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Case Report

Intracranial Rosai-Dorfman Disease - a Case Report and a Review of the Literature

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Rosai-Dorfman disease (RDD) is an uncommon, non-neoplastic benign lymphoproliferative disease characterized by prominent, painless cervical lymphadenopathy with fever and leukocytosis. RDD is histologically characterized by emperipolesis, where large histiocytes become infiltrated with lymphocytes and plasma cells. Intracranial RDD is extremely rare. Only 79 cases have been reported, including the present case. Intracranial RDD is associated with headaches, seizures, and numbness caused by increased intracranial pressure. A 67-year-old Japanese woman presented with dizziness and was diagnosed with a tumor in the cerebral falx. The preoperative radiological diagnosis was meningioma. She had no lymphadenopathy. The patient underwent a craniotomy and tumor resection. The tumor consisted of lymphoid tissue with scattered lymph follicles. The infiltrating histiocytes showed emperipolesis. The histiocytes were immunoreactive for S-100 protein and CD68 and negative for CD1a, leading to the diagnosis of intracranial RDD. The postoperative course was uneventful without further therapy. The dizziness had not re-appeared and MRI demonstrated no recurrence of tumors for 7 months. Intracranial RDD shows a male predominance and occurs later in life than nodal RDD. The clinical manifestations and prognosis are variable depending on the location of the tumor and treatment. Most intracranial RDD have a benign course, but long-term follow-up is important, because recurrence has been observed.

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Introduction

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is an uncommon, non-neoplastic benign lymphoproliferative disease characterized by prominent painless cervical lymphadenopathy with fever and leukocytosis.[1] Other clinical features of RDD are elevated erythrocyte sedimentation and polyclonal hypergammaglobulinemia.[2]

RDD is histologically characterized by emperipolesis, where large histiocytes become infiltrated with lymphocytes and plasma cells.[3] The disease involves extranodal sites including the skin, salivary glands, orbit, liver and upper respiratory tract.[2] RDD involving intracranial sites is rare, and intracranial Rosai-Dorfman without lymphadenopathy is very rare.[4] Intracranial RDD presents headaches, seizures, and numbness caused by increased intracranial pressure. We report a case-study of a patient with intracranial RDD lacking lymphadenopathy and other organ involvement. We also provide a short review of the RDD literature.

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Case Report

A 67-year-old woman presented with dizziness and was diagnosed with a tumor in the cerebral falx. A preoperative MRI revealed a mass (1.5×1.3×1.0 cm) in the left frontal side of the longitudinal fissure. The lesion demonstrated high signal intensity on T1-weighted images (Figure 1A) and isointensity to hypointensity on T2-weighted images (Figure 1B). On contrast-enhanced T1-weighted images, the lesion was heterogeneously enhanced and showed dural tail signs, indicating the extra-axial location (Figure 1C). There was no evidence of brain edema surrounding the lesion. The preoperative radiological diagnosis was meningioma.

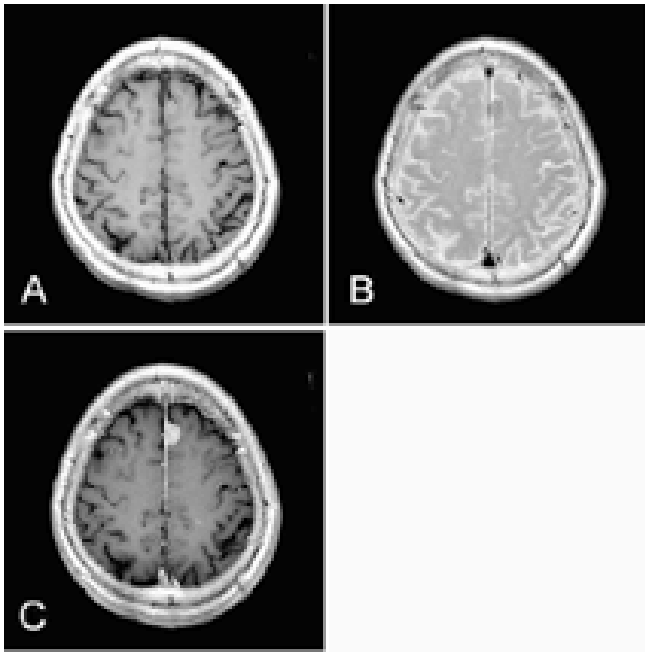

Figure 1. Axial MRI findings.
The lesion demonstrates high signal intensity on T1-weighted image (A), isointensity to hypointensity on T2-weighted image (B), and heterogeneous enhancement and dural tail sign on contrast-enhanced T1-weighted image (C).

Physical examination disclosed no lymphadenopathy. Hematological examination showed a white blood cell count of $6.5\times10^3/\mu l$ ($3.5\text{-}9.1\times10^3$), platelet count of $10.4\times10^4/\mu l$ ($13.0\text{-}36.9\times10^4$), platelet distribution wide of 11.5 fl (12.3-15.2), red blood cell count of $3.32\times10^6/\mu l$ (3.76-5.00 ×10), Hb of 10.3 g/dl (11.3-15.2), and Hct of 30.4% (33.4-44.9). C reactive protein (CRP) was elevated at 1.67 (<0.17).

The patient underwent a tumor resection after craniotomy. The tumor was reddish and well-differentiated from the brain, and was attached to the cerebral falx. Total resection was achieved.

The tumor consisted of lymphoid tissue with scattered lymph follicles. In the sinus area between the follicles, there were pale-staining areas. There were also fibrous scar-like areas (Figure 2A). The infiltrating histiocytes contained abundant pale-staining cytoplasm and round nuclei, each with a defined nucleolus. This specific feature of engulfment of the erythrocytes and lymphocytes is called emperipolesis (Figure 2B). There were no Russell bodies or Dutcher bodies in the plasma cells.

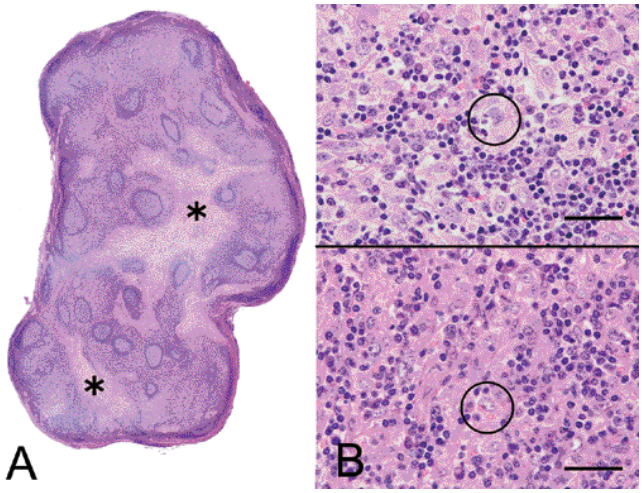

Figure 2. Pathological findings.
A) Scanning magnification.
The tumor consisted of lymphoid tissue with scattered lymph follicles. In the sinus area between the follicles, there are pale-staining areas. There also are fibrous scar-like areas (*).
B) High-power view of sinus area.
The pale sinus areas contain lymphocytes, plasma cells, and histiocytes. The infiltrating histiocytes contain abundant pale-staining cytoplasm and round nuclei with a definite nucleolus. They show the distinguishing feature of engulfment of the lym phocytes (upper) and erythrocytes (lower), so-called emperipolesis (circle). (Original magnification ×400, bar = 50 m)

On immunohistochemical examination, infiltrating plasma cells showed small amounts of kappa and lambda light chains. The infiltrating lymphocytes were a mixture of B and T cells, showing CD20 and CD3 positivity, respectively. The histiocytes were immunoreactive for S-100 protein and CD68, but negative for CD1a (Figure 3A, B, C). There were no meningeal cells immunoreactive for epithelial membrane antigen (EMA) in the lesions (Figure 3D). Periodic acid-Schiff and Grocott methenamine silver stain failed to identify any organisms.

Based on these histological and immunohistochemical findings, the lesion was diagnosed as intracranial RDD.

The postoperative course was uneventful and the patient was discharged 7 days after the operation without further therapy. Seven months after the operation, the dizziness had not re-appeared and MRI demonstrated no recurrence of the tumor.

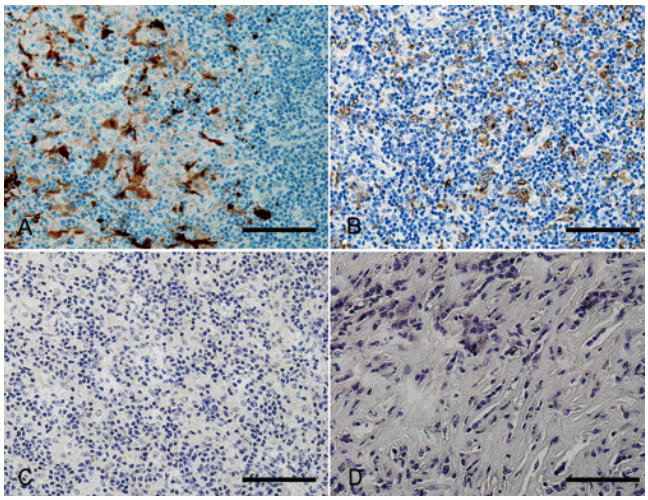

Figure 3. Immunohistochemical findings.
The infiltrating histiocytes in the sinus are immunopositive for S-100 protein (A). The reactivity varies from strong to weak. The histiocytes are also immunoreactive for CD68 (B), and are immunonegative for CD1a (C). There were no EMA-positive cells in the lesion including spindle cell elements (D). (Original magnification ×200, bar = 100μm)

Literature review

We found a total of 79 reported cases of intracranial RDD, including the present case (Table 1).

Nearly all cases occurred in the intracranial meningeal regions. Other locations included 12 cases in sellar and pituitary areas,[5-11, 21] 5 cases in intraparenchymal areas,[12-16] and 1 case in the fourth ventricle.[17] Sixteen cases showed multiple intracranial lesions.[11, 18-26] 20 cases had extracranial lymphadenopathy and other extra-nodal lesions.[4, 10, 11, 18, 20, 25, 27-36] The sizes of lesions ranged from 1.2 to 8.0 cm.

Cases ranged in age from 2 to 78 years (mean = 38.9 years). There was a male predominance (48 males and 30 females, male:female ratio = 1.6:1). There was no difference in the age between intracranial RDD with or without extracranial lesions.

Symptoms included headaches, seizures, visual problems, and others. (Table 1)

Most of the cases were diagnosed as meningioma preoperatively. Other preoperative diagnoses included meningitis,[10, 45] glioma,[13] and lymphoproliferative lesion.[3]

Treatments included total resection in 34 cases, subtotal resection in 26 cases, resection without comments in 10 cases, and biopsy sampling in 8 cases. For additional therapy after total resection, 2 cases received radiotherapy[9, 15] and 1 case received steroid therapy.[9] After subtotal resection, 5 cases received radiotherapy[22, 32, 35, 40, 44] and 4 cases received steroid therapy.[23, 35, 39, 42] After biopsy sampling, 6 cases were treated with steroid therapy,[6, 16, 33, 36, 46] 2 cases with radiotherapy[16, 36] and 1 case with anti-epileptic therapy,[47] and 1 case with no treatment.[7]

Tumors recurred in 4 cases of the 33 cases with total resection[9, 25, 30] and in 3 cases of the 26 cases with subtotal resection,[27, 29, 48] regardless of postoperative therapy. There was only 1 case that resulted in death, probably not caused by RDD, that occurred 5 days postoperatively.[21, 47]

Table 1. Published cases of intracranial Rosai-Dorfman disease.

No.	Author, year	Age and Sex	Symptoms	Location/Size(cm)/Extracranial	Preoperative Diagnosis*1	Treatment*2	Prognosis*3
1	Foucar 1982[4]	21M	lt facial numbness, lt decreased hearing	lt C-P angle	-	R	-
2		28F	headaches, dizziness	lt parietal/-/lymphadenopathy	-	R+Ste	-
3		55F	headaches	frontal/-/skin, rt nasal fossa	-	-	-
4	Trudel 1984[40]	28M	lt facial numbness and paresthesias, lt hearing loss	lt petrous bone	meningioma	SR+Rad	14 M G
5	Carey 1987[27]	35M	seizures	parasagittal/-/lymphadenopathy	meningioma	SR	3 Y Rec
6	Asai 1988[51]	39M	lt proptosis, lateral gaze disturbance	lt occipital	-	SR	3 Y RS
7	Song 1989[18]	30M	seizure, visual and hearing loss, focal palsy	cranial base/-/lymphadenopathy	-	SR	-
8	Lopez 1989[52]	35M	lt headaches	lt C-P angle and cavernous sinus/2.0	-	SR	12 M G
9	Bhattacharjee 1992[5]	78M	visual impairment	sellar	-	SR	7 M NR
10	Shaver 1993[42]	5M	III, IV, V, VI, VII palsies	lt cavernous sinus	-	SR+Ste	1 M G
11	Kim 1994[19]	50M	seizures	rt parietal double	meningioma	TR	6 M NR
12	Ng 1995[6]	22M	polydipsia, polyuria	pituitary/1.2	-	Bx+Ste	-

13	Clark 1996[53]	38F	headaches	lt parietal	meningioma	TR	4 Y NR
14	Panicker 1996[37]	58F	headache, dizziness	lt middle cranial fossa/3×2	meningioma	R	-
15	Resnick 1996[20]	38M	visual loss, headaches	multiple lesions/-/orbital	-	SR	2 Y NC
16	Simos 1998[43]	62M	seizure, headaches, dysphasia, rt hemiparesis	lt parietal	meningioma	TR	24 M NR
17	Deodhare 1998[54]	41M	seizures	lt parieto-occipital	meningioma	SR	4 Y NR
18		38M	headaches	lt parieto-occipital double/each 4	meningioma	TR	6 M NR
19	Haung 1998[55]	38M	seizures	rt parietal/8.0×5.0×5.0	meningioma	TR	1 Y G
20	Udono 1999[28]	67F	diplopia, headaches	rt frontal/-/nasopharynx	meningioma	TR	20 M NR
21	Woodcock 1999[7]	15F	headaches, visual impairment, amenorrhea	suprasellar	pseudotumor	Bx	9 M Rec
22	Gaetani 2000[12]	67F	ataxia	rt cerebellar	meningioma	TR	1 M NR
23	Kattner 2000[56]	33M	seizures, headache	rt parasagittal	-	TR	1 Y NR
24	Natarajan 2000[13]	45F	seizures	rt frontal lobe	glioma	TR	5 M NR
25	Morandi 2000[17]	22F	diplopia, lt VI, VII palsies	fourth ventricle	-	TR	3 Y NR
26	Andriko 2001[21]	50M	headaches	lt petroclinoid	Meningioma in 7 cases One case no report	SR	17 M NP
27		22M	seizures	lt frontotemporal and rt parietal		SR	10 M G
28		63F	headaches	rt frontal		TR	-
29		25F	headaches	posterior third falx		TR	42 M NR
30		31F	seizures, lt arm numbness	multiple lesions		SR	5 days dead
31		62M	rt visual loss, papilledema	rt parasellar		SR	6 M NP
32		43F	headaches	rt parietal		TR	9 M NR
33		24M	seizures	lt occipital		TR	2 M NR
34	Petzold 2001[22]	47M	rt visual loss	multiple lesions	meningioma	SR+Rad	2 Y RE T
35	Wu 2001[29]	35M	headaches, seizures, memory problem	lt occipital/-/multiple foci total-body	-	SR	3 Y Rec
36	Juri 2003[14]	39M	vertigo, visual disturbances	rt temporal/4.5	meningioma	TR	10 M NR
37	Konishi 2003[47]	68F	seizures	lt frontal	-	Bx+ anti-epileptic	11 M NP
38	Chang 2003[30]	55M	nasal obstruction, epistaxis	bilateral cavernous sinuses, dura, cerebellar tentorial/-/nasal cavity	-	TR	3 M Rec
39	Dodson 2003[31]	56F	headaches, seizure	rt parietal/5×4×3cm/nasopharynx	meningioma	TR	6 M NR
40	Hadjipanayis 2003[32]	52M	headaches, malaise	lt petroclival/-/lymphadenopathy	meningioma	SR+Rad	15 M RE T
41	Ture 2003[39]	29M	nasal obstruction, visual impairment	anterior fossa	-	SR+Ste	2Y NR
42	Yetiser 2004[33]	7M	hearing loss	rt parietal/-/lymphadenopathy	-	Bx+Ste	NC
43		6M	hearing loss	occipital/-/lymphadenopathy	-	Bx+Ste	NC
44	Kinoshita 2004[45]	69M	seizures	lt frontal	pachymeningitis	SR	2 Y NR
45	Griffiths 2004[57]	9M	seizures, headaches	rt frontal/3.5×2.5	meningioma	TR	43 M NR
46	Kayali 2004[58]	31M	headaches	lt temporal	meningioma	TR	NR
47	Toh 2005[38]	60F	dizziness, vertigo	rt occipital and cerebellar hemisphere/3×6	-	SR	4 M NP
48		59F	seizures	lt frontal	-	SR	30 M G
49	Gies 2005[59]	40F	headaches	lt parietal	-	TR	10 M NR
50	Kaminsky 2005[34]	32M	nasal obstruction, trigeminal pain	petroclival region and cavernous sinus/-/paranasal sinus and rt C5-6	meningiomatoses	TR	-
51	Gupta DK 2006[26]	15M	headaches, visual deterioration	bilateral petroclival and cavernous masses	meningioma	SR	12 M RE T
52	Z'Graggen 2006[46]	35M	lt headaches	lt cerebral convexity	-	Bx+Ste	3 M regredience
53	McPhlerson 2006[23]	53M	headaches, visual loss	multiple lesions	meningioma	SR+Ste	11 M G
54	Seyednejad 2007[41]	43F	quadriplegia and leg paresthesias	anterior-to-posterior cranial fossa	-	R+Rad	5 Y Rec at spine
55	Ghosal 2007[24]	26M	seizures	multi/largest 2.7×1.4	meningioma	TR	1 Y NR
56	Di Rocco 2007[48]	13F	lt headache	lt frontal	-	SR	3 M Rec
57	Miletic 2008[16]	8-	symptoms of increased intracranial pressure	lt insular/2.5	-	Bx+Ste+Rad	3 Y disappear
58	Wan 2008[8]	43M	lt visual blurring, headache	suprasellar/3×3×2.5	meningioma	TR	3 M NR

59	Scumpia 2008[49]	22M	headaches, visual deterioration	rt middle fossa /3.7×2.6×3.7+4.2×2×2.3	meningioma	SR	-
60	Theeler 2008[60]	56F	lt arm and leg motor impairment	rt frontoparietal	meningioma	TR	12 M NR
61	Raslan 2008[25]	50M	lt diplopia, numbness	Lt frontal and both temporal/-/L1	meningioma	TR	26 M Rec at spine
62	Hinduja 2008[35]	42M	visual loss	anterior cranial fossa/3×1.8/intraorbital	meningioma	SR+Ste+Rad	1.5Y RE T
63	Majdoub 2009[15]	10F	headache, dizziness	lt basal ganglia/2.5	-	TR+Rad	49 M NR
64	Russo 2009[3]	71M	upper rt arm paresis	lt frontal/2×3	-	TR	12 M NR
65		72M	headaches, mild hemiparesis	bilateral frontal/3×3	lymphoproliferative disease	TR	14 M NR
66	Lungren 2009[61]	2F	bilateral papilledema	frontal	-	R	-
67	Wang 2010[9]	10F	polydipsia, polyuria, hypopituitarism	sellar	-	TR+Rad	5 M Rec
68		27M	polydipsia, polyuria, headaches, visual impairment	sellar	-	TR+Ste	43 M Rec
69	Lu 2010[62]	48F	no	lt frontal	meningioma	TR	11 M NR
70	Zhang 2010[10]	27M	headaches, binocular visual impairment	sellar	meningioma	TR	4 Y NR
71		38F	headaches	pituitary	-	R	2 Y G
72		26F	headaches	pituitary	meningitis	TR	3 Y NR
73		30M	headaches, rt nasal obstruction, epistaxis	anterior cranial fossa/-/rt maxillary sinus, rt nasal cavity, orbit	-	R	-
74	Waild 2010[44]	60F	bipolar disease, paranoia	rt frontotemporal/7.4×5.1	meningioma	SR+Rad	-
75	Cavuoto 2011[36]	25M	headache, seizure, vertigo	lt temporal lobe and frontal gyrus/-/lt and rt eye	-	Bx+Ste+Rad	-
76	Raslan 2011[11]	50M	headache, diplopia,	multiple lesions in skull base and convexity/-/spinal meningeal	-	R	-
77		54M	headache, rt deceased vision, rt decreased hearing, dysphagia	sellar and suprasellar lesions bilateral CPA lesions/-/epidural mass in cervical canal	-	R+Ste	-
78		50F	galactorrhea, headaches	pituitary/-/anterior mediastinum mass	-	R	-
79	Present case 2011	67F	dizziness	lt frontal/1.5×1.3×1.0	meningioma	TR	7 M NR

*1 -: not described
*2 TR: total resection, SR: subtotal resection, R: resection, not specified, Bx: biopsy, Ste: steroid therapy, Rad: radiotherapy, -: not described
*3 NR: no recurrence, Rec: recurrence, NP: no progression, Y: year, M: month, RE T: residual tumor, RS: residual symptom, NC: no change, G: good health, -: not described
Abbreviations: lt, left; rt, right;

Discussion

Since RDD was first reported by Rosai and Dorfman in 1969,[1] more than 750 cases have been reported.[11] RDD generally affects persons 20 years of age or younger (mean age 20.6) and shows a male predominance.[2,4] About 90% of cases have painless cervical massive lymphadenopathy. Extranodal RDD has been reported in approximately 43% of cases.[2] Intracranial RDD without lymphadenopathy or other organ involvement is extremely rare (0.5% of cases).[4] The male predominance for intracranial RDD is the same, but the mean age is higher (2 to 78 years, mean age 38.9).

Intracranial Rosai-Dorfman disease mainly develops in dural regions. On radiographical findings, the lesions appear to be dural-based, extra-axial and enhanced with surrounding vasogenic edema.[49] Therefore, the preoperative diagnosis for most cases is meningioma, other cerebral tumors or lymphoproliferative lesion. Of those with intracranial RDD, 7.6% of cases have lymphadenopathy and 17.7% show other systemic lesions. More than 70% of RDD cases have presented with only intracranial lesions.

The clinical manifestations of intracranial RDD are due to increased intracranial pressure. Most cases complain of headaches, followed by seizure and visual and hearing impairments. Those at sellar and pituitary areas may cause polydipsia, polyuria, and galactorrhea.

Histology and immunostaining are essential to reach a correct diagnosis of RDD.[10] Intracranial RDD needs more careful observation, since patients are reported to show more fibrosis, fewer typical histiocytes, and less emperipolesis than nodal RDD.[2]

Diagnosis of intracranial RDD can be difficult to differentiate from lymphoplasmacyte-rich meningioma, plasma cell granuloma, Langerhans cell histiocytosis, and plasmacytoma.[26,45,50] Lymphoplasmacyte-rich meningioma shows infiltrating meningeal cells immunoreactive for EMA,[50] which can distinguish it from RDD, in which structural cells do not express EMA. It may be difficult to differentiate

RDD from plasma cell granuloma.[24] Microscopic features of plasma cell granuloma reveal histiocytes that are negative for S-100 protein and do not show emperipolesis.[50] On the other hand, the histiocytes in RDD are immunopositive for S-100 protein and emperipolesis is present, differentiating it from plasma cell granuloma. Although S-100 protein-positive histiocytes showed variable reactivity, the intensity of S-100 protein is reported to be variable by the size of the histiocytes.[52] Langerhans cell histiocytosis is positive for CD1a, but the histiocytes in RDD are negative for CD1a.[50] Plasmacytoma shows a cytologically monotonous and monoclonal proliferation of plasma cells.[50] The plasma cells of our cases were polyclonal and immunopositive for kappa or lambda light chain.

Intracranial RDD mimics meningioma or other intracranial tumors radiologically and clinically.[49] Therefore, the first choice for treatment is surgical resection. Surgical treatment contributes not only to reducing increased intracranial pressure but also to confirming the diagnosis.[48] Surgical resection was very effective in many cases. After subtotal resection, some cases respond with tumor decreases following radiotherapy and steroid therapy, but other cases showed tumor recurrence in the same region and in other regions such as the spine. The prognosis of intracranial RDD is favorable.[26] Most patients have no recurrence after surgery, however, 14.3% of cases showed recurrence within the follow-up period of 3 months to 5 years. Therefore, intracranial RDD needs further study to facilitate discovery of additional treatment options. Our case has shown no signs of recurrence 7 months after surgery.

Intracranial RDD is very rare and the clinical course and treatment of this disease are therefore not well established. The clinical manifestations and prognosis are variable depending on the location and treatment. Most intracranial RDD are benign diseases, but long-term follow up is important.

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