Clinical Case Report

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# Intracranial Rosai-Dorfman disease with the petroclival and parasellar involvement mimicking multiple meningiomas

## A case report and review of literature

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**Abstract**

**Rationale:** Rosai-Dorfman disease (RDD) is a rare non-Langerhans cell histiocytosis. Petroclival RDD is extremely rare. To the best of our knowledge, only 7 cases of petroclival RDD have been reported so far. Herein, we present the 8th case of intracranial RDD with the petroclival and parasellar involvement mimicking multiple meningiomas.

**Patient concerns:** A 57-year-old woman presented with a 1-year history of vision diminution and 1-month hearing loss in her right ear.

**Diagnoses:** Contrast-enhanced Magnetic resonance imaging (MRI) of the brain demonstrated multiple well-defined, homogenous mass which closely related to the dura mater in the bilateral parasellar and petroclival regions range from the basement of anterior to posterior cranial fossa. The lesions were T1 isointense, T2 hypointense, and homogeneously enhanced. Initial diagnosis of multiple meningiomas was made according to MRI findings. Final diagnosis of RDD was confirmed by histopathological and immunohistochemical examinations after subtotal surgical resection.

**Interventions:** The patient received subtotal resection because multiple lesions were extensive.

**Outcomes:** The vision diminution recovered well after the surgery but the hearing loss in her right ear was still persisted.

**Lessons:** Although rare, a standard RDD typically are dural-based, extra-axial, well-circumscribed masses mimicking meningioma, and presenting with characterized hypo to isointense on T1-weighted images, hypo to isointense on T2-weighted images, and obvious enhancement. Resection of the intracranial lesion is the most effective treatment. In case of subtotal resection, the application of adjunctive radiotherapy and/or steroid agents should be advised. Final diagnosis of RDD should be confirmed by histopathological and immunohistochemical examinations.

**Abbreviations:** CNS = central nervous system, CT = computed tomography, GFAP = glial fibrillary acidic protein, MRI = magnetic resonance imaging, RDD = Rosai-Dorfman disease.

**Keywords:** central nervous system, magnetic resonance imaging, meningiomas, petroclival region, Rosai-Dorfman disease

## 1. Introduction

Rosai-Dorfman disease (RDD) is a rare non-Langerhans cell histiocytosis first described in 1965.[1] Then in 1969, 2 pathologists Juan Rosai and Ronald Dorfman described 34 cases of the same entity under the name sinus histiocytosis with massive lymphadenopathy.[2] RDD can involve any nodal or extranodal site in all age groups, but most commonly presents as bilateral cervical lymphadenopathy in adolescents and young adults.[3] Approximately 43% of patients with RDD present with extranodal involvement with the orbits, superior airway, bones, skin, gastrointestinal tract, genitourinary tract, endocrine glands and central nervous system(CNS).[4] CNS involvement is rare and described in about 5% of cases. CNS involvement of RDD without nodal disease is very unusual, which presents with single or multiple lesions.[5,6] Although RDD cases in the CNS have been reported, petroclival RDD is very rare. To the best of our knowledge, only 7 cases of petroclival RDD have been reported so far.[6–12] Herein, we present a case of intracranial RDD with the petroclival and parasellar involvement mimicking multiple meningiomas both clinically and radiologically in a 57-year-old woman. A literature review was also performed to investigate clinical data, radiological findings, treatment protocols and disease prognosis pertaining to petroclival and parasellar RDD.

## 2. Case presentation

This study was approved by the ethics review board of the Second Affiliated Hospital, School of Medicine, Zhejiang University, and informed written consent was obtained from the patient for

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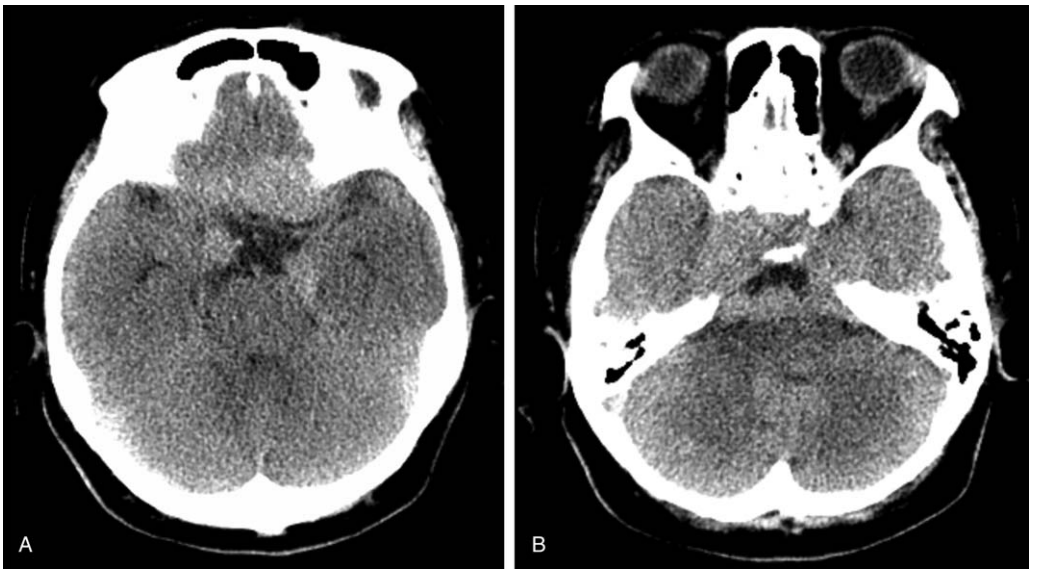


**Figure 1.** Brain computed tomography (CT) demonstrated multiple well-defined, homogenous, hyperdense masses in the basement of anterior-middle-posterior cranial fossa.

publication of this case report and accompanying images. A 57-year-old female was admitted at our hospital with a 1-year history of vision diminution and 1-month hearing loss in her right ear. There was no lymphadenopathy on physical examination. Brain computed tomography (CT) demonstrated multiple well-defined, homogenous, hyperdense masses in the basement of anterior-middle-posterior cranial fossa (Fig. 1). Contrast-enhanced Magnetic resonance imaging (MRI) of the brain showed multiple well-defined, homogenous mass which closely related to the dura mater in the bilateral parasellar and petroclival regions range from the basement of anterior to posterior cranial fossa (Fig. 2). The lesions were isointense on T1-weighted images (Fig. 2A), hypointense on T2-weighted images (Fig. 2B), and showed homogeneous enhancement following the intravenous administration of gadolinium (Fig.2C–D). The left optic nerve is compressed by the mass in the left basement of anterior cranial fossa. The diagnosis of multiple meningiomas in the basement of anterior-middle-posterior cranial fossa was made before surgery.

The lesion in the left basement of anterior cranial fossa was removed surgically via the left fronto-temporal approach, because this lesion compressed the left optic nerve and caused significant vision diminution. Histolopathological examination showed fibrous tissue with an infiltrate of inflammatory cells composed of histiocytes, lymphocytes, and plasma cells (Fig. 3). The histiocytes contained abundant cytoplasm within intact lymphocytes, which was called emperipolesis. Immunohistochemical results showed stained positive for S100, CD163 and CD68, but negative for CD1a, glial fibrillary acidic protein (GFAP). These pathological presentations of the lesion were compatible with the diagnosis of RDD.

Postoperative MRI scan was performed on the 3rd day after the operation revealed residual multiple solid lesions in the basement of anterior-middle-posterior cranial fossa and the lesion in the left basement of anterior cranial fossa had been removed (Fig. 4). The vision diminution recovered well after the surgery but the hearing loss in her right ear was still persisted. Considering the treatment cost and the curative effect uncertainty of adjuvant therapy, this patient was not treated using adjuvant therapy because the patient refused further treatment after subtotal resection. The patient was followed up by telephone after 18 months, and the patient was alive with disease, but the hearing loss in her right ear was still persisted.

## 3. Discussion

RDD is considered as a benign, non-neoplastic, self-limiting histiocytic disease. Its etiology and pathogenesis remain poorly understood. CNS manifestations of RDD only account for 5% of cases.[13] RDD can occur in both adults and children with a male predominance.[13] The clinical symptoms depend on the location (such as the cerebral convexity, parasagittal region, cranial base, suprasellar region, cavernous sinus, and petroclival region), and includes headache, nausea and vomiting, dizziness, seizures, cranial nerve deficits, and weakness.[6,14] Our case involving the suprasellar region and petroclival region produced visual loss in her left eye and hearing loss in her right ear, because the lesions involved the left optic nerve and the right acoustic nerve.

To the best of our knowledge, only 7 cases of RDD with petroclival region involvement have been reported in literature so far (Table 1). Of these 7 cases of petroclival RDD, only 1 case showed multiple mass which involved the petroclival region, cavernous sinuses, suprasellar region, anterior cranial fossa, paranasal sinuses, nasal cavity, and the spinal cord.[9] We present the second case of RDD with multiple mass which involved the petroclival and parasellar regions.

Of these 7 cases of petroclival RDD in Table 1, two other cases received total resection. In addition, 6 cases just received subtotal resection. After subtotal resection, 2 cases further received radiation, 1 case further received steroid medications, and the other 3 cases had subtotal resection without adjunctive therapy. The 2 cases of total tumor removal were without recurrence. Petroclival RDD can be safely and effectively treated using adjuvant radiotherapy when the residual lesion is located in a critical location.[8] Moreover, the application of adjuvant

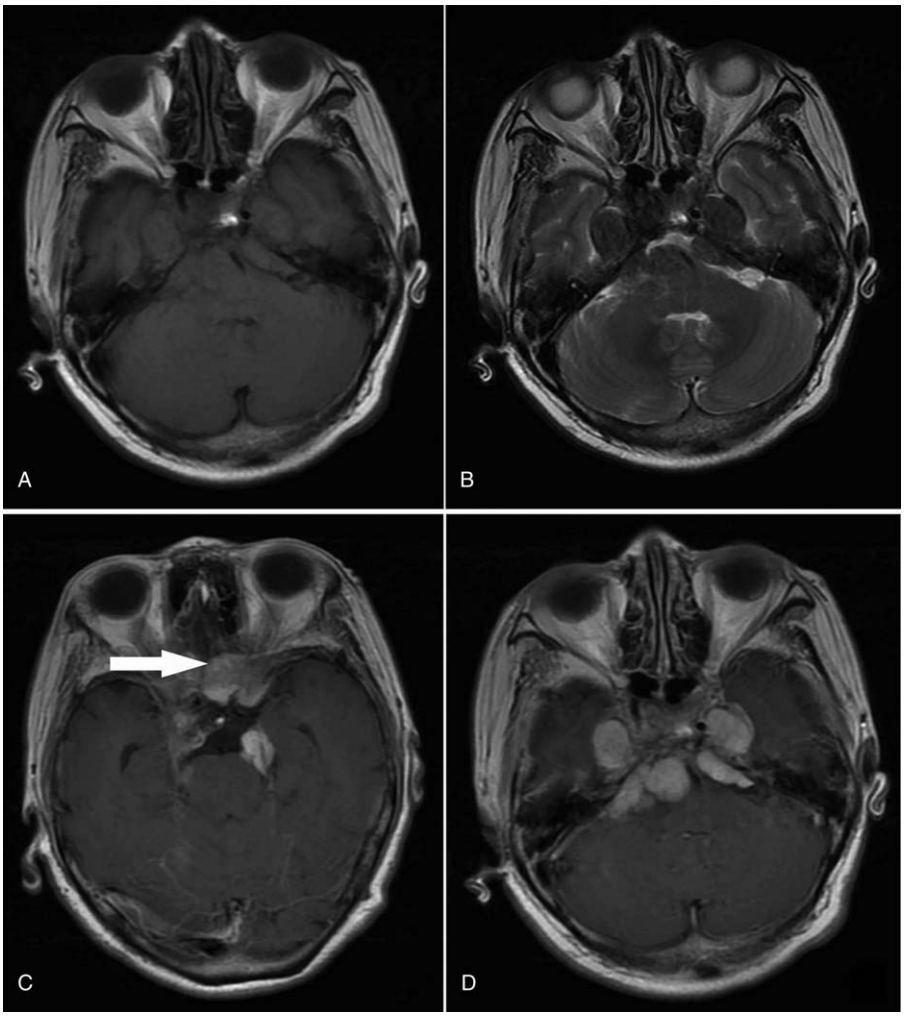


**Figure 2.** Contrast-enhanced Magnetic resonance imaging (MRI) of the brain showed multiple well-defined, homogenous mass which closely related to the dura mater in the bilateral parasellar and petroclival regions range from the basement of anterior to posterior cranial fossa (A–D). The lesions were isointense on T1-weighted images (A), hypointense on T2-weighted images (B), and showed homogeneous enhancement following the intravenous administration of gadolinium (C–D). The left optic nerve is compressed by the mass in the left basement of anterior cranial fossa (arrow).

treatment with steroid agents may be beneficial for the cases of subtotal resection.[4,8] Our case received subtotal resection because multiple lesions were extensive in the bilateral parasellar and petroclival regions across the basement of anterior-middle-posterior cranial fossa. The patient was followed-up by telephone after 18 months, and the patient was alive with disease, but the hearing loss in her right ear was still persisted.

Although the disease is proven to be benign, there are individual differences in the prognosis of patients. A previous study found intracranial lesions regrowth or recurrence of symptoms in 14% of 29 patients with a mean follow-up period of 10.1 years.[15]

The typical radiological findings of intracranial RDD show dural-based, extra-axial, well-circumscribed masses mimicking meningioma.[16–19] On CT, intracranial RDD are homogeneous hyperdense or isodense masses. MRI is currently the optimal diagnostic modality for evaluating lesions of intracranial RDD. On T1-weighted images, the lesions usually appear as isointense or hyperintense masses with clear borders relative to the peripheral brain parenchyma; on T2-weighted images, the lesions usually appear as isointense masses with possible intralesional hypointense foci. On contrast-enhanced T1-weighted images with gadolinium, the lesions demonstrate homogeneously or inhomogeneously obvious enhancement, and the dural tail sign can commonly be found.[16–19] In the present case, CT demonstrated multiple well-defined, homogenous, hyperdense massed; MRI showed multiple well-defined, homogenous masses which closely related to the dura mater in the bilateral parasellar and petroclival regions; the lesions were isointense on T1-weighted images, hypointense on T2-weighted images, and homogeneously enhanced. These above radiological findings closely mimic meningiomas, thus our case was misdiagnosed as meningioma before surgery, in agreement with other cases reported in the literature.[12,20] Preoperative radiological findings using the current MRI sequences are difficult to preoperatively distinguish RDD from meningiomas, but the absence of hyperostosis, bony erosion, or calcification should suggest RDD as a differential diagnosis of meningiomas.[17] Final diagnosis of RDD can only be confirmed by histopathological and immunohistochemical examinations. Typically, the histiocytes contained abundant cytoplasm within intact lymphocytes (so-called emperipolesis) and histiocytes

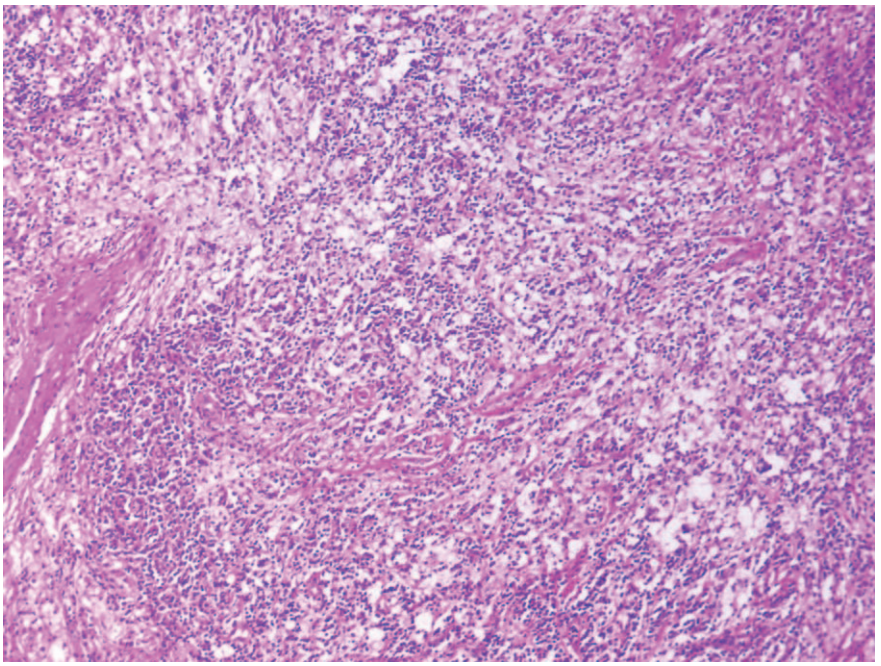

**Figure 3.** Histolopathological examination showed fibrous tissue with an infiltrate of inflammatory cells composed of histiocytes, lymphocytes, and plasma cells. hematoxylin and eosin staining; magnification, ×10.

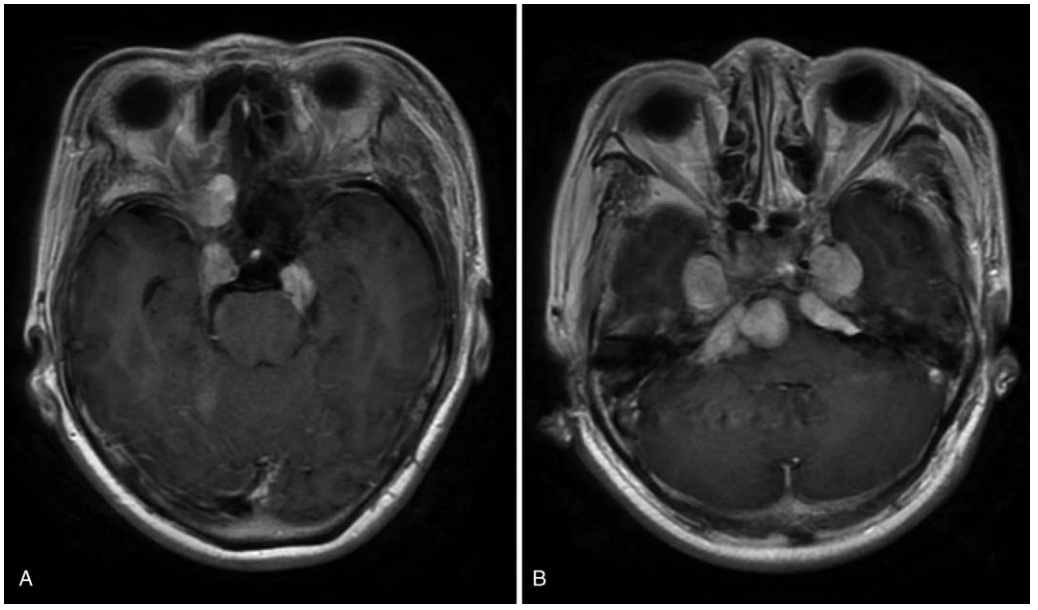


**Figure 4.** Postoperative MRI scan was performed on the 3rd day after the operation revealed residual mutiple solid lesions in the basement of anterior-middle-posterior cranial fossa and the lesion in the left basement of anterior cranial fossa had been removed.

**Table 1**

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

| | Authors | Sex | Age | Location | Presentation | Treatment | Follow-up/outcomes |
|---|---|---|---|---|---|---|---|
| 1 | Andriko et al[6] | M | 50 | Left petroclival | Headache | Subtotal resection | 17 mo, AWD |
| 2 | Kitai et al[10] | F | 42 | Petroclival | Headache | Total resection | NA, NED |
| 3 | Hadjipanayis et al[8] | M | 52 | Petroclival,left cavernous sinus | Fever, headache, diplopia, left facial paresthesias | Subtotal resection, and radiotherapy | 15 mo, AWD |
| 4 | Kaminsky et al[9] | M | 32 | Petroclival,cavernous sinus,suprasellar region. | Chronic nasal obstruction, left trigeminal pain | Subtotal resection | NA, AWD |
| 5 | Gupta et al[7] | M | 15 | Bilateral petroclival | Headache, vomiting, visual deterioration | Subtotal resection, and steroids | 12 mo, AWD |
| 6 | Wang et al[11] | M | 47 | Left petroclival | Trigeminal neuralgia | Total resection | 6 mo, NED |
| 7 | Yang et al[12] | F | 14 | Petroclival, left cavernous sinus | Dizziness, headache, walking instability | Subtotal resection, and radiotherapy | 18 mo, AWD |
| 8 | Present case | F | 57 | Bilateral petroclival, parasellar | visual loss and hearing loss | Subtotal resection | 18 mo, AWD |

AWD = alive with disease; NA = not available; NED = no evidence of disease.

stain positive for S-100 protein and CD68, but negative for CD1a.[21]

## 4. Conclusion

A standard RDD would typically be dural-based, extra-axial, well-circumscribed masses mimicking meningioma, and presenting with characterized hypo to isointense on T1-weighted images, hypo to isointense on T2-weighted images, and obvious enhancement. Resection of the intracranial lesion is the most effective treatment. In case of subtotal resection, the application of adjunctive radiotherapy and/or steroid agents should be advised. Final diagnosis of RDD should be confirmed by histopathological and immunohistochemical examinations.

## Author contributions

**Conceptualization:** Chao Wang.
**Data curation:** Chao Wang.
**Resources:** Chao Wang.
**Supervision:** Pingding Kuang.
**Validation:** Fangfang Xu.
**Writing – original draft:** Ling Hu.
**Writing – review & editing:** Ling Hu.

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