

Case Report

A Rare Case of Isolated Intracranial Rosai–Dorfman Disease Mimicking Optic Nerve Meningioma: A Case Report and Literature Review

Abstract

Sinus histiocytosis with massive lymphadenopathy, or Rosai–Dorfman disease (RDD), was first described in 1969 as a reactive condition of unknown etiology that is characterized by a proliferation of histiocytes exhibiting emperipolesis of both lymphocytes and plasma cells. It usually presents with painless cervical lymphadenopathy either with or without extranodal manifestations. Intracranial involvement of this disease is extremely rare. Intracranial RDD occurs in <5% of all patients with extranodal disease. Here, we report a case of RDD with isolated intracranial involvement. A 67-year-old male presented with a long-standing headache, retro-ocular pain, and progressive visual loss of the left eye. Magnetic resonance imaging showed features of optic nerve meningioma. The histopathology revealed sheets of histiocytes displaying emperipolesis. These histiocytes were S100 positive; however, a CD1a and epithelial membrane antigen were negative.

Keywords: Intracranial tumor, optic nerve meningioma, Rosai–Dorfman disease, sinus histiocytosis

Introduction

Rosai–Dorfman disease (RDD) was first described in 1969 as an idiopathic histiocytic proliferative disorder.^[1] It is characterized by bilateral cervical lymphadenopathy, fever, leukocytosis, increased erythrocyte sedimentation rate, and hypergammaglobulinemia. Isolated extranodal disease occurs in approximately 23% of cases and affects the skin (12%), paranasal sinuses (11%), soft tissue (9%), bone (9%), salivary glands (5%), central nervous system (CNS) (5%), oral cavity (3%), kidneys (2%), lower respiratory tract (2%), larynx (1%), and orbit (5%–10%).^[2,3,5] To the best of our knowledge, fewer than 100 cases of RDD that were restricted to the CNS without associated adenopathy have been reported in literature.^[6,7] Due to its rarity, RDD is never the first diagnosis in intracranial lesions. It mimics meningioma, neurosarcoidosis, and metastasis radiologically. Surgery has been applied as the first therapy, and it is essential for diagnosis as well. After total removal, although the outcome is usually good, postoperative corticosteroids have been recommended, and some cases may require additional chemotherapy and/or radiation.

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Case Report

A 67-year-old male presented with a headache, retro-ocular pain, and transient blurred vision of 3 months' duration. He had no cervical lymphadenopathy and no fever in his recent history. Neurological examination revealed a visual acuity defect and proptosis of the left eye. His neurological examination was otherwise unremarkable. Laboratory tests revealed the following results: white blood cells, 4320 cells/mL; hemoglobin, 12.2 g/dl; and platelet count, 300,700 platelets/ml. His C-reactive protein was negative. Magnetic resonance imaging (MRI) showed an isointense T1 and hypointense T2 extra-axial space-occupying lesion in the posteromedial retro-ocular region of the left eye, which measured 2.3 cm × 2.6 cm, with homogeneous contrast enhancement and enhancing dural tail [Figure 1]. A systemic computed tomography (CT) scan of the chest, abdomen, and pelvis was negative. Presurgery, the primary diagnosis was meningioma. The mass was totally resected using a fronto-orbito-temporal approach, and during debulking, the lesion was found to be firm and partially well defined. A routine postoperative CT scan of the skull showed gross total mass resection [Figure 2].

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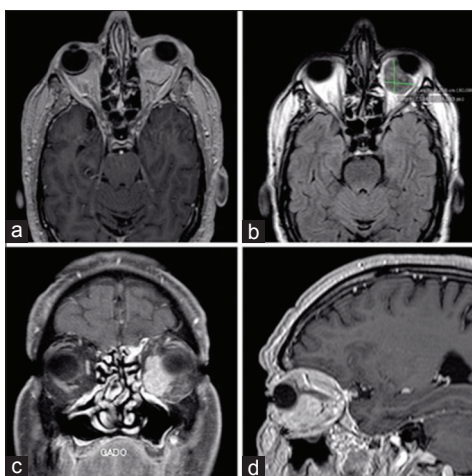


Figure 1: Preoperative magnetic resonance imaging of the head of a 67-year-old male with Rosai–Dorfman disease. (a) Axial T1-weighted gadolinium-enhanced magnetic resonance imaging showing an isointense well-defined lesion in the left medial retro-ocular space. (b) The lesion is hypointense on axial fluid-attenuated inversion recovery imaging. (c) Coronal T1-weighted gadolinium-enhanced magnetic resonance imaging demonstrating enhancement of the lesion. (d) Sagittal T1-weighted gadolinium-enhanced magnetic resonance imaging demonstrating enhancement of the lesion

The histiocytes of a frozen section of the lesion were immunopositive for S-100 protein and CD68 and negative for CD1. These findings were consistent with extranodal RDD.

The patient was discharged 7 days postsurgery. A follow-up MRI examination 3 months later revealed no recurrence.

Discussion

RDD is a condition of unknown origin that predominantly affects children and young adults and shows a predilection for males. The disease can appear with massive painless cervical lymphadenopathy, fever, anemia, leukocytosis, an elevated erythrocyte sedimentation rate, and polyclonal hypergammaglobulinemia. Extranodal involvement, including the skin, respiratory tract, visceral organs, genitourinary tract, bones, CNS, and orbit, occurs in 43% of cases.^[8]

Intracranial involvement of RDD is extremely rare. The most common imaging appearance of intracranial RDD is a dural-based enhancing mass that mimics a meningioma. The MRI characteristics of this intracranial dural-based space-occupying lesion are quite similar to those of meningioma.

A differential diagnosis of the imaging findings includes meningioma, eosinophilic granuloma, lymphoproliferative disorder, plasma cell granuloma, and dural-based metastasis.

Histopathologically, RDD is characterized by a variety of chronic inflammatory cells that are dominated by lymphocytes and plasma cells. There are also scattered giant foamy macrophages and histiocytes that engulf lymphocytes,



Figure 2: Postoperative computed tomography image of the head of a 67-year-old male with Rosai–Dorfman disease. (a) Axial plain computed tomography scan with contrast showing a total resection of retro-ocular mass of the left eye. (b) Noncontrast axial plain computed tomography scan. (c) Sagittal plain computed tomography scan. (d) Coronal plain computed tomography scan

plasma cells, and polymorphonuclear leukocytes; notably, emperipolesis may occur.^[9] Immunohistochemical studies in RDD have revealed positivity for the S100 protein and CD68 (KP1), yet negativity for CD1a.^[10,11] The presence of chronic inflammatory infiltrate, emperipolesis, and positive S100 staining is consistent with RDD [Figure 3]. It can be differentiated ultrastructurally from Langerhans cell histiocytosis by the absence of Birbeck granules^[12] and negativity for CD1a.

Isolated intracranial RDD is a benign disease with a good prognosis. Recurrence and development of neurological deficits are rare. Since surgical resection is the best modality of therapy, follow-up MRI is recommended to evaluate the long-term outcome. The lesion is extremely radiosensitive. Progressive disease and nonresectable lesions are treated with radiotherapy and chemotherapy. Postsurgical complications, such as bleeding, may occur when the lesions are highly vascular, as shown radiologically by perilesional edema. In a review of the follow-up data of 43 patients with RDD, 58% were alive with the disease. Only two patients had died, and neither death was reported as a result of isolated intracranial RDD.^[13]

Conclusion

Although isolated intracranial RDD is rare, it should be included in the differential diagnosis of a dural mass that resembles meningioma. The first approach is usually a surgical one because preoperative differentiation from meningiomas on imaging criteria alone is extremely difficult, and debulking of a space-occupying tumor is indicated. Special stains and immunohistochemistry are needed for a definitive diagnosis because both the prognosis and treatment aspects differ for the various differential diagnoses.

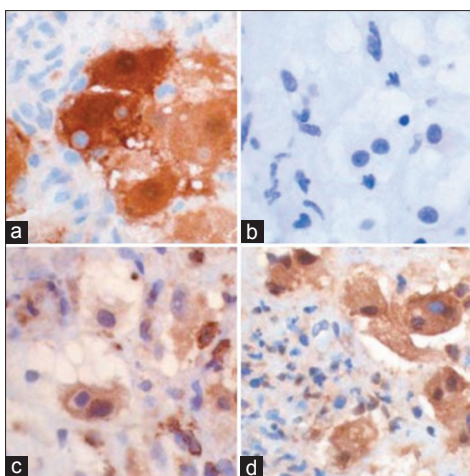


Figure 3: Immunohistochemistry of histiocytes in our case consistent with RDD (400x). (a) Stain for CD-1a was negative. (b) Stain for lysozyme was positive. (c) Stain for alpha-1-antitrypsin was positive. (d) Stain for S-100 was positive

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Conflicts of interest

There are no conflicts of interest.

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