

CASE REPORTS

Pediatric isolated intraparenchymal cerebellar Rosai-Dorfman disease

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ABSTRACT

We illustrate the imaging findings of a case of isolated intraparenchymal cerebellar Rosai-Dorfman disease (RDD) in a child, which to our knowledge is the first reported case of such lesion in a child.

Key Words: Rosai-Dorfman disease, Intraparenchymal, Cerebellar, Pediatric, Child

1. INTRODUCTION

Rosai-Dorfman disease (RDD) was first described in 1969 as a benign proliferative lesion with systemic symptoms and lymphadenopathy.^[1] The disease affects mostly children and young adults with a slight male predilection.^[2] While RDD often involves lymph nodes, it can have extranodal involvement. Isolated intracranial RDD is extremely rare.^[3] It is especially rare in children where there are only about 10 cases reported in the literature.^[4-13] When lesions are intracranial, they are usually extra-axial with dura involvement, not in the brain parenchyma.^[14] Isolated intracranial RDD without dural attachment is extremely rare.^[15] We found only a few reported cases in the literature of isolated intraparenchymal RDD involving the cerebellum, all cases are in adults, even though the disease predominantly affects children and young adults.^[16] We present here a case of RDD isolated to the brain parenchyma of the cerebellum in a child, which to our knowledge has not been reported in the literature.

2. CASE PRESENTATION

The patient is a 14-year-old right-handed boy without significant past medical history who noted a sudden headache in the back of his head while watching TV. When the symptoms became more severe he went to a local emergency room. He underwent blood and urine tests and was sent home, because the tests were within normal limits. Two days later he had severe headaches again. He saw a physician and received injection of pain medication which helped the pain.

A week later, he underwent a CT scan, followed by an MRI. The imaging studies showed a right cerebellar lesion. After discussion with family, the decision was made to surgically resect the lesion, both for treatment of the lesion and for pathologic diagnosis. The patient underwent a suboccipital craniotomy, and the lesion was resected without complication. The most recent follow up MRI showed gross total resection of the cerebellar lesion with no evidence of recurrence. Patient returned to normal life and had no residual symptoms during his last clinical follow up 2 years later.

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3. IMAGING FINDINGS

CT showed the presence of a hyperdense intraparenchymal lesion in the right cerebellar hemisphere, with surrounding with edema (see Figure 1A).

MRI demonstrated a solitary 3.0 cm × 2.6 cm homogeneously enhancing lesion in the right cerebellar hemisphere (see Figure 1B). On the T2 FLAIR image, the lesion was slightly low in signal compared with cerebellum with sur-

rounding edema (see Figure 1C). On the T2 gradient echo image (GRE), a tiny focus of susceptibility signal void is present anteriorly, which could represent a tiny focus of hemorrhage (see Figure 1D). On the T1 weighted image, the lesion is isointense to the brain tissue (see Figure 1E). On the diffusion image (DWI), there is no evidence of restricted diffusion (see Figure 1F). There is no displacement of the fourth ventricle or hydrocephalus.

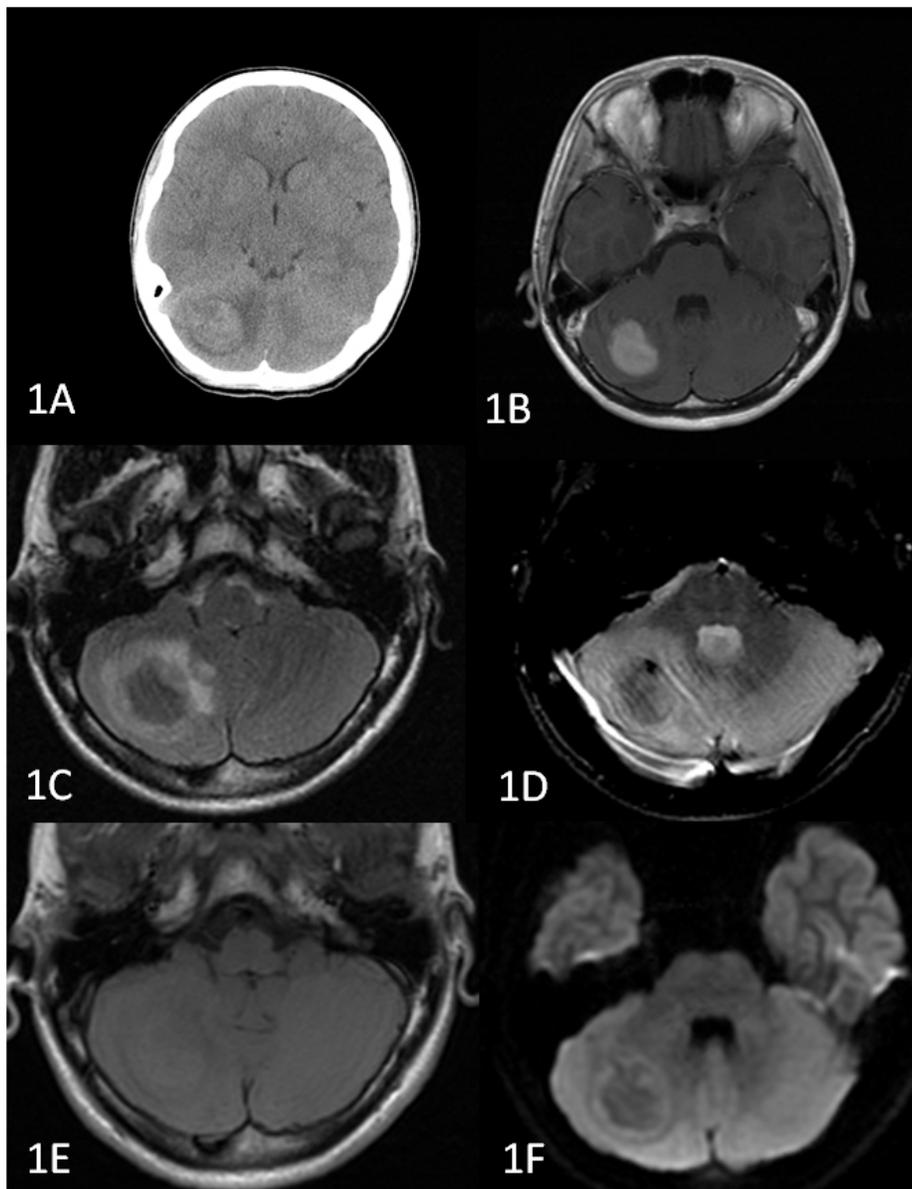


Figure 1. 1A: Noncontrast CT demonstrates a hyperdense intraparenchymal lesion in the right cerebellar hemisphere, with surrounding hypodensity compatible with edema. 1B: Post contrast T1 image shows a solitary 3.0 cm × 2.6 cm homogeneously enhancing lesion in the right cerebellar hemisphere away from the 4th ventricle. 1C: On T2 FLAIR image, the lesion is slightly low in signal compared with cerebellum; the hyperintensity surrounding the lesion is most consistent with edema. 1D: On T2 GRE, a tiny focus of susceptibility signal void anteriorly could represent a tiny focus of hemorrhage. 1E: On T1, the lesion is isointense to the brain tissue. 1F: On DWI, there is no evidence of restricted diffusion.

4. PATHOLOGICAL FINDINGS

Gross pathology shows an irregular ovoid resection of cerebellum measuring 3.5 cm × 2.0 cm × 1.5 cm. The specimen is firm. The fragments of cerebellum attached to the external

surface are gray-white and soft. The normal foliar pattern is noted focally. Within the specimen is a firm, yellow-tan tumor which is well-circumscribed. It measures up to 3.0 cm × 2.0 cm. In the center of the tumor is an area of stellate gray-white discoloration.

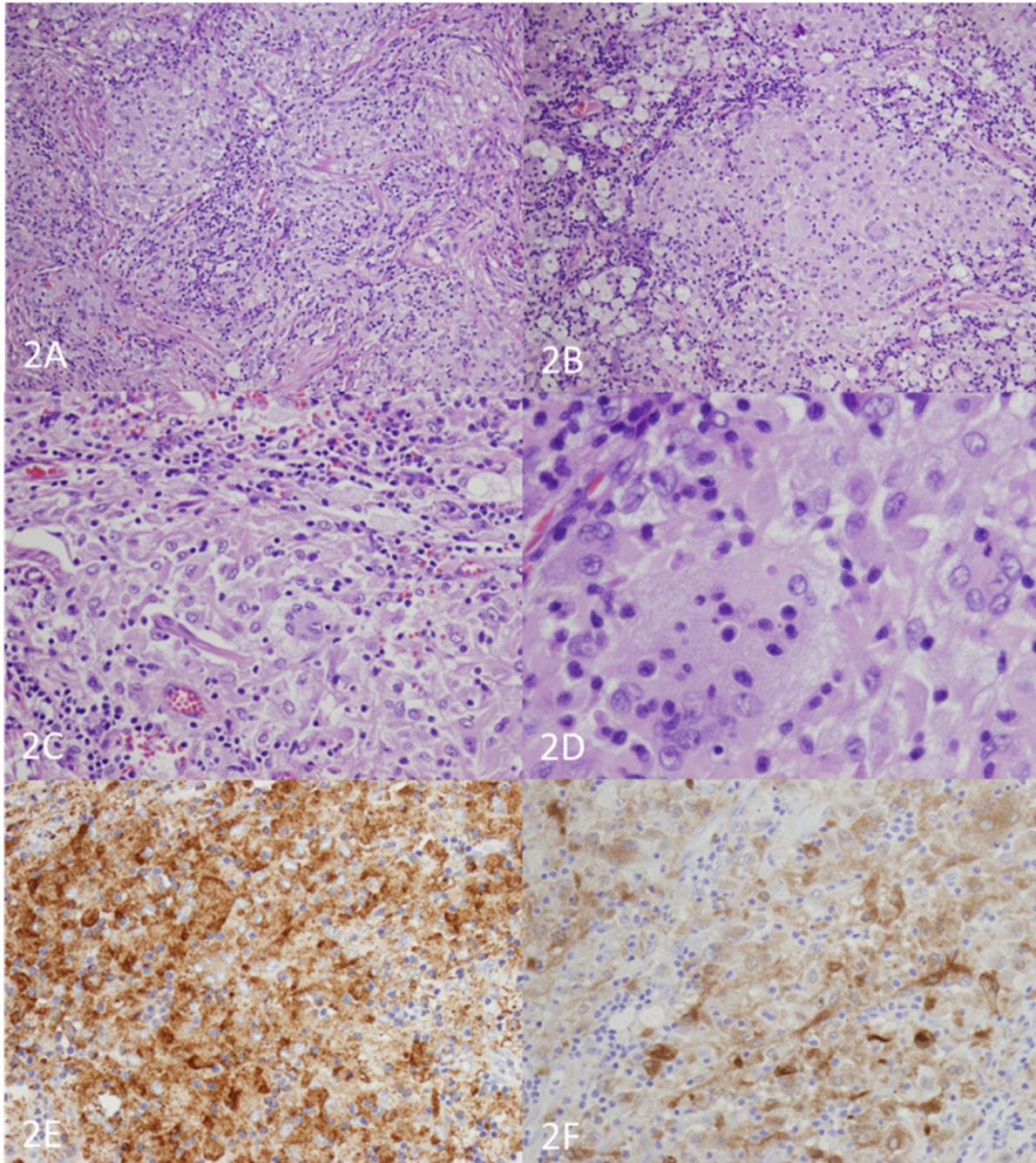


Figure 2. 2A: Histological section (H&E) shows lobules of histiocytes with pale granular cytoplasm, surrounded by mononuclear inflammatory cells. 2B: Histological section (H&E) shows multinucleated giant cells, foamy histiocytes, lymphocytes, plasma cells, and Touton-like giant cells. 2C: Histological section (H&E) shows giant cells, histiocytes, and mononuclear cells. 2D: Histological section (H&E) shows scattered lymphocytes within the cytoplasm of histiocytes (emperipolesis). 2E: Immunohistochemistry shows CD68 antibody+. 2F: Immunohistochemistry shows S-100 antibody+.

Histologic sections show the classic appearance of Rosai-Dorfman disease with lobular aggregates of histiocytes surrounded by thin connective tissue septae containing mature lymphocytes. There are nodules of granulomatous-type non-caseating lymphohistiocytic inflammation. Some of the histiocytes are multinucleated. Within the cytoplasm of histiocytes, scattered lymphocytes are noted (emperipolesis). The adjacent cerebellum shows Rosenthal fiber formation and reactive gliosis (see Figures 2A-2D).

Immunohistochemistry shows strong diffuse reactivity in the histiocytes for CD68 (see Figure 2E) and focal strong reactivity for S-100 (see Figure 2F) protein. A stain for organisms (GMS) is negative.

5. DISCUSSION

RDD disease, sinus histiocytosis with massive lymphadenopathy, rarely involves the intracranial structures without other sites of involvement. When it does, it commonly is dural-based, involving the frontal and parietal regions and the skull base. Extranodal disease usually involves eyes, salivary glands, thyroid and upper respiratory tract rather than brain.^[2] Isolated intraparenchymal Rosai-Dorfman disease is extremely rare.

This case is unique because it only involved cerebellum without nodal involvement, which has not been described before in children. Although the disease is characterized by sinus

histiocytosis and massive lymphadenopathy, our case does not have lymphadenopathy; his full lymph node imaging search was negative. His presentation is limited to brain parenchyma, not just intracranial space. We found only one other reported case of isolated intracranial parenchymal involvement in a child, which involved supratentorial thalamus and insula.^[12] We could not find another similar reported case of isolated intraparenchymal involvement in the infratentorial compartment in a child, as in our case.

This lesion is highly cellular on pathology, which explains why it is slightly hyperdense on CT and slightly hypointense on the MRI T2 and FLAIR sequences. A range of low to hyperintensity on T2 sequence has been described. In our case, gross total surgical resection resulted in no recurrence on follow up.

6. SUMMARY

To our knowledge, this case is the first reported case of isolated intraparenchymal cerebellar Rosai-Dorfman disease in a child. Despite of its rarity, physicians should be aware of this entity and take it into consideration when treating solitary cerebellar lesions even in children.

CONFLICTS OF INTEREST DISCLOSURE

The authors have no conflict of interest related to this publication.

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