

ISOLATED INTRACRANIAL ROSAI-DORFMAN DISEASE: CASE REPORT

Enfermedad de Rosai-Dorfman intracraneal aislada: Reporte de casoJORGE ZUMAETA S.^{1a}, FERNANDO PALACIOS S.^{1b}, WILLIAM ANICAMA L.^{2c}, CLAUDIA BURGOS J.^{2c}¹Department of Neurosurgery, Service of Vascular and Brain Tumors, ²Department of Pathology, Service of Surgical Pathology and Necropsies, Guillermo Almenara Irigoyen National Hospital, Lima, Peru.^a Resident of Neurosurgery, ^b Neurosurgeon, ^c Pathologist.**ABSTRACT**

Introduction: Rosai-Dorfman disease is a pathology of histiocytic, proliferative, idiopathic and benign type characterized by sinus histiocytosis and massive lymphadenopathy. The most frequent clinical presentation is painless bilateral cervical lymphadenopathy. Extra-nodal involvement occurs in 43% of cases and central nervous system (CNS) involvement in 4%. CNS involvement is more common in men and manifests itself as a mass in the cranial dura, which may or may not be associated with lymph node involvement.

Clinical case: We present the case of a 51-year-old woman with a history of sinusitis, with a clinical picture of holo-cranial headache, associated with periods of disorientation and ideomotor apraxia. MRI showed a right parieto-occipital extra-axial lesion, contrast sensor with implantation in the cranial dura. A right parietal craniotomy was performed with subtotal resection of the lesion. The pathological anatomy was reported as Rosai-Dorfman disease of meninges. The evolution after surgery was favorable with remission of symptoms.

Conclusion: Rosai-Dorfman disease should be within the differential diagnosis of lesions based on implantation in the dura. Its diagnosis is eminently histological. Although there is no specific therapy, surgical removal is the most effective treatment. Adjuvant therapies such as steroids and radiation can help control residual or recurrent disease.

Keywords: Histiocytosis Sinus, Lymph Nodes, Dura Mater, Meninges, Craniotomy (source: MeSH NLM)

RESUMEN

Introducción: La enfermedad de Rosai-Dorfman es una patología de tipo histiocítica, proliferativa, idiopática y benigna que se caracteriza por presentar histiocitosis sinusal y linfadenopatía masiva. La presentación clínica más frecuente es linfadenopatía cervical bilateral indolora. La afectación extranodal ocurre en el 43% de los casos y el compromiso del sistema nervioso central (SNC) en un 4%. El compromiso del SNC es más común en los varones y se manifiesta como una masa en la duramadre craneal, que puede estar asociada o no con afección ganglionar.

Caso Clínico. Presentamos el caso de una mujer de 51 años con antecedente de sinusitis, con cuadro clínico de cefalea holocraneal, asociado a periodos de desorientación y apraxia ideomotora. La Resonancia Magnética Cerebral mostró una lesión extra-axial parieto-occipital derecha, captadora de contraste con implantación en la duramadre craneal. Se realizó una craneotomía parietal derecha con resección subtotal de la lesión. La anatomía patológica fue informada como Enfermedad de Rosai-Dorfman de meninges. La evolución luego de la cirugía fue favorable con remisión de los síntomas.

Conclusión. La enfermedad de Rosai-Dorfman debería estar dentro del diagnóstico diferencial de lesiones con base de implantación en la duramadre. Su diagnóstico es eminentemente histológico. A pesar de que no existe una terapia específica, la extirpación quirúrgica es el tratamiento más eficaz. Las terapias adyuvantes como los esteroides y la radiación pueden ayudar a controlar la enfermedad residual o recurrente.

Palabras clave: Histiocitosis Sinusal, Ganglios Linfáticos, Duramadre, Meninges, Craneotomía (fuente: DeCS Bireme)

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Rosai-Dorfman Disease is a benign, idiopathic, and proliferative histiocytic disorder characterized by sinus histiocytosis and massive lymphadenopathy.^{1,2,3} Its

importance was recognized until in 1969 Rosai and Dorfman described 4 cases they called sinus histiocytosis with massive lymphadenopathy, entity which is different from that of histiocytosis X.⁴ It mainly affects patients in their second decade of life and there seems to be a slight

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predominance of males. The most common clinical presentation is painless bilateral cervical lymphadenopathy.^{5,6} The most common sites of involvement are the skin, paranasal sinuses, soft tissues, bone, and central nervous system.^{1,4} Extranodal disease can occur in 43%, however, in less than 5% of cases, the disease has been reported in the central nervous system and in the majority with lymph node involvement, with reports of isolated intracranial involvement being exceptional.⁷⁻⁹ Rosai-Dorfman disease with involvement of the central nervous system occurs more in men in the fourth decade of life and typically manifests as a mass in the cranial dura, which may or may not be associated with lymph node involvement.^{4,10} Neurological manifestations due to involvement of the central nervous system, they are extremely rare (4%), with intracranial involvement being even less frequent in the absence of node involvement (0.5%).¹¹ Symptoms due to central nervous system involvement include headaches, seizures, loss of vision, fever and motor deficit.¹²

Intracranial lesions are usually presented as extra-axial lesions with a dural base both at the convexity and at the cranial base,^{13,14} cases with bone erosion, compromise of the dural sinuses and intraparenchymal location have been reported.^{10, 15, 16, 17} On Cerebral Tomography, the lesion appears as a homogeneous and lobulated mass with strong contrast uptake. On brain magnetic resonance imaging (MRI) the T1 image is typically isointense to the adjacent brain parenchyma with strong enhancement after contrast administration. Likewise, in the T2 image, the lesion appears hypo or isointense with surrounding vasogenic edema.^{10,18} Perfusion-weighted images and MRI spectroscopy have been used to differentiate Rosai-Dorfman disease as an inflammatory process instead of a neoplastic one. However, these modalities cannot be used to reliably diagnose based solely on image.^{12,19}

The diagnosis is confirmed with histopathology which shows a characteristic emperipolesis; immunohistochemical staining reveals characteristic histiocytes, positive for protein S100 and CD68 and negative for CD1a.^{10, 20} The

prognosis is good, 40% patients experience spontaneous remission with treatment with oral corticosteroids, although it can also be managed with low doses of fractionated radiotherapy with good long-term response, or even radical surgery of the lesion.¹ In intracranial Rosai-Dorfman disease, surgical resection seems to be the most effective therapy.^{21, 22} The exact etiology and adjuvant therapy for recurrent injuries have not yet been established.

In this article we present a case of intracranial Rosai-Dorfman disease without lymph node involvement.

CLINICAL CASE

History and examination: 51-year-old female patient with a history of sinusitis, without previous surgeries, without drug allergies. He presents a clinical picture of 2 years of evolution with an insidious onset and a progressive course characterized by intermittent holocranial headache of predominantly right periorbital origin associated subsequently with disorientation, difficulty in describing objects, apraxia and loss of the ability to perform voluntary movements for 3 to 5 minutes, which remit spontaneously. On physical examination, the patient was awake, spontaneously ventilating, Glasgow 14 points, without motor or sensory deficits, photoreactive and isochoric pupils, no adenopathies were observed. Symptoms improved after corticosteroid treatment. Cerebral magnetic resonance imaging (MRI) showed an extra-axial expansive lesion dependent on the meninges in the right parietal-occipital region with subcortical cortical extension, in slightly hyperintense T1, in isointense T2, the lesion eagerly highlights the contrast in a homogeneous way, vasogenic edema is observed around the lesion (**Fig. 1**). Cerebral tomography (CT) showed an extra-axial lesion adjacent to the right parietal lobe, homogeneously receiving contrast, without apparent bone erosion, associated with vasogenic edema (**Fig. 2**). These characteristics suggested the diagnosis of meningioma.

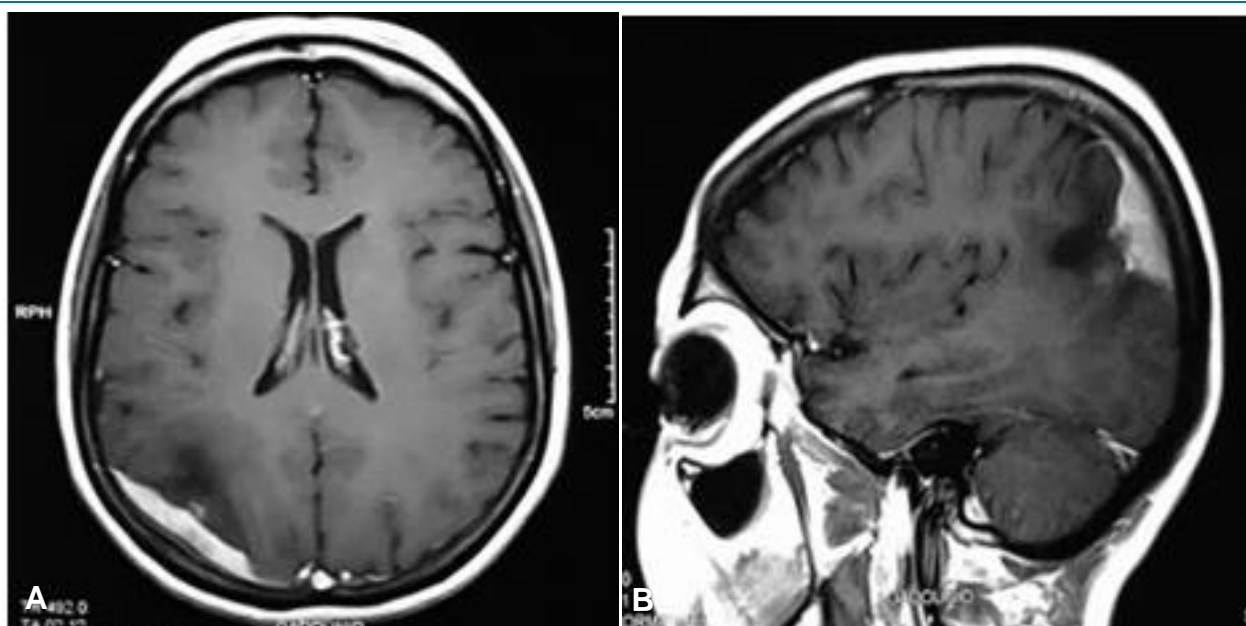


Fig 1. Brain MRI in axial (A) and sagittal (B) showing right parieto-occipital lesion that show contrast enhancement and implantation in the cranial dura.

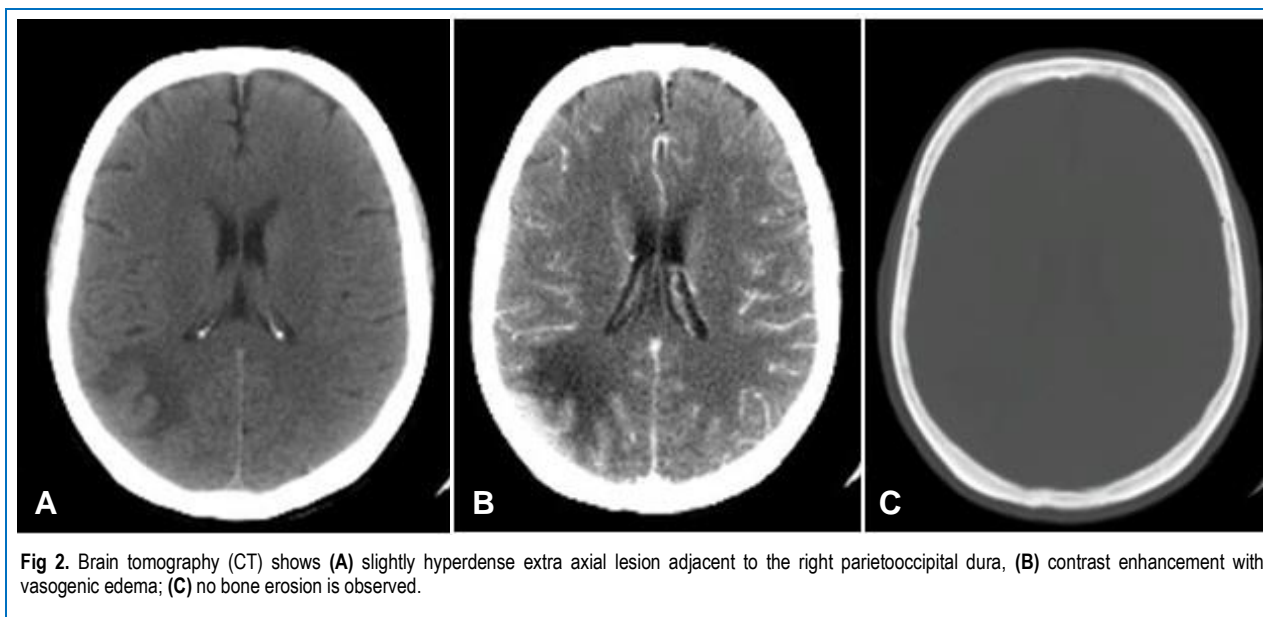


Fig 2. Brain tomography (CT) shows (A) slightly hyperdense extra axial lesion adjacent to the right parietooccipital dura, (B) contrast enhancement with vasogenic edema; (C) no bone erosion is observed.

Treatment: A right parietal craniotomy and subtotal resection of the right parieto-occipital lesion (Simpson III) plus duroplasty with galea were performed. A pseudo capsule of hard fibrous tissue was found. A 1 cm tumor fragment was left because it was very attached to a cortical drainage vein, the fragment was coagulated. After surgery, a control brain CT was performed, showing the right parietal craniotomy and subtotal excision of the right parieto-occipital lesion (**Fig. 3**).

Clinical evolution: Subsequently, the patient was transferred to the neurosurgical intensive care unit where she remained under sedo-analgesia. She presented volume depletion requiring blood transfusion with which it was

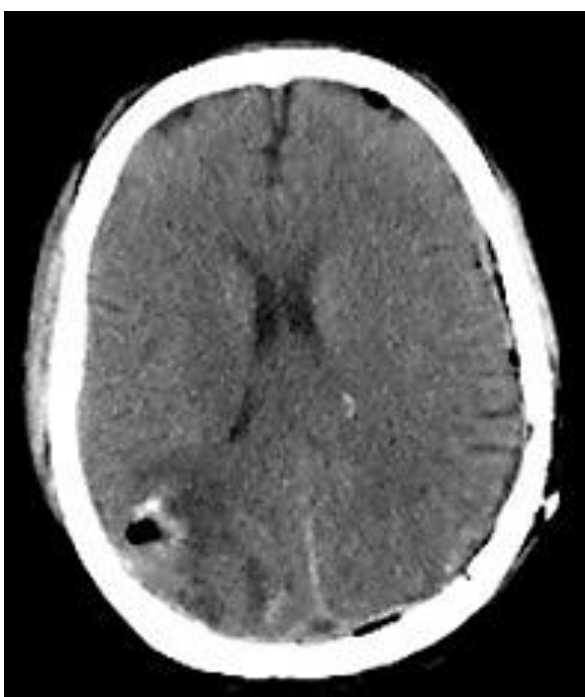


Fig 3. Brain CT scan in the immediate postoperative period showing parietal craniotomy and subtotal resection of the right parieto-occipital lesion.

compensated. He also had respiratory failure that resolved with medical treatment. In the following days, the patient presented a favorable evolution, being awake, oriented, Glasgow scale: 15, wound with no signs of infection, and therefore discharge from hospital was decided.

The initial report of Pathological Anatomy indicated connective tissue with severe nonspecific chronic inflammation, so it was decided to carry out an additional immunohistochemical study. The results were of a lesion showing emperipolesis and histiocytes positive for the S100 and CD68 protein, being negative for CD1a confirming the diagnosis of Rosai-Dorfman disease (**Fig. 4**). Patient in his outpatient control has continued with a favorable evolution, remitting symptoms, control images at one month and at 6 months they have not shown recurrence of the disease (**Figs 5 and 6**).

DISCUSSION

Rosai-Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy, was first recognized as a distinct clinicopathological entity by Rosai and Dorfman in 1969. It is considered a self-limiting, benign, nonneoplastic histiocytic disorder, although its etiology and pathogenesis continue being little known.^{5,20} Involvement of the central nervous system in Rosai-Dorfman disease is rare (approximately 5% of all cases), with 210 cases reported worldwide, of which 174 cases presented isolated and These 134 were isolated intracranial involvement.²³ In our hospital it is the first reported case and there is no record of any other case in our country.

This pathology has a male predominance²⁴ and more commonly involves young and adult patients with an average age of 35.7 years.²⁵ Cases have also been reported in children.²⁶ In our case, it was a female patient which is the gender less affected, and her age was 51 years old which is the upper limit of the presentation range. Common symptoms include headache, seizures, limb weakness, and cranial nerve dysfunction depending on the location of the lesions.^{27, 28, 29, 30} Our patient presented symptoms that match the description in the literature, characterized mainly

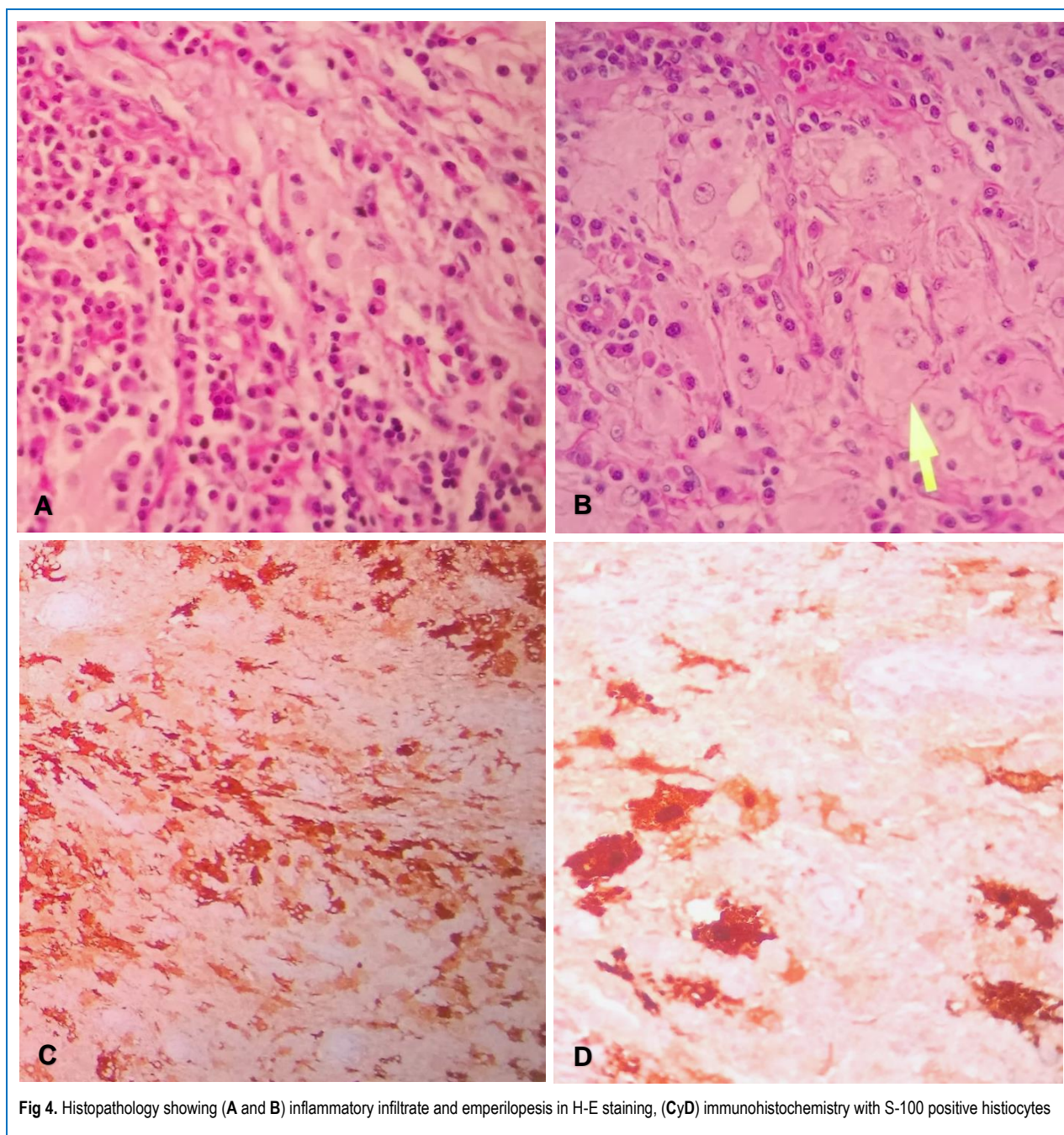


Fig 4. Histopathology showing (A and B) inflammatory infiltrate and emperipolesis in H-E staining, (CyD) immunohistochemistry with S-100 positive histiocytes

by headache and periods of confusion associated with difficulty in making voluntary movements for short periods of time with spontaneous recovery that suggest us to think of simple partial convulsive episodes since there was no loss of consciousness.

Rosai-Dorfman disease with involvement of the central nervous system usually presents as a lesion associated with the dura, extra-axial, with homogeneous contrast enhancement that mimics a meningioma, this added to its infrequency makes diagnosis by neuroimaging difficult.^{3,31,32,33} The lesions are generally located at the convex level,²⁴ other rare locations are the parasagittal, suprasellar, cavernous, petroclival, and spinal regions.^{34,35,36} Extremely rare cases of intraventricular and intraparenchymal lesions have also been reported,^{37, 38} including a fatal case of injury at the level of the

brainstem.³⁹ The injury is generally unique although cases of multiple intracranial injuries have been reported.^{40, 41}

T1 Magnetic Resonance images generally show an isointense or hyperintense lesion with well-defined edges and with significant contrast enhancement and in a homogeneous manner. In T2, the lesions are usually isointense.⁴² In our patient, the MRI lesions were slightly hyperintense in T1 and isointense in T2; standing out with the contrast in an avid and homogeneous way, which coincides with what was previously mentioned. It should be noted that these characteristics led to the pre-operative diagnosis of meningioma, which is also reported in the literature as the main differential diagnosis, additionally including histiocytosis X, lymphoproliferative disorders, plasma cell granuloma, and infectious diseases. During surgery, an injury can be found with a clearly thick and hard tunic, different from what is commonly found in a meningioma.²⁴

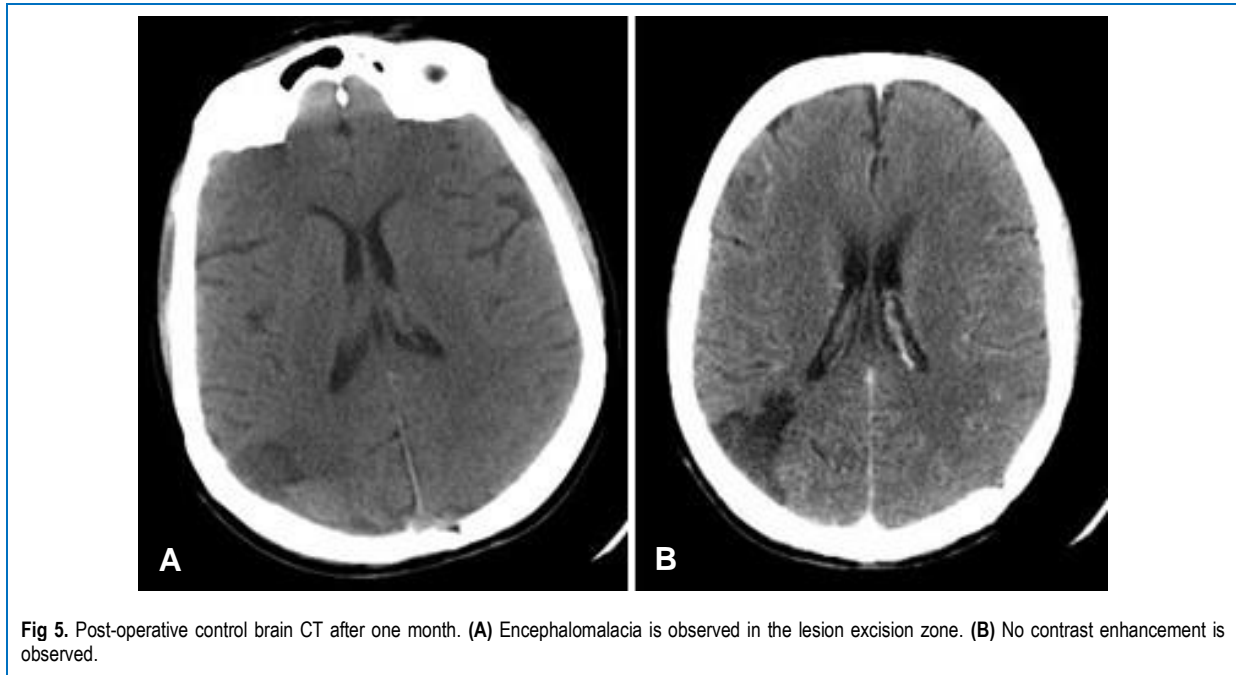


Fig 5. Post-operative control brain CT after one month. (A) Encephalomalacia is observed in the lesion excision zone. (B) No contrast enhancement is observed.

Histologically, Rosai-Dorfman's disease is characterized by the presence of multinucleated histiocyte sheets with emperipolesis, which is an entrapment of lymphocytes in the cytoplasm of the histiocytes. The histiocytes that are the antigen presenting cells show the histochemical record characterized by being positive for the S-100 protein and the phagocytic histiocytes positive for CD 68, α -1 antitrypsin, lysozymes, MAC-378 but negative for CD1a. Emperipolesis, however, could be seen in around 70% of RDD patients.⁴³

In the case of our patient, it was initially reported as nonspecific inflammatory tissue, which is why the study with immunohistochemical markers was deepened, the sample presenting emperipolesis and being positive for S-100 and CD 68, and negative for CD1a. These results confirmed the diagnosis of Rosai-Dorfman disease.

The treatment of systemic Rosai-Dorfman's disease is recommended only in patients with symptomatic lesions or masses that have damaged the function of vital organs since this disease is considered to be a benign, non-neoplastic lymphoproliferative disorder.²³ In addition, there are reports of complete spontaneous resolution in approximately 20% of patients with systemic disease.⁵ However, complete spontaneous resolution has not been reported in intracranial disease. Consequently, surgery is still the first treatment option in intracranial disease for diagnostic purposes, also achieving improvement in neurological symptoms.^{44, 45} Complete resection should be attempted when the lesions do not adhere to critical surrounding structures, subtotal resection should be carried out for pathological diagnosis and remission of symptoms.

After surgery, the majority of patients can achieve stabilization of the disease and therefore conservative treatment and follow-up by means of periodic controls are recommended.²¹ Furthermore, radiotherapy, chemotherapy and corticosteroid therapy have been used in patients who did not obtain improvement in their neurological symptoms after surgery.⁴⁶

Both stereotactic fractional radiotherapy and common radiotherapy have been used in intracranial disease after surgery, however, the results were variable, and some patients even showed no improvement.^{21,47} Although steroid therapy has shown some therapeutic benefit in treating patients with systemic disease and in some patients with intracranial disease, randomized trials are still required to standardize its use. Chemotherapy has also been used in systemic and intracranial disease⁴⁸ with a generally ineffective result, although it has been recommended as adjuvant treatment after subtotal resection.⁴⁹ In our case, surgical resection of the right parieto-occipital lesion was performed.

In our case, until the last clinical evaluation of the patient surgical treatment has been shown to be optimal and first-line, coinciding with the literature. The patient has improved from her initial symptoms and has not presented a recurrence in her imaging controls. Despite this, close follow-up is maintained since, being a rare pathology, post-surgery management has not yet been fully elucidated. Our patient has not required additional management that includes corticotherapy, radiotherapy or chemotherapy.

CONCLUSION

Intracranial Rosai-Dorfman Disease is a rare, benign, non-infectious histiocytic disease that mimics meningioma and other granulomatous conditions. While systemic disease affects children and young adults the most, intracranial involvement is a more frequent disease in adults. The etiology and pathogenesis remain unclear despite various hypotheses. Intracranial disease presents with symptoms of increased intracranial pressure, focal or generalized seizures. Radiologically it appears usually as a dura-based injury mimicking a meningioma. Histopathology is essential for diagnosis.

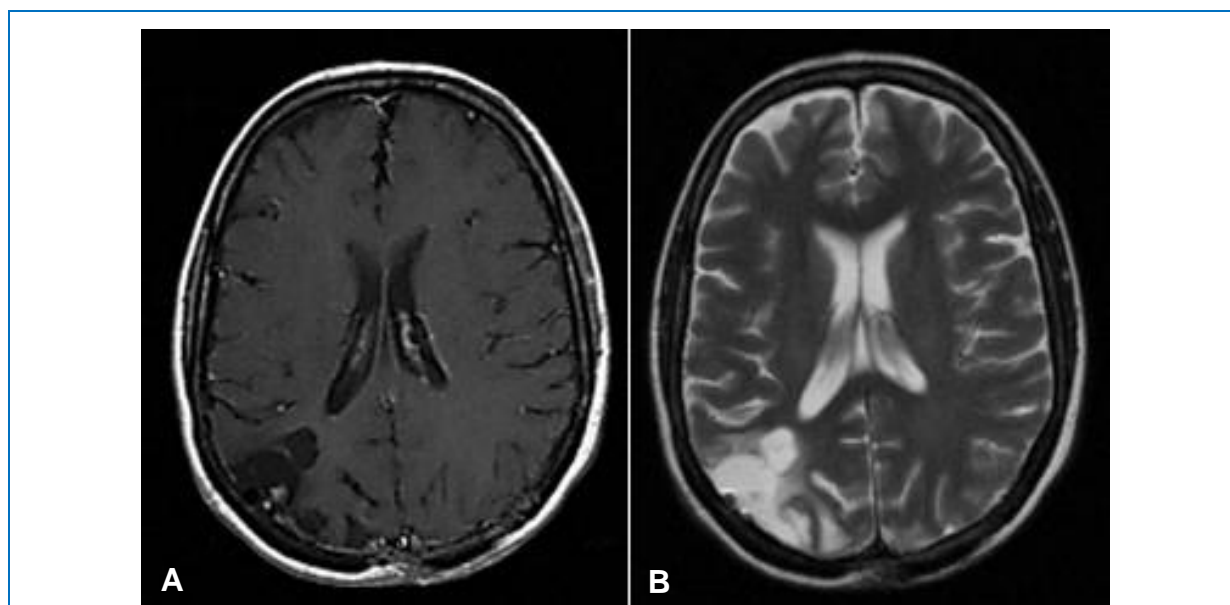


Fig 6. Control brain MRI at the sixth month after surgery. (A) In T1 with contrast, there is an area of encephalomalacia in the tumor area and lack of contrast enhancement. (B) In T2 where hyperintensity is seen in the resection area of the tumor lesion.

Surgical removal is the most effective treatment in most cases, since it eliminates the mass effect and provides tissue diagnosis. Adjuvant therapies like steroids and radiation can help control residual or recurrent disease. Simple observation is also an acceptable strategy for stable disease. The prognosis of Rosai-Dorfman's disease is good where there is no nodal or multi-organ involvement. Recurrences are rare after initial surgical resection, but when they do occur, they occurred after incomplete resection. The precise diagnosis of this disease and the differentiation from other common conditions are very important since, the natural history and the prognosis seem to be more favorable for this pathology.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

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