





Case Report

Rosai-Dorfman sphenoorbital histiocytosis with intraparenchymal invasion: Do we have to consider this skull base pathology as a malignant disease?

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ABSTRACT

Background: Rosai-Dorfman disease (RDD) is a rare type of histiocytosis that can manifest with diverse symptoms. It usually presents with systemic involvement, and only a few cases have been reported at the level of the skull base. RDD typically follows a benign course during the progression of the disease. In this particular case reported, after the skull base invasion, the disease started to infiltrate the brain parenchyma. Our objective for this case report was to present this particular progression pattern and the nuances of its surgical treatment. In addition, a revision of the current literature was performed about skull base RDD with intracranial invasion and brain parenchyma infiltration not previously described.

Case Description: We are presenting the case study of a 57-year-old male patient who was experiencing severe headaches and an increase in volume in the right fronto-orbital region. On clinical examination, no neurologic clinical symptoms were observed. Contrast computed tomography and magnetic resonance imaging showed a tumor mass that affected the right orbit, frontal paranasal sinus, greater sphenoid wing, and right frontal lobe with moderate adjacent brain edema. The patient underwent surgery using an extended pterional approach with intracranial, orbital decompression, and frontal sinus cranialization, accompanied by frontal lobe tumor resection. Neuropathologic diagnosis revealed a Rosai-Dorfman histiocytosis disease.

Conclusion: The etiopathogenesis of RDD is still not completely understood. The current literature considers this disease to have a predominantly benign course. Nevertheless, as we have shown in this case, it may, in some cases, present direct parenchymal invasion. We consider that prompt surgical treatment should be ideal to avoid the local and systemic progression of the disease.

Keywords: Histiocytosis, Intraparenchymal invasion, Rosai-Dorfman disease, Skull base, Spheno-orbital

INTRODUCTION

Rosai-Dorfman disease (RDD) is a rare type of benign lymphoproliferative disorder, usually with a self-limiting course. Most cases involve lymph nodes in the cervical region, but it can also affect other areas of the body, representing 25–43% of cases outside of the lymph nodes.^[8] When it is present in the orbit, it usually causes affections in the eyelids.^[9] In rare cases, RDD can

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spread from the skull base area and invade the intracranial compartment.^[5,13,20] Until now, RDD brain parenchyma invasion extending from the skull base has not been reported.

Scarce literature on this disease has been reported concerning when it starts to compromise brain structures. This case aims to present the development and possible extensions of this type of disease. We describe some intricacies for the appropriate and timely treatment of this skull base lesion to avoid a local progression and intraparenchymal invasion.

LITERATURE REVIEW

We analyzed articles from PubMed from inception to 2024. Search terms included Rosai–Dorfman, orbit, intracranial invasion, surgical resection, skull base, and intraparenchymal invasion. Irrelevant and duplicated articles were removed. Incomplete cases and missing data were excluded from the study. This search yielded 46 articles, representing 67 cases in the skull base. From this, we only included 58 cases. To analyze the invasion of parenchyma from the extension of the skull base, for each case, magnetic resonance imaging (MRI) and surgical reports were visualized and analyzed by the authors. Demographic data and the location of the tumor were collected. Additional information concerning treatment was registered and divided into the type of surgical treatment (biopsy, complete resection, and incomplete/subtotal resection), surgery + radiotherapy, surgery + steroids, and surgery + radiotherapy + steroid treatment. Recurrence was considered if the patient had new clinical symptoms or a new finding was detected on MRI. Statistical analysis for demographic data and recurrence of the disease about the type of surgical treatment was performed using the software Statistical Package for the Social Sciences (version 25.0; IBM Corporation, Armonk, New York, USA).

RESULTS

We found 58 cases with RDD in the skull base area with intracranial invasion with a mean age of 36.74 (± 18.073) years, corresponding to 65.5% ($n: 38$) males and 34.5% ($n: 20$) females. In order of frequency, the sellar region was the most common affected area (27.6%; $n: 16$), followed by parasellar/middle fossa (25.9%; $n: 15$), and suprasellar (22.4%; $n: 13$). The intraorbital location with skull base involvement was only found in 6.9% ($n: 4$). In respect to the invasion, we only found a compromise of the cavernous sinus in 13.8% ($n: 8$).

Surgical treatment and recurrence

In all patients that we analyzed ($n: 58$), surgical treatment was divided into three modalities: complete resection (32.8%; $n: 20$), incomplete/subtotal resection (46.6%, $n: 27$), and biopsy (20.7%; $n: 12$). Patients received surgical resection without

adjuvant therapy in 72.4% ($n: 42$), surgical resection + radiotherapy in 13.8% ($n: 8$), surgical treatment + steroids in 6.9% ($n: 4$), and surgical treatment + radiotherapy + steroids in 6.9% ($n: 4$).

From the total of cases ($n: 58$), recurrence was present in 20.7% ($n: 12/P: 0.051$) in an average follow-up time of 28 months (± 26.878). For the patients from which only surgery was considered, recurrence for complete resection ($n: 20$) was found in 1 patient (5%). For incomplete/subtotal resection ($n: 27$), recurrence was present in 9 cases (33.3%), and for biopsy ($n: 12$), recurrence was found in 2 cases (16.7%) [Figure 1].

CASE DESCRIPTION

A 57-year-old man presented with persistent headaches and a noticeable increase in volume in the right fronto-orbital region, progressing over the past 3 months. The limitation of the ocular movements accompanied this progression of the disease. Clinical examination reveals only a slight right eye proptosis, with no visual deficit. Computed tomography (CT) and MRI show a solid mass involving the orbit, extending to the skull base, frontal sinus, and greater sphenoid wing with intracranial invasion, infiltration of the inferior frontal gyrus, and moderate brain edema [Figure 2].

Surgical resection

An extended pterional approach was performed on the site of the lesion to expose the greater sphenoid wing, roof, and lateral orbital walls.^[3] The tumor was infiltrating the frontal lobe parenchyma; this was completely resected with subpial dissection and extensive skull base bone drilling until negative pathology margins were achieved, leaving only a small portion from the intraorbital compartment, which was invasive to the intraconal structures. The dural graft was placed after the surgical resection, followed by the placement of a

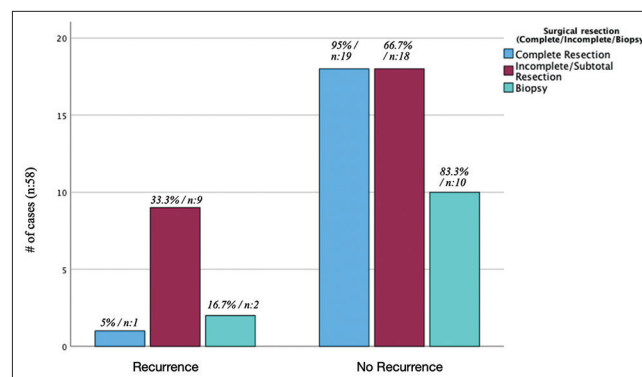


Figure 1: This graph shows the number and percentage of recurrence for patients with skull base Rosai–Dorfman disease after complete resection, subtotal resection, and biopsy.

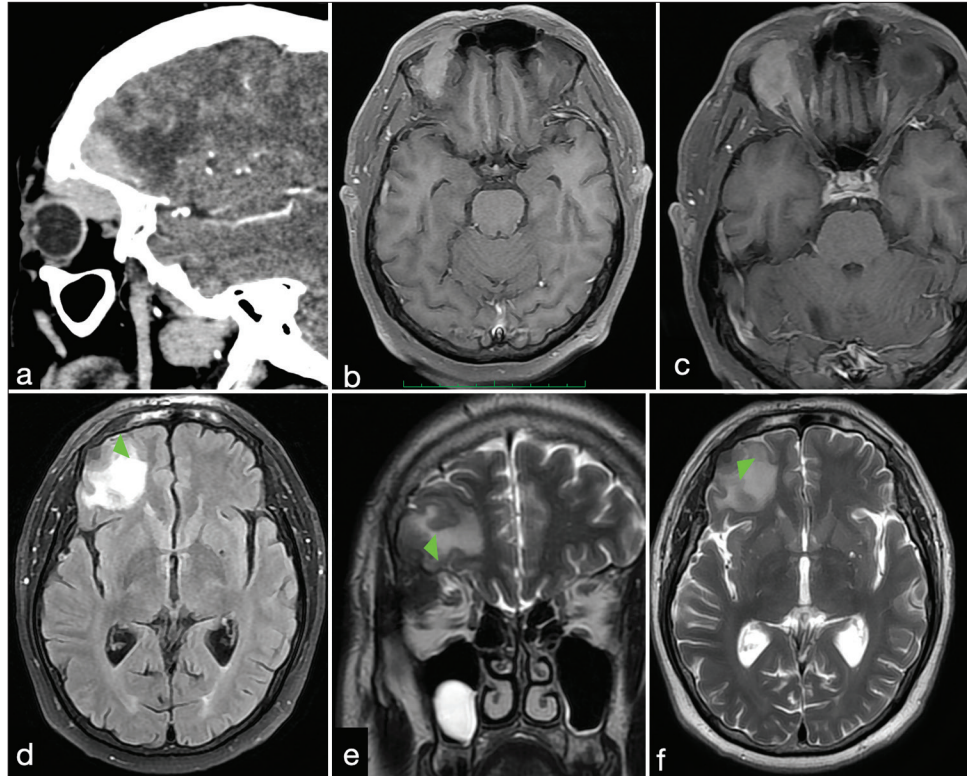


Figure 2: Computed tomography (CT) and magnetic resonance imaging (MRI). (a-c) CT and contrast MRI T1-weighted imaging with contrast shows enhancement of the lesion from the intraorbital area extending to the intracranial compartment with compromise of the frontal sinus. (d) Fluid-attenuated inversion recovery-MRI shows a hyperintense lesion adjacent to the tumor. Green arrowheads indicate the site of infiltration over the parenchyma. (e and f), This last was present on coronal and axial T2-weighted MRI, accompanied by peritumoral hyperintensity suggestive of perilesional edema. Green arrowheads indicate the site of infiltration over the parenchyma.

titanium mesh plate on the site of the craniotomy [Figure 3]. Pathology samples, accompanied by immunohistochemical analysis (S100, CD68 with IgG4), were consistent with RDD [Figure 4]. During the outpatient follow-up at 6 months, the patient did not have any clinical deficit.

DISCUSSION

In this study, we describe the surgical treatment of a patient who had intracranial fronto-orbital invasive histiocytosis (RDD). Due to its rarity, performing adequate surgical treatment for RDD can be challenging, mainly when it has an involvement of the orbit, skull base, paranasal sinus, and brain parenchyma invasion. To the best of our knowledge, this is the first article that reports and remarks on the potential for brain invasion by this disease.

RDD is commonly regarded as a benign, non-neoplastic, self-limiting, and extracranial histiocytic disease. Its etiology and pathogenesis remain poorly understood.^[15,17] In Latin

America, this kind of pathology is underdiagnosed. RDD is a rare proliferative histiocytic disorder first described in 1969.^[18] This disease has an incidence of 1:200,000, and the most common affected site corresponds to the glandular component, being the extra glandular presentation in 23%. Within these, affecting in order of frequency, skin in 12%, paranasal sinuses in 11%, orbit in 11%, soft tissues in 9%, and intracranial in ~3%.^[7] Concerning the skull base affected area, Hollon *et al.*,^[13] reported in their revision that the mean age of presentation was 36.7 years, with the most commonly located area in the sellar region.

After conducting a thorough analysis of the previous reports and their radiologic studies, we found that there were 58 cases of skull base RDD with intracranial extension and only five cases with suggestive brain involvement, even though not specifically commented on in the referenced literature.^[13,11,19,21,23] It is important to notice that in these last five cases, a disease portion was left over the brain parenchyma. In our case, we performed a complete intracranial surgical resection.

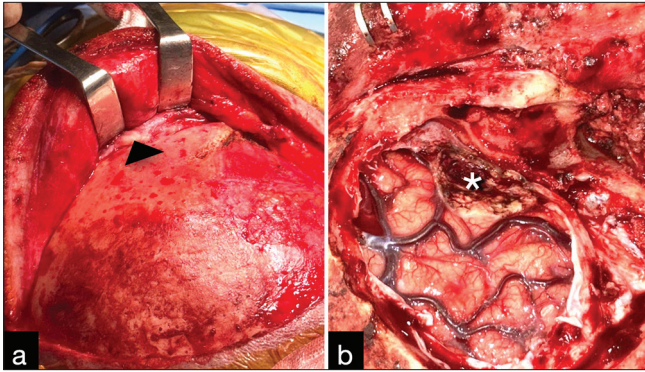


Figure 3: Surgical resection. (a) Cutaneous flap retraction with superior periorbital disease involvement. Interfascial dissection was performed to get adequate exposure (the head arrow indicates part of the orbital extension of the tumor). (b) Asterisk shows the resected tumor bed that was infiltrating the inferior frontal gyrus parenchyma.

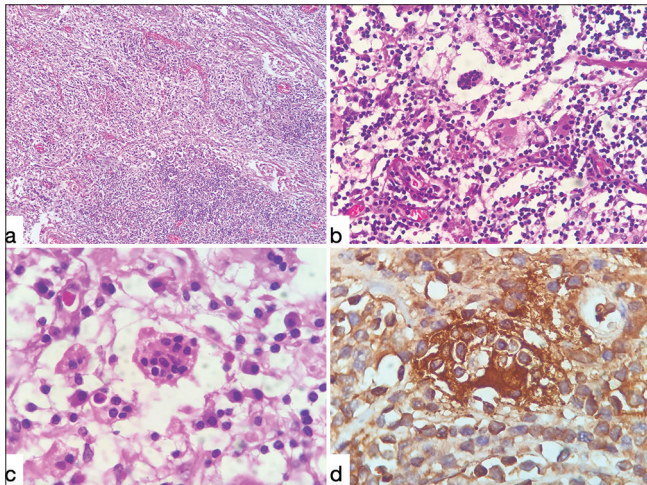


Figure 4: Histopathological findings. (a) Hematoxylin and eosin stain (H.E.). Magnifying glass panoramic photomicrographs recall the appearance of lymph node distribution. Small and large cells with abundant vessels and collagen fibers are noted. (b) (H.E. $\times 10$). Under medium magnification, histiocytic cells with clear or eosinophilic cytoplasm, with some lipids, and a nucleus with moderate pleomorphism can be noted. (c) (H.E. $\times 40$). Here, we can note the phenomenon of emperipolesis or lymphophagocytosis characteristic of this disorder. (d) (Immunohistochemical with S100 protein). This photomicrograph demonstrates the positivity of the cells that characterize the disease. (Courtesy from Professor Laura Chavez-Macias, Neuropathologist, Mexico City).

The cause of RDD is still unknown, and while there is more information available, it remains unclear. Various etiologies have been described, including herpesvirus and Epstein-Barr virus;^[4,14] with no identifiable clinical or demographic

history, the etiology may be consistent with genetic variations.^[4]

The clinical presentation of this disease can belong to several differential diagnoses.^[13] It usually requires a directed clinical suspicion. For a main instance, we had previously thought of metastases, meningioma, plasmacytoma, hemangiopericytoma, lymphoma, and multiple myeloma.^[16] For diagnosis, it may be necessary to complement with radiologic studies (CT, MRI) in addition to neuropathologic and immunohistochemical analysis. When the cranial vault or the skull base area is involved, it can mimic either a destructive or blastic lesion, orientating to a malignant disease.^[5,14,20] CT and MRI T1-weighted imaging may show a dural enhancement, which can suggest a meningioma.^[1,22] However, this finding was not observed in our case. MRI, in contrast, can help us to identify the brain parenchyma infiltration and guide the appropriate surgical treatment.^[5,13] This last finding could change the prognosis in terms of morbidity.

Intracranial RDD is a rare disease with no established treatment. In some reports, surgery, radiation, and chemotherapy have been effective in reducing neurological deficits.^[6,12,20] Descriptions in the literature report that approximately 78% of patients could have a stable course or regress after the initial surgical resection, regardless of the extent of resection.^[2] In the case we report, the disease remained stable with a self-limiting course. Some authors comment that after the surgical procedure, they may present disease stability,^[13] notwithstanding if the 2surgery was consistent with complete, incomplete/subtotal resection. Our extensive review of the literature on skull base RDD indicates that a complete surgical resection results in a 5% recurrence rate, while incomplete resection can lead to a higher recurrence rate of 33.3%. The combined treatment with steroids or radiotherapy may be a valuable therapeutic strategy in cases of incomplete resection or recurrence.^[5] In our analysis, the recurrence did not vary between the combination of modalities complemented with the surgical treatment. Relapse of central nervous system (CNS) RDD, after a complete surgical resection, has been reported in up to 11% of cases with a certain rate of complications up to 44%.^[5] After our analysis, we found that relapse of CNS RDD after non-total resection can be as high as 50%. Due to this last finding, a close follow-up of the progression of the disease is mandatory.

The main objective for the treatment of this pathology is to achieve a complete surgical resection, with the main objective to keep the neurovascular structures intact. The extent of resection apparently directly affects the disease progression and the morbidity of the patient.

If complete resection is not an option, adjuvant chemotherapy or radiotherapy may assist in the treatment of the disease.^[5,8,10]

Further research is necessary to gain a better understanding of the disease and lead to the development of more effective treatments to treat the disease opportunely without allowing it to cause damage to the brain tissue.

CONCLUSION

The cause of intracranial RDD is still not well understood. It is always important to treat this disease promptly to avoid its progression into brain parenchyma invasion. Complete surgical removal is the preferred treatment due to a lower recurrence rate, and its success depends on the extent of invasion and the surgeon's experience. For subtotal resection, radiotherapy or steroids may be considered as an adjuvant therapy.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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