Extensive Multifocal Rosai-Dorfman Disease Involving the Central Nervous System and Paranasal Sinuses

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Abstract

Introduction: Rosai-Dorfman disease (RDD) is a rare benign lymphocytic disorder characterized by idiopathic histiocytic proliferation affecting lymph nodes. RDD in the central nervous system (CNS) without nodal disease is exceedingly uncommon. We describe a rare case of extensive multifocal RDD involving the CNS, skull base, and paranasal sinuses. We review the literature and propose a surgical strategy to manage extensive multifocal disease.

Methods: This 39-year-old male with known RDD and multiple previous surgeries for intracranial and spinal RDD lesions presented with worsening headache, gait disturbance, and left sided weakness. Neuroimaging demonstrated multifocal enhancing dural-based lesions in the brain including both cerebellopontine angle, left clinoidal, left petroclival and bilateral Meckel's cave lesions (Figures 1A & B). The patient also had obstructive sleep apnea due to RDD occupying the entire nasal and paranasal sinuses.

Results: The patient underwent a left-sided retrosigmoid-transpetrosal-transtentorial approach to remove the left petroclival tumor and decompress the brainstem. The mass was carefully dissected off of cranial nerves IV through XI with excellent decompression of the brainstem. Endoscopic resection of the paranasal sinus masses throughout the paranasal sinuses, multiple extra-axial enhancing masses at the skull bases, and worsening spasticity and gait dysfunction for the past 2 years. Previous treatments include intravenous hypergammaglobulinemia, and elevated erythrocyte sedimentation rate (2). Extensive multifocal RDD has been reported in up to 43% of cases with or without lymphadenopathy in diverse sites, most commonly the skin, bone, and upper respiratory tract (2). Management of RDD involving the brain and space without lymphadenopathy is exceedingly rare (2). In 1996, Foscar, Rosai, and Dorfman developed a S100 region, describing 423 RDD patients. Twenty-five of 423 patients had RDD in intracranial or intraspinal structures. Only 10 out of those patients had no lymph node involvement (2).

Conclusions: Extensive multifocal RDD involving the brain, spine, and paranasal sinuses is exceedingly rare and often mimics meningiomas on imaging. Surgical management can be challenging. We recommend a conservative approach with close observation and reserve surgical treatment for enlarging symptomatic lesions.

Discussion

Sinus histiocytosis with massive lymphadenopathy (SHML), also known as Rosai-Dorfman-Disease (RDD), was first described in 1966 by Rosai and Dorfman. It is an idiopathic histiocytic proliferative disorder affecting the lymph nodes that is currently defined by its histologic features. Classically, RDD presents as massive and painless cervical lymphadenopathy in young adults (mean age 20 years). Initial clinical presentation can be associated with fever, anemia, hypergammaglobulinemia, and elevated erythrocyte sedimentation rate (2). Extensive disease has been reported in up to 43% of cases with or without lymphadenopathy in diverse sites, most commonly the skin, bone, and upper respiratory tract (2).

 RDD involving the brain and space without lymphadenopathy is exceedingly rare (2). In 1996, Foscar, Rosai, and Dorfman developed a S100 region, describing 423 RDD patients. Twenty-five of 423 patients had RDD in intracranial or intraspinal or intracranial or intraspinal structures. Only 10 out of those patients had no lymph node involvement (2).

RDD in the CNS most commonly occurs in adults in the fourth decade (5). Initial clinical presentation can include headaches, seizures, numbness, vision loss, symptoms of raised intracranial pressure, or paraplegia (3, 4) and the differential diagnosis is broad. The differential diagnosis includes a wide variety of disease including metastatic cancers, a variety of infectious disease, and sinus hypophysitis (2). Most commonly, RDD intracranial manifestations are extra-axial and dural-based masses that mimic meningiomas on imaging (7, 4, 6, 7). Distinguishing RDD from meningomas on imaging is subtle; RDD has low signal intensity on T2-weighted images hypothesized to be due to free radicals released by inflammatory macrophages (9).

For the management of Rosai-Dorfman disease includes radiation therapy, chemotherapy, corticosteroids, and surgery. Clinical observation without treatment is preferred when possible (10) because RDD is considered a benign disease. There has been no reported evidence of malignant transformation of RDD and many patients continue to live long lives after diagnosis (11). However, the clinical course of RDD is variable and can include regression, persistence, or progression. Although RDD is considered a benign disease, if a patient has an aggressive disease course, it may cause clinical problems due to prominent mass formation. The disorder can have a propensity to form large masses and disseminate to both nodal and extranodal sites (2). If a surgical approach is taken, biopsy allows for establishment of the definitive diagnosis. Features suggesting the diagnosis of RDD include large nuclear size, nuclear prominence, acidophilic cytoplasm, and uniform S-100 staining (2). On histology, the pathognomonic finding is well preserved lymphocytes contained in the cytoplasm of histiocytes, a phenomenon known as emperipolisis (2).

RDD in the central nervous system (CNS) without nodal disease is exceedingly rare (3, 4). Of the 423 patients reported in the sinus histiocytosis with massive lymphadenopathy registry, 20 cases had evidence of CNS involvement and only 10 of those had no lymph node disease (2). We describe a unique case of a patient with extensive multifocal RDD involving the CNS, skull base, and paranasal sinuses with no evidence of lymphadenopathy on clinical examination. We review the literature and propose a surgical strategy to manage extensive multifocal disease in the symptomatic patient.

Case Report

A 39-year-old Hispanic male with known RDD and multiple previous surgeries for intracranial, spinal, and paranasal tumors presented with left facial numbness, obstructive sleep apnea, seizures, and worsening spasticity and gait dysfunction for the past 2 years. Previous treatments include resection of right nasal masses, left frontotemporal craniotomy, laminectomy (C2-C5 levels), placement of a ventriculoperitoneal shunt catheter to alleviate obstructive hydrocephalus, and orbitozygomatic craniotomy for resection of a left posterior fossa intradural tumor, and removal of a right mandibular radiculary cyst. Due to the progression of his symptoms, he expressed interest in surgical options.

On examination, his general physical examination was normal. No lymphadenopathy was appreciated on exam. On neurological examination, the patient was awake, alert, and oriented to person, place, and time. His short-term memory, extended short-term memory, ability to calculate, and language function were all normal. Cranial nerve examination revealed left facial sensory deficit with dysphasia especially in the V1 and V2 distribution and decreased left palpebral sensation. The patient was noted to have bilateral clonus in all extremities (Aushclle score 2.5 in upper extremities and 3.5 in lower extremities), as well as bilateral Hoffmann’s and Babinski’s signs. The remainder of his neurological examination was normal.

Neuroimaging with MRI with and without gadolinium demonstrated extensive enhancing soft tissue masses throughout the paranasal sinuses, multiple extra-axial enhancing masses at the skull bases, multifocal enhancing dural-based lesions in the brain including right frontal, right temporal, right cerebellopontine angle, left clinoidal, left petroclival and bilateral Meckel’s cave lesions (Figures 1A & B). Postoperative coronal and sagittal (C) and post-gadolinium T1-weighted MRI of the brain and paranasal sinuses.

Conclusion

Extensive multifocal RDD involving the brain, spine, and paranasal sinuses is exceedingly rare and often mimics meningiomas on imaging. Surgical management can be challenging. We recommend a conservative approach with close observation and reserve surgical treatment for enlarging symptomatic lesions.

References