Surgical Management of Skull Base Rosai–Dorfman Disease

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Key words
- Rosai–Dorfman disease
- Sellar/parasellar tumor
- Skull base tumor
- Vision loss

Abbreviations and Acronyms
MRI: Magnetic resonance imaging

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■ BACKGROUND AND IMPORTANCE: **Rosai–Dorfman disease is a rare benign histiocytic proliferative disorder with a self-limiting clinical course. Skull base Rosai–Dorfman disease presents with intracranial lesions that often mimic meningiomas and other benign skull base tumors. The disease is difficult to diagnose radiographically, and tissue diagnosis exposes patients to significant perioperative risk. Surgical resection may require a large skull base exposure that risks significant surgical morbidity. Aggressive surgical resection, although often attempted, is of unproven efficacy. Our objective was to determine the optimal surgical management of skull base Rosai–Dorfman disease.**

■ CASE DESCRIPTION: **We present 2 cases of skull base Rosai–Dorfman disease: a 26-year-old man with a middle fossa tumor and a 15-year-old teenage girl with a hypothalamic tumor. In addition, we reviewed 39 cases of skull base Rosai–Dorfman disease reported in the literature.**

■ CONCLUSIONS: **Tumors commonly occur in the sellar/parasellar region and result in loss of vision. Regardless of extent of resection, the majority of patients (>78%) have subsequent tumor regression or stable disease. Steroids and/or radiation are effective treatments for tumor recurrence. Tumor biopsy followed by observation, steroids, and/or radiation may be the most appropriate surgical management of skull base Rosai–Dorfman disease.**

INTRODUCTION

Rosai and Dorfman first described sinus histiocytosis with massive lymphadenopathy in 1969.[1] It initially was described as a benign histiocytic proliferative disorder that presented with massive painless cervical lymphadenopathy, fever, weight loss, anemia, and polyclonal hypergammaglobulinemia. Biopsy of affected tissue is required to make a definitive diagnosis. Clinical observation without treatment is advisable when possible, because the disease generally is nonprogressive and self-limiting.[2] Surgical resection may be needed to control focal symptomatic disease.

With only a little more than 100 cases reported in the literature, central nervous system Rosai–Dorfman disease likely is an underrecognized diagnosis.[3] These lesions tend to occur in the parasagittal and skull base region and often mimic common benign dural-based lesions, particularly meningiomas. Although the majority of patients undergo surgical resection, the optimal surgical management and long-term natural history of intracranial Rosai–Dorfman disease has yet to be determined.

Skull base Rosai–Dorfman disease presents a clinical challenge for 2 main reasons. First, these benign lesions are prone to causing debilitating neurologic symptoms. Cranial nerve palsies,[4,5] endocrine dysfunction,[6,7] and vision loss attributable to optic apparatus compression[8,9] are common presenting symptoms. Second, aggressive resection often requires a large skull base approach because the location of the lesion. Standard surgical approaches to the skull base, such as subcranial,[8] orbitozygomatic,[10] or retrosigmoid[11,12] craniotomies can result in significant perioperative morbidity. Patients are exposed to a high degree of perioperative risk in an attempt at tumor resection for a disease that is nonprogressive and self-limiting.[2,13,14] Although needle or burr-hole biopsy techniques may not be feasible for some skull base locations (e.g., the infratemporal fossa), tissue biopsy without aggressive resection may be the most appropriate surgical management of skull base Rosai–Dorfman disease.

To investigate this claim, we present 2 cases of skull base Rosai–Dorfman disease and reviewed 39 cases from the literature. The clinical presentation, treatment, and outcome of this rare intracranial disease were recorded. Our objective was to determine the optimal surgical management of skull-base Rosai–Dorfman disease.

CASE STUDIES

Case 1

A 26-year-old man presented with sudden onset of dizziness, nausea, and visual scotoma described as flashing lights that resolved spontaneously after 30 minutes. The patient was neurologically intact at time of evaluation. Computed tomography of the head demonstrated a middle fossa osteolytic mass near the temporal pole involving the pterion and sphenoid wing. Magnetic resonance imaging (MRI) of the brain showed a heterogeneously contrast-enhancing mass of the middle fossa with extracranial extension into the subtemporal fossa (Figure 1). Differential diagnosis included atypical meningioma, hemangiopericytoma, eosinophilic granuloma, and sarcoma.

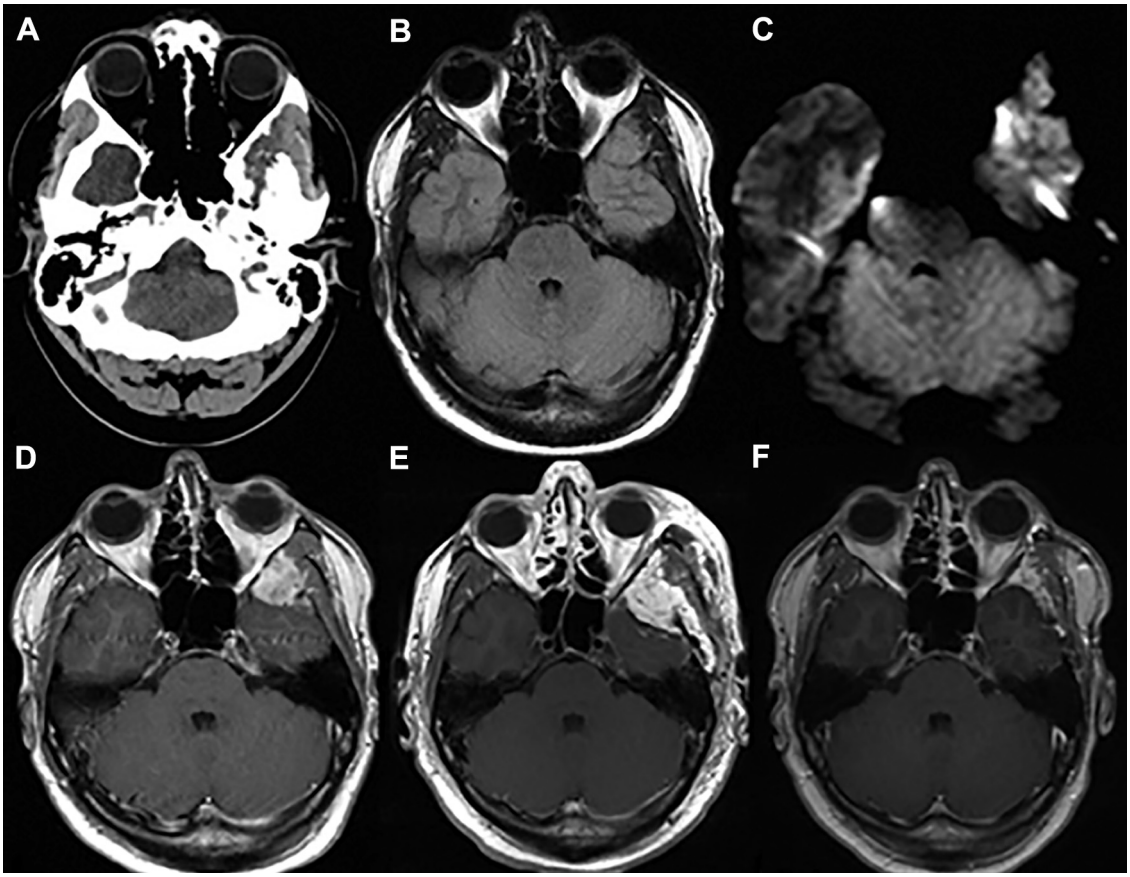

Figure 1. Axial images for case 1. (**A**) Computed tomography of the head shows osteolytic lesions involving the middle cranial fossa with extension into the infratemporal fossa. (**B**) Fluid attenuation inversion recovery sequence demonstrates isointense lesion at the anterior pole of the middle fossa. (**C**) Diffusion-weighted magnetic resonance imaging (MRI) notable for subtle hyperintensity compared with contralateral temporal lobe. (**D**) Preoperative post-gadolinium T1-weighted MRI shows avidly enhancing mass. (**E**) Immediate postoperative post-gadolinium T1-weighted MRI shows fat graft without clear evidence of residual tumor. (**F**) Two-year postresection MRI shows no evidence of tumor recurrence.

Pterional craniotomy with zygomatic osteotomy was completed to access both the middle temporal and infratemporal fossa. Tumor was noted to be within the dural leaflets and did not resemble meningioma. Intraoperative frozen section demonstrated benign dense fibrous tissue and chronic inflammation with frequent giant cells, favoring histiocytic lesion. The surgeon elected to perform biopsy only to minimize morbidity, given the ambiguity of the intraoperative pathologic findings. The patient experienced the expected postoperative trismus, but had an otherwise uncomplicated course. He was discharged to home on postoperative day 2.

Pathology demonstrated a mixed inflammatory infiltrate composed of macrophages and lymphocytes. Some macrophages were multinucleated and demonstrated emperipolesis (i.e., lymphocytophagocytosis). Macrophages were immunoreactive for S-100 protein staining but not CD1a. Lymphocytes were a mixture of T and B cells. Special stains for bacteria, fungus, and acid-fast bacilli were negative. Pathologic findings were definitive for Rosai–Dorfman disease.

At the patient's routine outpatient follow-up visit, postoperative paresis of the frontalis branch of the facial nerve was noted due to retraction injury. This subsequently resolved. Outpatient work-up was negative for systemic disease on position emission tomography scan. The patient's case was presented at our multidisciplinary brain tumor board and our radiation oncologists suggested that he be treated with low-dose radiotherapy to prevent recurrence. The patient elected to forgo radiation and was followed with serial surveillance MRIs. MRI follow-up at 2 years demonstrated no evidence of residual or recurrent disease. The patient had complete resolution of his symptoms.

Case 2

A previously healthy 15-year-old girl presented with a 6-month history of polydipsia, polyuria, and 50-pound weight gain. Her primary care physician referred her to an endocrinologist, who diagnosed her with hypothyroidism and prediabetes mellitus. She subsequently developed altered mental status and generalized weakness. She later presented to the emergency department dehydrated, tachycardic, and hypernatremic, with a sodium level of 155 mg/dL. MRI of the

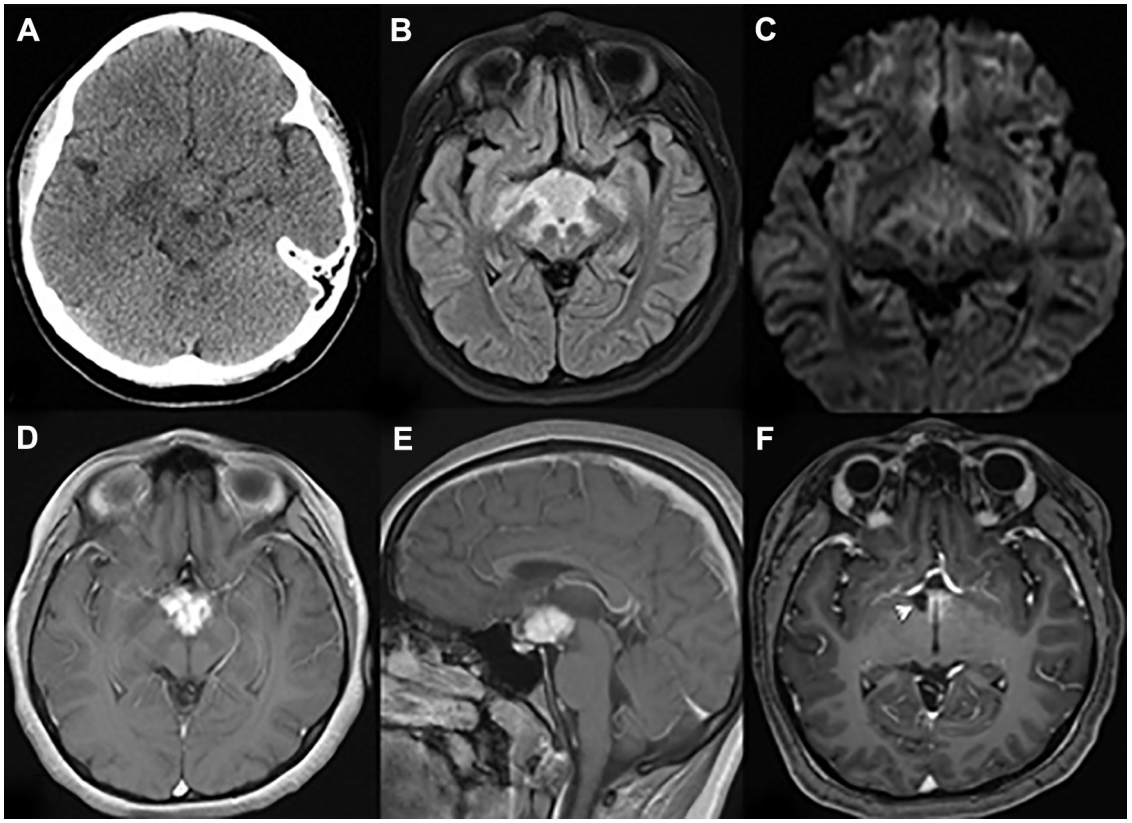

Figure 2. Imaging for Case 2. (**A**) Computed tomography of the head shows mild fullness in the suprasellar/hypothalamic region. (**B**) Brain fluid attenuation inversion recovery magnetic resonance imaging (MRI) sequence demonstrates dramatic hyperintensity involving the midbrain, visual apparatus, and hypothalamus. (**C**) Diffusion-weighted MRI shows subtle hyperintensity in similar regions. (**D**) Axial and (**E**) sagittal preoperative postgadolinium T1-weighted MRIs show avidly enhancing mass in the hypothalamic region. (**F**) Postgadolinium T1-weighted MRI shows regression of enhancing mass afterg radiation. Biopsy cavity can be seen lateral to mass (*arrowhead*).

brain demonstrated a 2.4 cm × 2.2-cm suprasellar/hypothalamic mass (Figure 2). Differential diagnosis included pilocytic astrocytoma or other low-grade glioma, germ cell tumor, or lymphoma. Endocrinologic evaluation demonstrated panhypopituitarism (H = high, N = Normal, L = Low): prolactin 44 ng/mL (H), morning cortisol 1.0 µg/dL (N), adrenocorticotropic hormone 12 pg/mL (N), insulin-like growth factor-1 80 ng/mL (L), follicle-stimulating hormone 0.7 (L), luteinizing hormone 0.4 (L), thyroid-stimulating hormone 0.14 (L), free T4 1.14 ng/dL (L), estradiol less than 20 (L). She was admitted to the pediatric intensive care unit for close monitoring and further work-up. Findings of the neuro-ophthalmology examination did not reveal any deficits.

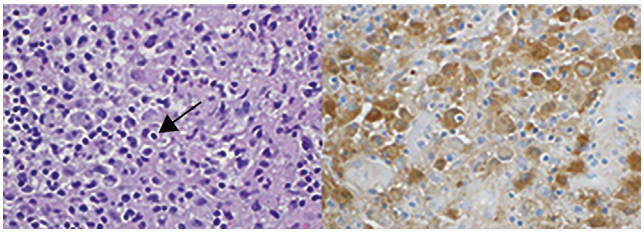

Figure 3. Pathology for Case 2. *Left*: Hematoxylin and eosin stain showing mixed inflammatory infiltrate composed predominately of macrophages and lymphocytes. *Arrow* shows a mononuclear cell engulfed by a macrophage (emperipolesis). *Right*: S100 protein stain highlights macrophages and demonstrates the clear halo surrounding mononuclear cells engulfed within macrophages (emperipolesis).

Pediatric neurosurgery was consulted, and recommended serum beta-human chorionic gonadotropin, placental alkaline phosphatase, and alpha-fetoprotein to evaluate for possible germ cell tumor. All levels were within normal limits. Cerebrospinal fluid specimen was collected via lumbar puncture with the following test results: alpha-fetoprotein less than 0.5 ng/mL (N) and beta-human chorionic gonadotropin less than 0.5 IU/L (N). Cerebrospinal fluid cytology revealed small mature lymphocytes with no evidence of malignant cells. Full spinal MRI showed

Table 1. Summary of Skull-Base Rosai–Dorfman Disease Reported in the Literature

Authors and year	Age, years	Sex	Signs and Symptoms	Location	Nodal	Initial Treatment	Follow-Up and Clinical Course
Andriko et al., 2001[15]	50	M	Headaches	Petroclinoid region	None	Subtotal resection	17 months, residual stable tumor near carotid
	62	M	Rapid vision loss, bilateral papilledema	Parasellar	N/A	Subtotal resection	6 months, stable residual disease with worsening vision
Bhattacharjee et al., 1992[16]	78	M	Vision loss	Suprasellar	None	Subtotal resection	1 year, stable
Chivukula et al., 2014[4]	66	F	Headaches, diplopia with bilateral 6th nerve palsies	Suprasellar/ hypothalamic	None	Stereotactic brain biopsy followed by radiation, steroids	19 months, stable
Douleh et al., 2015[17]	39	F	Hearing loss, otitis externa	Middle fossa, EAC/IAC	None	Biopsy, radiation	6 years, tumor regression, no recurrence
Foucar et al., 1982[11]	21	M	Facial palsy, deafness	CPA	None	N/A	N/A
Friedman et al., 1984[18]	32	M	Headache	Suprasellar	LA	Subtotal resection	14 months, stable
Gaetani et al., 2000[19]	67	F	Ataxia	Cerebellum/ posterior fossa	None	Gross total resection	1 month, complete resolution
Gupta et al., 2006[10]	15	M	Visual loss	Bilateral petroclival	None	Orbitozygomatic craniotomy	12 months, improvement in vision with residual tumor
Gupta et al., 2011[20]	14	M	Vision loss, hypopituitarism	Suprasellar, cavernous sinus	None	Pterional craniotomy with orbital osteotomy, subtotal resection	N/A
Hadjipanayis et al., 2003[21]	52	M	Facial paresthesias	Petroclinoid region, cavernous sinus	LA	Petrosal/translabyrinthine craniotomy, stereotactic radiosurgery	2 months, regression
Kaminsky et al., 2005[22]	32	M	Trigeminal pain, epistaxis	CPA, cavernous sinus, suprasellar	None	Transnasal biopsy, frontal craniotomy	N/A
Katz et al., 1993[23]	17	M	Headache	Posterior fossa	None	Subtotal resection	8 years, no recurrence
Kelly et al., 1999[24]	45	F	Headaches, fever, hyponatremia, polydipsia	Sellar	None	Gross total resection	3 years, no recurrence
Kidd et al., 2006[25]	37	F	Headaches, visual loss	Parasellar, skull base	None	Frontal craniotomy, subtotal resection	4 years, recurrence, resolved with craniospinal radiation
	68	M	Visual loss	Sellar/suprasellar	None	Craniotomy, subtotal resection, WBRT	Tumor regression, improvement in visual fields
Lopez and Estez, 1989[27]	35	M	Facial pain	CPA, cavernous sinus	None	Subtotal resection, steroids	12 months, stable
McPherson et al., 2006[28]	53	M	Vision loss	Suprasellar, clivus, CPA	None	Orbitozygomatic craniotomy	10 months, recurrence that resolved with prednisone
Mir et al., 1985[29]	13	M	Headache	Suprasellar	LA	Subtotal resection	N/A
Morandi et al., 2000[30]	22	F	Diplopia, 6th and 7th nerve palsy	Floor of 4th ventricle	None	Posterior fossa craniotomy	Improvement of diplopia
Nalini et al., 2012[31]	35	M	Visual loss, hearing loss	Parasellar, tentorium, clivus	None	Biopsy, radiation	N/A
Ng & Poon, 1995[32]	22	M	Diabetes insipidus	Sellar	None	Needle biopsy	N/A
Panicker et al., 1996[33]	58	F	Headache, dizziness	Middle fossa	None	Gross total resection	N/A

Petzold et al., 2001[8]	47	M	Right visual loss	Suprasellar, CPA	None	Subtotal resection of suprasellar mass	1 year, recurrence with worsening visual deficits, radiation after recurrence
Resnick et al., 1996[34]	38	M	Deafness, proptosis	CPA, orbit	None	Subtotal resection x2	2 years, stable
Sakai et al., 1998[16]	60	M	Tinnitus	CPA	LA	Multiple resections	31 months, recurrence treated with resection
Scumpia et al., 2009[9]	22	M	Visual loss	Middle cranial fossa, orbit	None	Pterional craniotomy	Planned staged resection of orbital portion
Sharma et al., 2005[35]	40	M	Seizure	Sphenoid wing	None	Pterional craniotomy, subtotal resection	12 weeks, seizure-free with no residual disease
Shaver et al., 1993[5]	5	M	Multiple cranial nerve palsies	Cavernous sinus	None	Subtotal resection	6 months, no recurrence
Song et al., 1989[36]	30	M	Seizure	Skull base	None	Gross total resection	N/A
Sundaram et al., 2005[37]	56	M	Headache, weakness	Sphenoid wing	None	Frontal craniotomy, subtotal resection	11.5 years, no recurrence
Trudel, 1984[38]	28	M	Facial pain, deafness	Middle fossa	None	Subtotal resection	14 months, stable
Wang et al., 2011[6]	10	F	Hypopituitarism, polydipsia, polyuria	Sellar	None	Transsphenoidal resection, radiation	5 months, local recurrence treated with steroids, resolution
	27	M	Vision loss, polydipsia, polyuria	Sellar	none	Subfrontal craniotomy	3 years, recurrence treated with radiation, resolution at 5 years
Woodcock et al., 1999[7]	15	F	Hypothyroidism, primary amenorrhea	Suprasellar/ hypothalamic	none	Subfrontal craniotomy for open biopsy	9 months, mild enlargement
Zhang et al., 2010[39]	26	F	Headache, CSF pleocytosis	Sellar/suprasellar	none	Transsphenoidal resection	3 years, no recurrence
	27	M	Headache, visual loss	Sellar/suprasellar	none	Surgery	4 years, no recurrence
	30	M	Headaches, nasal obstruction	Anterior skull base, orbits	none	Surgery	N/A
	38	F	Headache, nausea, vomiting	Sellar/suprasellar	none	Transsphenoidal resection	2 years, no recurrence

M, male; N/A, not available; F, female; EAC, external auditory canal; IAC, internal auditory canal; CPA, cerebellopontine angle; LA, lymphadenopathy; WBRT, whole-brain radiotherapy; CSF, cerebrospinal fluid.

no evidence of spinal leptomeningeal disease or drop metastasis.

A right frontal stereotactic needle biopsy was completed for tissue diagnosis. The patient tolerated the procedure well without complications. Pathology was consistent with diagnosis of Rosai–Dorfman disease (Figure 3). Because of her continued symptoms related to panhypopituitarism, treatment with focal external beam radiation therapy with total dose of 36 Gy was given for local disease control. At her 6-month follow-up visit, the patient had stable disease radiographically and continues to require total hormone replacement.

REPORTS IN THE LITERATURE

A PubMed search was completed for *intracranial Rosai–Dorfman disease* with English filter. The search resulted in 115 articles that were then screened for skull base location. Any references used within these articles that included cases of skull base Rosai–Dorfman disease also were included. A total of 39 cases of skull base Rosai–Dorfman disease were found in the literature since 1982.[4–9,11,12,15–39] A summary of each case can be found in Table 1.

Mean age at presentation was 36.7 years (range, 5–78 years). Mean follow-up time was 2.3 years (range, 0.2–11.5). Seven pediatric patients (<18 years) have been reported. Men were affected more commonly than women (72% vs. 28%, respectively). The most common presenting symptoms were headache and cranial nerve deficits. Summary of presenting symptoms is listed in Table 2. Tumor location was most common in the sellar region (Table 3). Nodal involvement (systemic lymphadenopathy, usually in the cervical region) was uncommon, present in only 4 patients.

Treatment, Recurrence, and Outcome

Treatment and follow-up information was available for 32 patients, including the 2 cases presented here. Skull-base craniotomy for tumor resection was performed in 25 of 32 patients (18 subtotal resection, 5 unknown, 2 gross total resection). Transsphenoidal surgery and biopsy were performed in 3 and 4 patients, respectively. Initial treatment included radiation in 5 patients (3 focal, 1 whole-brain radiation therapy, 1 stereotactic radiosurgery) and steroids in 2 patients.

Twenty five patients (78%) had tumor regression or stable disease after the initial surgical intervention. Seven patients (3 subtotal resection, 2 unknown, 1 transsphenoidal, 1 biopsy) had progression of disease or recurrence during their clinical course. Of the 5 patients who received radiation, 1 had recurrence. Recurrence treatment consisted of radiation (3 patients), steroids (2 patients), re-resection (1 patient), or observation after mild asymptomatic enlargement (1 patient). All patients who underwent steroid or radiation therapy for recurrence had resolution of their tumors and improvement of symptoms. No adverse event from treatment for recurrence was noted.

DISCUSSION

We report 2 cases of skull-base Rosai–Dorfman disease. Both patients had stable or self-limiting disease after treatment. These cases serve as illustrative examples of the presentation, radiographic features, surgical management, and clinical course of this rare benign skull-base tumor. A total of 39 cases of skull base Rosai–Dorfman disease were reviewed, showing that 78% of tumors remain stable or regress after initial surgical resection, regardless of extent of resection.

Intracranial and skull base tumors constitute 5%–10% of all cases of Rosai–Dorfman disease diagnosed each year.[3,15,40] Men are affected more commonly, at a mean age of approximately 40 years. Tumors tend to be dural-based, involving the parasagittal region and skull base. Presenting symptoms vary with tumor location. Endocrine dysfunction, especially diabetes insipidus, is a common presenting symptom for sellar/suprasellar tumors and may preferentially affect children and young adults. Wang et al.[6] reported a 10-year-old and 22-year-old patient with sellar Rosai–Dorfman disease who both presented with endocrine dysfunction. Of the 7 patients with endocrine dysfunction in our literature review, all required prolonged hormone therapy despite regression of tumor.

The common radiographic finding is an enhancing dural-based mass on T1-weighted postgadolinium MRI. Enhancing tumor that is most easily accessible should be the target for tissue diagnosis. The literature has underemphasized some radiographic features that distinguish these tumors from meningiomas. Multiple reports have found these lesions to have some distinguishing radiographic characteristics: hyperintense on T1-weighted and fluid attenuation inversion recovery sequences,[41] hypointense on T2-weighted imaging,[41,42] and mild diffusion restriction on diffusion-weighted imaging. Intracranial Rosai–Dorfman disease may have multiple discrete lesions[10,26,28] or have an en plaque appearance.[28,43] Although the listed findings are not universally present, these imaging characteristics may help to differentiate skull base Rosai–Dorfman disease from a standard meningioma. Clinical suspicion should be particularly high with concurrent lymphadenopathy.

In our experience, the intraoperative findings on gross pathology differ from

Table 2. Summary of Presenting Symptoms

Signs and Symptoms	No. Patients (N = 39)
Cranial nerve (CN) deficits	24
CN II	11
CN V	4
CN VI	2
CN VII	2
CN VIII	5
Headache	9
Hypothalamic/pituitary dysfunction	6
Seizures	2

Table 3. Summary of Tumor Locations

Location	No. Patients (N = 39)
Sellar region	20
Suprasellar	12
Sellar	5
Parasellar	3
Middle fossa	7
Cavernous sinus	5
Cerebellopontine angle	4
Posterior fossa	3
Petroclival	2

meningioma. Tumor does not directly involve the dura and lacks a dural tail, a classic finding in meningioma. It has been described as "pink soft mass" in other reports.[17] Frozen specimen shows fibrous tissue matrix with histiocyte predominance, allowing differentiation from neoplastic lesion. Preoperative imagining, intraoperative findings, and frozen pathology provide sufficient clinical data to make appropriate diagnosis of Rosai–Dorfman disease.

Vision loss caused by involvement of the optic apparatus is a common and debilitating feature of skull-base Rosai–Dorfman disease. Significant visual symptoms were reported even with small tumors and recurrences. One patient had stable residual disease on MRI but continued to have worsening vision.[15] Petzold et al.[8] report 2 cases of skull base Rosai–Dorfman disease affecting the suprasellar region causing vision loss. Both patients underwent bifrontal/transglabellar craniotomy for resection of suspected meningioma. Subtotal resection was completed with residual tumor near the optic apparatus. At their 1-year follow-up visit, both patients had progression of disease with significant worsening of vision. Low-dose focal radiation was then provided with partial recovery of vision and resolution of tumor. The authors suggest postoperative radiation be considered for all patients as prophylaxis against disease recurrence and progressive vision decline. Whether the degree of vision loss is proportionally greater than other benign skull base lesions has not been determined; we suspect that the inflammatory nature of these lesions may result in injury to the optic apparatus. Postoperative radiation and/or steroids may be warranted in these cases, particularly in the setting of residual/recurrent tumor or progressive symptoms.

Management of intracranial Rosai–Dorfman disease has consisted of surgical resection with or without postoperative radiation and steroids.[3] Large skull base craniotomies are usually required to safely expose these tumors. The perioperative complication rate for open skull base surgery ranges up to 40%.[13,44] These complication rates vary depending on tumor location and aggressiveness of resection, both of which are directly relevant to the perioperative morbidity associated with tumor resection of Rosai–Dorfman disease. Previous authors have stated that the morbidity associated with gross total resection is too high, considering the uncertain therapeutic benefit.[21,43]

Tomio et al.[43] reported on the complications associated with aggressive resection of intracranial Rosai–Dorfman disease. A 53-year-old man with right parietal convexity Rosai–Dorfman disease suffered left hemiparesis and intracranial hemorrhage following craniotomy for attempted gross total resection. The complications were attributed to tumor hypervascularity, brain invasion, and overaggressive resection. Although symptoms related to mass effect generally improve with resection, it is unclear if aggressive surgical intervention alters natural history and improves patient prognosis.

The majority of patients with skull base Rosai–Dorfman disease reported in the literature had stable or regression of disease (78%) after initial surgical treatment. Andriko et al.[15] reported the largest case series of central nervous system Rosai–Dorfman disease completed to date. Nine patients with surgically treated intracranial or spinal disease had follow-up data. No patient had radiographic evidence of disease progression or recurrence. The majorityof patients (56%) had subtotal resection and were alive with disease. Gross total resection did not provide additional therapeutic benefit. These findings lend further evidence that gross total resection is unlikely improve patient outcome and may increase complication rates.

In conclusion, skull base Rosai–Dorfman disease most commonly remains stable or regresses following initial surgical management, regardless of extent of resection. Although tumor debulking is indicated in the setting of progressive vision loss due to optic apparatus compression, aggressive gross total resection may expose patients to unnecessary perioperative risk with lack of therapeutic benefit. Open or stereotactic tumor biopsy followed by observation, steroids, or radiation (initially or at recurrence) may be the optimal surgical management of skull base Rosai–Dorfman disease.

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