Clinical Case Report

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Isolated intracranial Rosai–Dorfman disease mimicking petroclival meningioma in a child

Case report and review of the literature

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Abstract

Rationale: Rosai –Dorfman disease (RDD) is a rare, idiopathic, and non-neoplastic histioproliferative disease with distinctive entity of unknown etiology. Central nervous system (CNS) RDD is uncommon, hence, isolated intracranial RDD is extremely rare. So far only 6 cases of CNS RDD with the lesions originating from petroclival region have been reported. We present a case of isolated intracranial RDD mimicking petroclival meningioma.

Patient concerns: A 14-year-old girl was admitted at our hospital with a 3-month history of dizziness, slowly progressing headache, and 2-month history of instability in walking. Cranial nerve deficits, including left facial paralysis, left facial numbness and left hearing loss, were evident on examination.

Diagnoses: Initial diagnosis of petroclival meningioma was made according to preoperative magnetic resonance imaging.

Interventions: The lesion was resected subtotally and pathology confirmed RDD. The patient received gamma-knife treatment for the residual lesion.

Outcomes: The patient recovered well and the residual lesion significantly retrogressed on follow-up images.

Lessons: Preoperative diagnosis of petroclival RDD is full of challenges. Although surgical resection of lesions is an effective treatment option, total resection is not highly recommended because the surgery-related defect must be minimal. Patient with residual lesion can be put on steroid therapy and/or radiotherapy, especially for IgG4 positive subset of RDD.

Abbreviations: CNS = central nervous system, EMA = epithelial membrane antigen, GFAP = glial fibrillary acidic protein, H–B = House–Brackmann, MRI = magnetic resonance imaging, RDD = Rosai–Dorfman disease.

Keywords: gamma-knife, petroclival meningioma, radiotherapy, Rosai–Dorfman disease, steroid therapy

1. Introduction

Rosai–Dorfman disease (RDD) was reported in 1965, but Rose and Dorfman in 1969 conducted a detailed pathological study on it and referred to it as "sinus histiocytosis because of massive lymphadenopathy."[1] It was a benign disease characterized with massive painless cervical lymphadenopathy, fever, weight loss, and leukocytosis. About 40% of cases are extranodal and usually involved with the skin, upper respiratory system, orbits, bones, and endocrine glands.[2] Although central nervous system (CNS) cases have been reported, isolated petroclival RDD is very rare. To the best of our knowledge, there are only 6 of such case reported so far.[3–8] We therefore present a case of RDD with the lesion originating from the petroclival region and review literature on this rare presentation.

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2. Case report

Previous written and informed consent were obtained from the patient, and this study was approved by the ethics review board of West China Hospital of Sichuan University. A 14-year-old girl was admitted at our hospital with a 3-month history of dizziness, slowly progressing headache, and 2-month history of instability in walking. Cranial nerve deficits, including left facial paralysis (House–Brackmann [H–B] II degree), left facial numbness, and left hearing loss, were evident on examination. There was no lymphadenopathy on physical examination. Magnetic resonance imaging (MRI) revealed homogeneously contrast-enhancing petroclival giant lesion ($5 \times 6 \times 4$ cm in size) with extension into the left cavernous sinus (Fig. 1). The mass was hypointense on T1-weighted images and hyperintense on T2-weighted images and fluid-attenuated inversion recovery images. The diagnosis of petroclival meningoma was made before surgery.

The lesion was removed surgically via the right suboccipital retrosigmoid approach. Pathological examination of the lesion showed fibrous tissue with an infiltrate of inflammatory cells composed of histiocytes, lymphocytes, and plasma cells. The histiocytes contained abundant cytoplasm within intact lymphocytes (so-called emperipolesis). The histiocytes stained positive for S100 and CD68, but negative for CD1a, glial fibrillary acidic protein (GFAP), and epithelial membrane antigen (EMA). Lymphocytes showed about 15/hpf IgG4 positive (Fig. 2). The pathological presentations of the lesion were consistent with the diagnosis of petroclival RDD.

Postoperative MRI scan done on the 2nd day after the operation revealed residual solid lesion in the left petrous apex and cavernous sinus (Fig. 3A and D). The patient recovered well

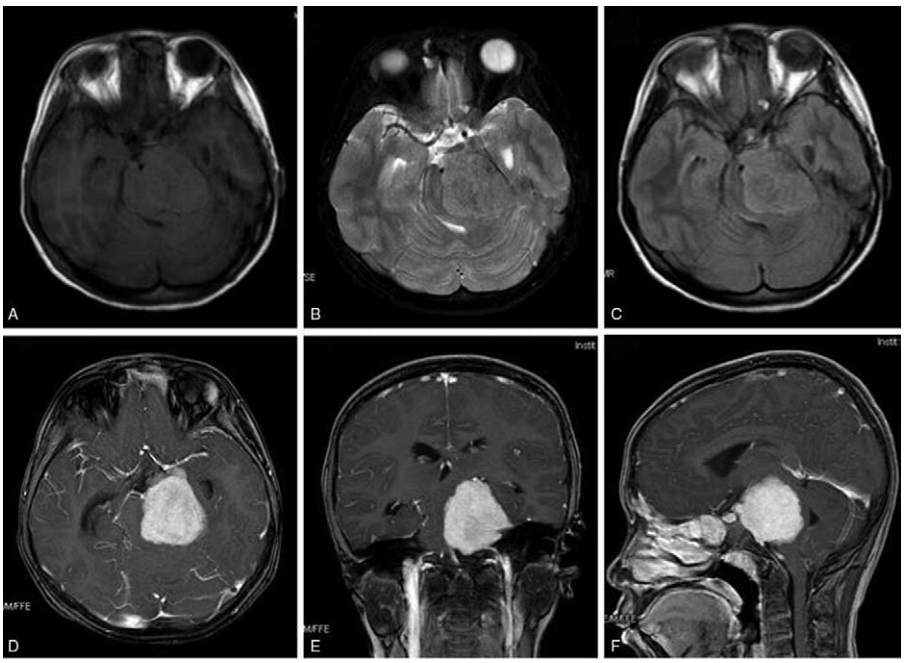

Figure 1. (A) Axial T1-weighted MRI showing a hypointense mass with severe compression of the brainstem in the left petroclival region. (B) Axial T2-weighted and (C) axial fluid-attenuated inversion recovery MRI showing the lesion as hyperintense. (D, E, F) Axial, sagittal, and coronal T1-weighted contrast-enhanced MRI showing the hyperintense petroclival lesion with extension into the left cavernous sinus. MRI = magnetic resonance imaging.

after the surgery with a left facial paralysis (H–B III degree). Gamma-knife surgery was performed to the residual tumor at 9th day after the subtotal resection. Scheduled clinical evaluation and imaging were done at 4, 12, and 18 months after gamma-knife surgery. Major symptoms like left facial paralysis and left hearing loss slightly improved at 4 months, but the left facial dysfunction recovered basically at 18 months after surgery. The residual lesion significantly retrogressed on follow-up images (Fig. 3B, C, E, and F).

3. Discussion

A total of about 219 patients with CNS RDD (including our case) have been reported in literature so far.[9–12] CNS manifestation of

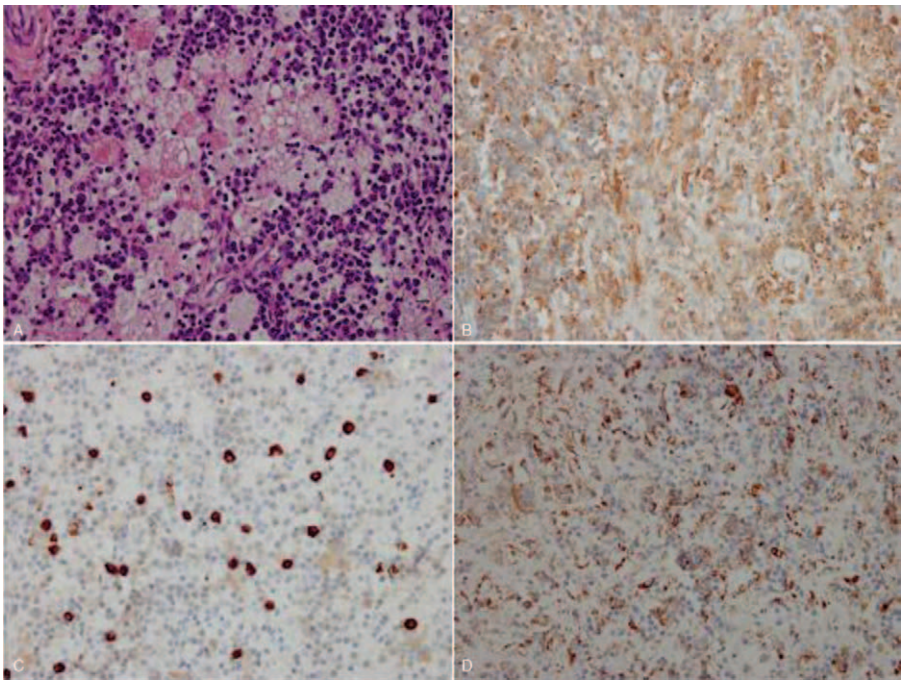

Figure 2. (A) Histologic sections demonstrating emperipolesis (lymphocytes within the abundant cytoplasm in histiocytes) (hematoxylin and eosin stain, ×400). The histiocytes showing positive reactivity for S-100 protein (B) and CD 68 (D) (×400). Lymphocytes showing about 15/hpf IgG4 positive (C) (×400).

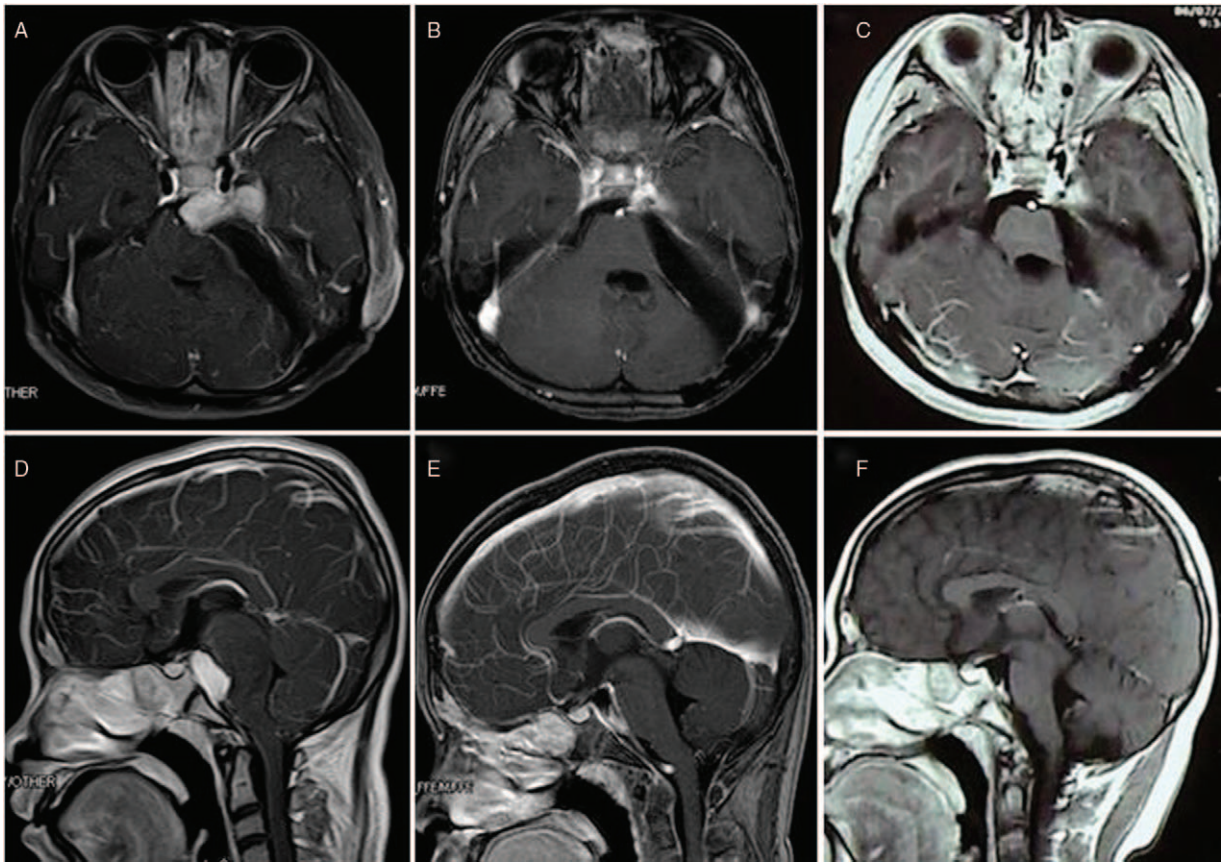

Figure 3. (A, D) Repeated T1-weighted contrast-enhanced MRI scan at the 2nd day after the operation showing residual lesion in the left petrous apex and cavernous sinus. The residual lesion significantly retrogressed on follow-up images at 4 months (B, E) and 12 months (C, F) after gamma-knife treatment.

RDD was reported in 180 cases while 39 cases had a systemic disseminated manifestation of the disease (Table 1). Furthermore, CNS manifestation of the disease has been reported in both adults and children with a male prevalence (M/F ratio: 1.8:1.0). Although majority of cases of CNS manifestation was located intracranial, extremely very few cases have been seen with spinal cord involvement. The lesions are usually multiple or single. Out of the 219 cases above, 174 (79.5%) presented with intracranial lesions while 26 (11.4%) had spinal lesions; 19 (9.0%) presented with both intracranial and spinal lesions (Table 1).

With regard to the intracranial presentations of the disease, very few cases have been reported with petroclival region involvement. So far only 6 cases of petroclival RDD have been reported in literature (Table 2). To the best of our knowledge, 1 case was reported but it was not a real petroclival RDD. The presentation of this particle case was unusual, because it involved a wide range of structures including the petroclival region, cavernous sinuses, suprasellar region, anterior cranial fossa, paranasal sinuses, nasal cavity, and the spinal cord.[6] We present the first case of RDD with the lesion originating from the petroclival region in a female child because all the 6 cases reported so far were in adult patients.

The first case of petroclival RDD was described by Kitai in 2001 in a 42-year-old woman who presented with progressing headache.[7] The mass was totally resected with good outcome. The severity of petroclival RDD depends on the degree of

Table 1

Characteristics of CNS RDD cases according to present and previous reports.

	Total RDD with CNS involvement (n = 219), N	RDD with isolated CNS involvement (n = 180), N	Systemic RDD with CNS involvement (n = 39), N
Sex			
Male	143	120	23
Female	76	60	16
Location			
Intracranial	174	149	25
Spine	26	17	9
Intracranial and spine	19	14	5

CNS = central nervous system, RDD = Rosai–Dorfman disease.

Table 2

Summary of intracranial petroclival Rosai–Dorfman disease cases in the literature.

Author, year	Age, y/sex	Presentation	Location	Treatment	Follow-up, mo
Kitai, 2001	42, F	Headache	Petroclival	Total resection	NA
Andriko, 2001	50, M	Headache	Left petroclival	Subtotal resection	17
Hadjipanayis, 2003	52, M	Fever, headache, diplopia, left facial paresthesias	Petroclival, left cavernous sinus	Subtotal resection + radiosurgery	15
Kaminsky, 2005	32, M	Chronic nasal obstruction, left trigeminal pain	Petroclival, cavernous sinus, spinal cord, . . .	Subtotal resection	NA
Gupta, 2005	15, M	Headache, vomiting, visual deterioration	Bilateral petroclival	Subtotal resection + steroids	12
Wang, 2010	47, M	Trigeminal neuralgia	Left petroclival	Total resection	6
Our case	14, F	Dizziness, headache, walking instability	Petroclival, left cavernous sinus	Subtotal resection + radiosurgery	18

NA = not available.

compression of the brainstem. Our patient presented the walking instability because the brainstem pyramidal tract was severely compressed by lesion. In most of the intracranial RDD, the lesions appeared to be dural-based. Therefore, petroclival RDD lesions are likewise closely related to the dura mater. Preoperative misdiagnosis of petroclival RDD seems inevitable because the radiological features of homogeneous contrast-enhancing meningeal-based mass on T1-weighted imaging highly mimic petroclival meningiomas.

Out of the 6 cases outlined in Table 2, only 2 patients had total resection without adjunctive therapy. Two other patients had subtotal resection without adjunctive therapy. One had neither radical resection nor radiosurgery because lesions were extensive. The other one had good prognosis which confirms the benign nature of the disease. The rest of 2 patients who had subtotal resection were put on steroid medications and/or radiosurgery after surgery. The outcome of total tumor removal was good without recurrence compared with subtotal resection. Petroclival RDD can be safely treated with radiosurgery when the residual lesion is located in a critical location. Radiosurgery is also a recommended treatment option for petroclival RDD when complete resection carries a risk of significant morbidity.[5] In our case, multiple factors such age, sex, cranial nerve deficits, size of the mass, as well as critical location were taken into broad consideration which made us hesitate to achieve complete removal of the lesion. Residual lesion in the left petrous apex and cavernous sinus was treated with gamma-knife surgery in our patient. Scheduled follow-ups revealed that gamma-knife surgery therapy was effective and the girl made a good recovery.

It is not difficult to make definite diagnosis pathologically because of emperipolesis. Microscopic examination revealed chronic inflammation with infiltrate of histiocytes, lymphocytes, and plasma cells in the fibrous stroma.[12] Emperipolesis exhibits that the majority of histiocytes contains well-preserved lymphocytes within their cytoplasm. The histiocytes react positively with S-100 protein and CD68 but negatively with GFAP and EMA. Though immunohistochemical examination revealed about 15/hpf IgG4 positive lymphocytes in our case, no extra evidence show that RDD could be IgG4-related disease.[13] The finding above has never been mentioned in earlier petroclival RDD. Only a small percentage of RDD cases have proven to be responsive to steroids, which means IgG4 positive subset of RDD represents a more steroid responsive group just like IgG4-related disease.[13] We did not give our patient steroids because she had gamma-knife surgery. Therefore, we propose that steroid drugs could be useful when radiosurgery has failed, especially for IgG4 positive subset of RDD. Although the disease is proven to be benign, there are individual differences in the prognosis of patients, so a longer follow-up of patients to monitor their post-treatment clinical presentations is absolutely necessary.

4. Conclusion

Preoperative diagnosis of petroclival RDD is puzzling. Although surgical removal of lesions is an effective treatment option, total resection is not highly recommended to the petroclival RDD to decrease the surgery-related deficits. Radiotherapy and/or steroid drugs may be useful for patients with residual lesion, especially for IgG4 positive subset of RDD.

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