipolesis, a phenomenon describing intact cells being present within the cytoplasm of another. In RDD, this occurs typically as intact lymphocytes within a sinus histiocyte.\(^1\) Immunohistochemical staining of RDD specimens display strong S100 and CD68 positivity.\(^10\)

Treatment

Rosai-Dorfman disease is observed in most patients. Surgical excision is reserved for symptomatic lesions causing mass effect on surrounding structures, though recurrence may occur.\(^9,11\) If patients are experiencing systemic symptoms such as fevers, arthralgias, and unremitting lymphadenopathy, short courses of oral steroids have been used to mitigate symptoms.\(^12\) In cases of disseminated RDD, limited data suggest chemotherapeutic agents such as methotrexate, 6-mercaptopurine, and imatinib may be useful.\(^4\)

Conclusion

We present the first reported case of RDD involving the parapharyngeal space. Rosai-Dorfman disease represents a unique clinical entity that presents a challenge for otolaryngologists and other clinicians who may encounter such patients. Increased familiarity of RDD and its presentation, diagnosis, self-limiting nature, and possible management strategies should help to improve treatment algorithms, prevent unnecessary surgery, and improve patient outcomes.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

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