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Rapid, *de novo* development of isolated intracranial rosai-dorfman disease: A case report

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Abstract

Isolated intracranial Rosai-Dorfman Disease is rare. Here, we describe a patient who developed an asymptomatic, right parietal RDD lesion over 18 months while being followed radiographically for another brain lesion. To our knowledge, rapid, *de novo* radiographic formation of isolated intracranial RDD has never been reported in an asymptomatic patient.

Keywords

de novo development; isolated; intracranial; rosai-dorfman disease; sinus histiocytosis with massive lymphadenopathy

Introduction

In 1969, Rosai and Dorfman described a clinical syndrome characterized by bilateral painless cervical lymphadenopathy, fever, weight loss, leucocytosis, elevated erythrocyte sedimentation rate, and hypergammaglobulinemia. ¹ Initially termed sinus histiocytosis with massive lymphadenopathy (SHML), this rare, benign lymphoproliferative disease now bears their name. Extranodal involvement is common in RDD and can affect the soft tissues, bone, salivary glands, paranasal sinuses, thyroid, orbit, upper respiratory tract, skin, and testes. While the precise etiology of RDD is unknown, viral, inflammatory, and immunosuppressive etiologies have all been proposed.

Isolated intracranial RDD is extremely rare, with approximately 50 reported cases in the existing literature. Patients with this form of RDD almost invariably present with signs and

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symptoms referable to their intracranial lesion. Also, these patients tend to have neither lymphadenopathy nor associated systemic disease.

In general, for patients with isolated intracranial RDD, surgery with gross total resection of the lesion is the treatment of choice. Recurrence after a complete surgical resection is uncommon, and fractionated radiation, stereotactic radiosurgery, chemotherapy, and steroid therapy have all been utilized as treatment adjuncts. Histopathology and immunohistochemistry are requisite for diagnosis. The prognosis for patients with isolated intracranial RDD is generally good.

In the present report, the authors describe the case of a rapidly progressive, isolated intracranial RDD lesion that presented *de novo* in an asymptomatic patient. To our knowledge, this is the first report of such an unusual presentation of RDD.

Clinical details

A 46-year-old right-handed woman was referred to our institution with intermittent episodes of speech arrest. The episodes had been present for one year, occurred infrequently (every 2–3 weeks), and were not associated with loss of consciousness or amnesia. Her past medical history was significant for migraine headaches and Crohn's disease. The migraine headaches were long-standing and associated with aura (scintillating scotoma) and left hemifacial paresthesias. Medical treatment included propranolol for headache prophylaxis and 6-mercaptopurine immunosuppressive therapy for Crohn's disease.

The physical examination was normal with no evidence of lymphadenopathy or hepatosplenomegaly. The results of hematological and biochemical studies were also normal. Electroencephalography revealed evidence of cerebral dysfunction in the left frontotemporal region.

Initial magnetic resonance (MR) imaging revealed a heterogeneous lesion on T1- and T2-weighted sequences in the left insular cortex, likely representing a cavernous malformation. MR angiography showed no evidence of a vascular malformation. No other lesions were identified. It was felt that the episodes of speech arrest were simple partial seizures referable to the perisylvian cavernoma near the language area. The patient was placed on anticonvulsant therapy (Levetiracetam) with amelioration of the speech arrest episodes.

A surveillance MRI scan taken 18 months later showed stability of the left perisylvian cavernoma, but revealed a new lobulated, extra-axial, dural-based mass in the right posterior parietal region (Fig. 1). This mass was seen as isointense to gray matter on both T1- and T2-weighted imaging and showed avid, homogeneous enhancement following gadolinium administration. The differential diagnosis included an *en plaque* meningioma, hemangiopericytoma, and duralbased metastasis.

A right parietal craniotomy and gross total resection of the lesion was performed. Macroscopically, the lesion was fibrous, avascular, and densely adherent to the underlying pia. Microscopically, the lesion consisted of a dense network of lymphoplasmacytic cells and scattered, often multinucleated, foamy histiocytes with occasional

lymphocytophagocytosis (e.g., emperipolesis; Fig. 2). Immunohistochemistry showed strong staining with CD68 and S100 (Fig. 2), but no staining with CD1 α (not shown) - a pattern most consistent with RDD. A post-operative MRI (72 h after surgery) confirmed gross total resection. The patient did well after surgery and was discharged home without neurological deficits. Subsequent MR imaging at 6 months did not show any recurrence of the lesion.

Discussion

In this report, we describe a patient with rapidly progressive, *de novo* development of isolated intracranial RDD. To our knowledge, of the ~50 reported cases of isolated intracranial RDD reported in the literature, no patient has presented in such a unique fashion. Most, if not all, patients with isolated intracranial RDD have presented with symptoms and signs referable to their intracranial lesion.

While most patients with isolated intracranial RDD have a benign clinical course, some authors have reported intracranial RDD with aggressive, infiltrative features. Moreover, many patients with isolated intracranial RDD who have undergone subtotal resection of their lesion have experienced rapid radiographic progression following surgery. Symms et al.² reported on three patients with isolated intracranial RDD. One patient, a 21-year-old man who presented with global headaches and vomiting, underwent a subtotal resection of an extensive infra- and supra-tentorial lesion that tracked anteriorly along the tentorium, bilaterally to the cavernous sinus. The patient presented 6 months later with recurrence of his symptoms and a contrasted head CT showed significant bilateral disease progression. Di Rocco et al.³ described a 13-year-old girl who presented with severe headache and nausea/vomiting from a left frontal RDD lesion. Following burr hole placement with subtotal debulking, the patient rapidly developed a large recurrence over the ensuing 3 months. She subsequently underwent a craniotomy for gross total resection of the lesion, with satisfactory clinical results.

Interestingly, the majority of cases of rapid radiographic progression of isolated intracranial RDD have been after a biopsy or subtotal debulking. This latter observation suggests that surgical intervention may, in fact, increase the biological aggressiveness of RDD if the lesion is not completely resected. Furthermore, in our review of the literature, we were unable to find other cases of isolated intracranial RDD where no surgical intervention was performed initially and the disease process was followed with serial MR imaging. Our report is therefore the first to document rapidly progressive, *de novo* development of isolated intracranial RDD, and may provide valuable insights into the enigmatic natural history of this rare disease.

Conclusions

While the natural history and radiographic progression of isolated intracranial RDD is unknown, this lesion can manifest asymptotically in a *de novo* fashion. A rapidly progressive, dural-based, *de novo* lesion should raise suspicion for isolated intracranial RDD.

References

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Fig. 1. MR imaging shows a new, lobulated, extra-axial, duralbased mass in the right posterior parietal area. The lesion is seen as isointense to gray matter on T1-weighted sequences (A) with avid, homogeneous contrast enhancement (B) and isointense on T2-weighted (C) and fluid-attenuated inversion recovery sequences (D).

Fig. 2.

Histopathology and immunohistochemistry of the right posterior parietal lesion.

Hematoxylin and eosin (H&E) staining (A, low magnification) shows scattered, often multinucleated, foamy histiocytes with a dense network of lymphoplasmacytic cells and occasional emperipolesis (B, high magnification, arrowhead). Cells stained strongly with CD68 (C) and S100 (D), but not with CD1 α (not shown).