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Case report

Primary isolated intracranial Rosai–Dorfman disease: Report of a rare case and review of the literature

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ABSTRACT

Background: Intracranial involvement is an uncommon manifestation of Rosai–Dorfman disease (RDD) and had been rarely reported. In this study, we explore clinical characteristics, imageology manifestations and pathological features of primary intracranial RDD so as to improve the understanding for this disease.

Methods: One case (16-years-old boy) with primary intracranial RDD was analyzed and studied retrospectively by MRI features, histopathological observation and immunohistochemical staining, and the related literatures were reviewed.

Results: The case was single lesion and involved the dura of the left middle cranial fossa base, which was iso-hypo signal intensity on T1WI and hypointense on T2WI and FLAIR image. The lesion was a homogeneous contrast enhancement mass with dural tail sign and had peritumoral brain edema. Pathological analysis showed the lesion consisted of variable numbers of mature lymphocytes, plasma cells and neutrophils. The characteristic histiocytes were emperipolesis and positively expressed for S-100 and CD-68 and negatively expressed for CD-1a by immunohistochemical analysis. Based on clinical presentations and histological findings after surgical excision, a final diagnosis of primary intracranial RDD was made.

Conclusion: Primary intracranial RDD, especially located in the cranial base, is exceptionally rare, which hard to be distinguished with meningoma by imageology and clinical manifestations, but could be diagnosed by pathological and immunohistochemical examinations. Surgery is of the most importance treatment and prognosis is optimistic for this disease.

1. Introduction

Rosai–Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy (SHML), which is an idiopathic benign histiocytic proliferation affecting lymph nodes, and was first described by Lucien Destombes in 1965 and was recognized as a distinct clinicopathologic entity by Rosai and Dorfman in 1969 [1]. Although extranodal involvement has been reported in diverse sites, intracranial presentation of this disease is extremely rare.

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Only a few cases have been reported in literatures so far. And it was basically occurred in adults. In this article, we described a 16-years-old boy who had an isolated intracranial involvement of RDD. The current literatures and treatment options were also reviewed.

2. Case report

A 16-years-old boy, without a significant past history, presented with a generalized tonic-clonic seizure. He was admitted to our neurosurgery department for an abrupt onset of tremor and jerking of his Limbs, lock-jaw and loss of consciousness. For about 3 min, he gradually regained consciousness. After the attack, he also complained of dizziness, headache and weakness. He had never experienced this symptom before. His family history was negative for tumors. The neurological examination showed no abnormalities.

Brain MRI showed a homogenous contrast enhancement mass measured approximately 3.3*3.2*1.2 cm and enhancing dura tail in the left temporal fossa region (Fig. 1A–C). T1-weighted MRI revealed the lesion to be iso-hypointense (Fig. 1D), and T2 (Fig. 1E–G) and FLARE (Fig. 1H) predominant hypointense and perilesion edema. DWI also showed circular

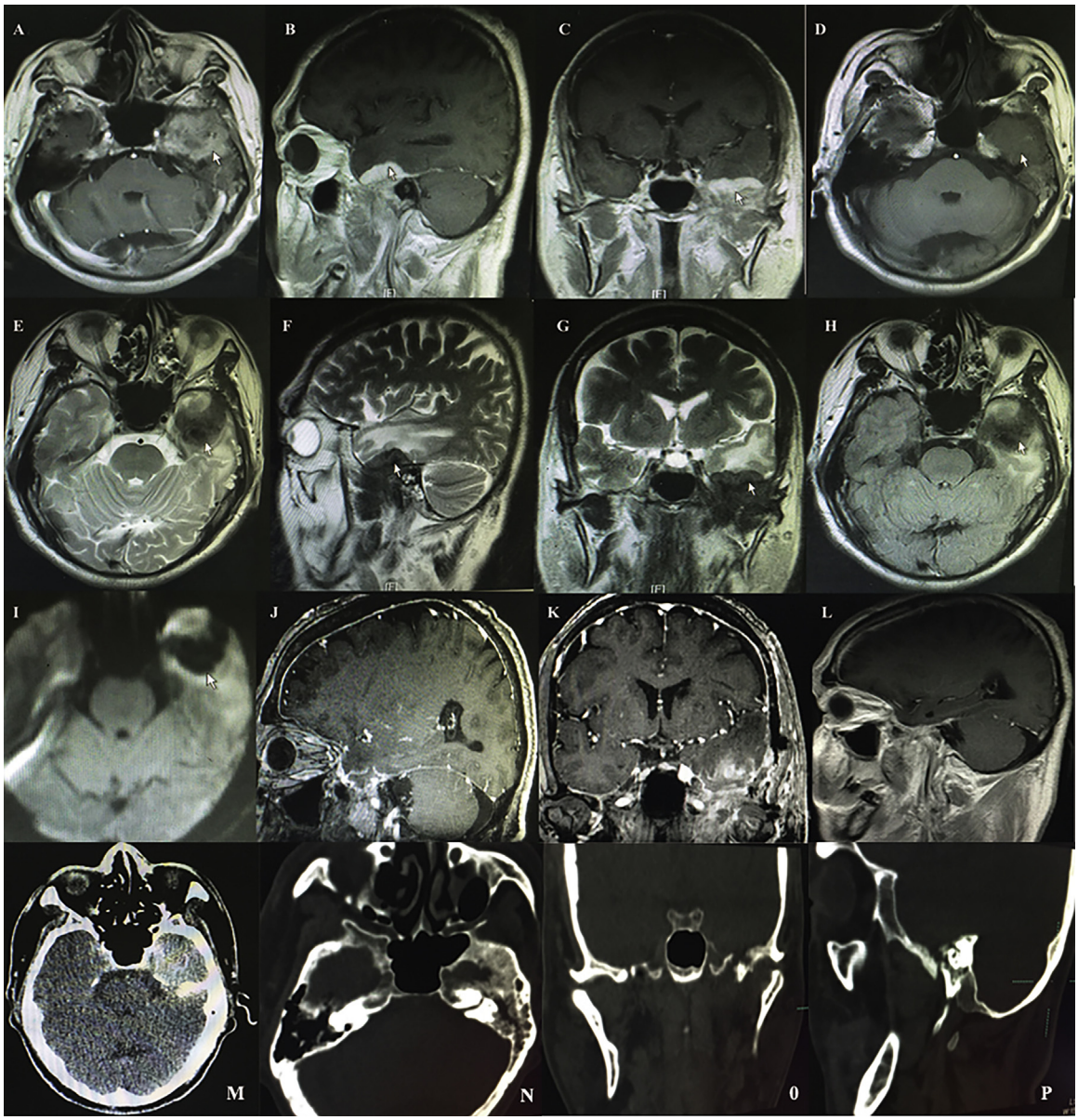

Fig. 1 – Brain MRI with contrast showed the enhancing lesion and the dural tail (A–C). T1 weighted MRI revealed the lesion to be iso-hypointense (D), and T2 (E–G) and FLARE (H) predominant hypointense and perilesion edema. DWI also showed circular hyperintense surrounding lesion and very hypointense for lesion (I). Non-contrast axial CT scan showed a homogeneously hyper-dense lesion without any internal calcifications and hemorrhage in the left middle cranial fossa base (M). Hyperplasia and destruction of skull base bone were seen according to three-dimensional CT scan (N–P). Post-operative MRI of the brain showed total excision of the lesion (J, K) and without evidence of recurrence after 5 years of follow-up (L).

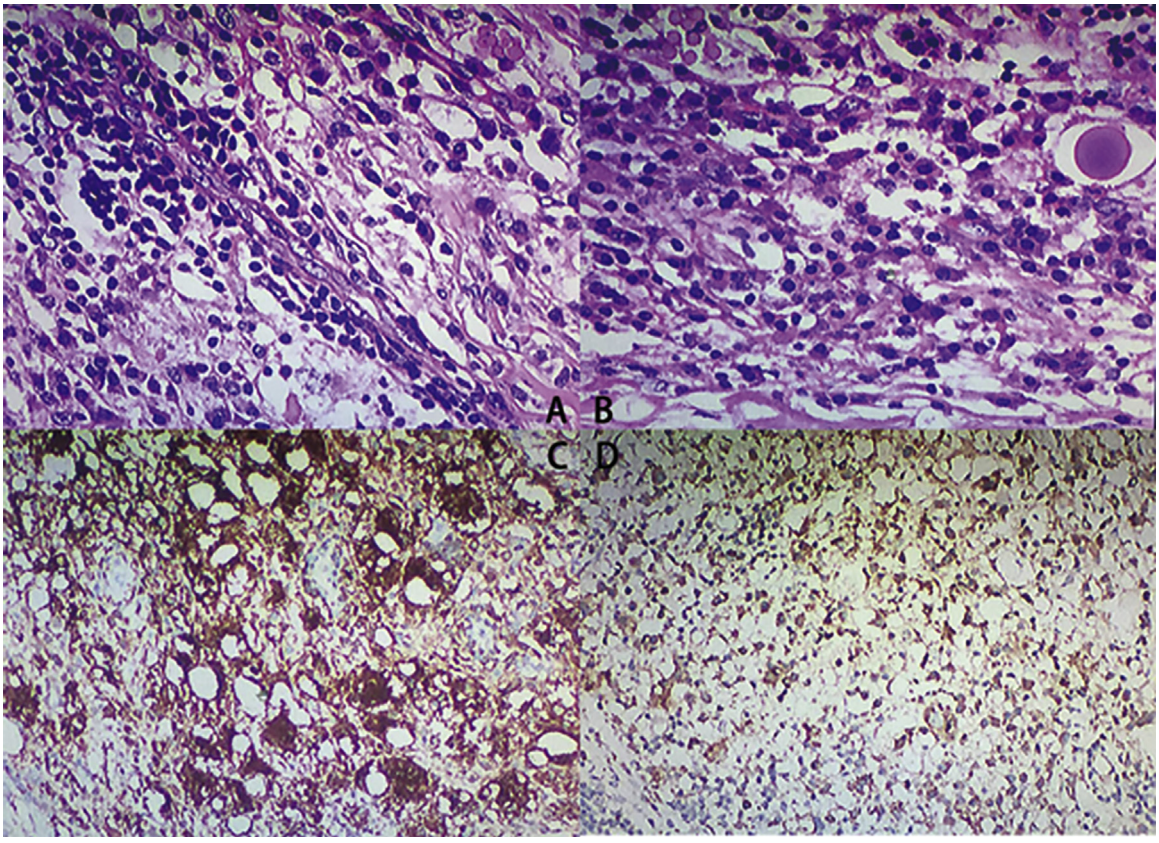

Fig. 2 – Microscopically, section showed infiltration of abundant lymphocytes, plasma cells and neutrophils, and some histiocytes scattered in fibrotic background. Histiocytic cells occasionally showed emperipolesis with intact lymphocytes, plasma cells and neutrophils (A, B, H&E, 400×). On immunohistochemical examination, histiocytes are positive for S-100 (C) and CD68 (D) (400×), but negative for CD1a.

hyperintense surrounding lesion and very hypointense for lesion (Fig. 1I). Axial plain CT scan showed a lesion which was relatively hyperdense located in the left skull base, and no evidence of any calcification and hemorrhage was observed within (Fig. 1M). Hyperplasia and destruction of skull base bone were seen according to three-dimensional CT scan (Fig. 1N–P).

Electroencephalogram did not show typical diffuse spike and slow waves activity. Blood examinations, Urine test, CSF analysis and others test were also normal.

The patient underwent craniotomy and total resection of the lesion. During the surgery, the tumor was gray-white, well-circumscribed, relatively avascular and tough in consistency. There was no clear arachnoid boundary between the lesion and the cortex, and the tumor tightly adhered to brain parenchyma and skull.

Histologic examinations showed that proliferation of histiocytic cells and infiltration of lymphocytes, plasma cells and neutrophils were seen (Fig. 2A and B). Immunohistochemical stainings were strongly positive for S-100 (Fig. 2C), CD68 (Fig. 2D), Vimentin, Kappa and Lambda, but negative for CD1a, CD30, EMA, and GFAP, and Ki67 less than 5%.

Based on these findings above, thus the final diagnosis was RDD. Post-operative MRI showed total excision of the lesion (Fig. 1J and K). After 5 years of follow-up, the patient was well and asymptomatic, without clinical evidence of recurrence (Fig. 1L).

3. Discussion

RDD often involve lymph nodes, and the most common extranodal sites are skin, nasal cavity, paranasal sinus, eyelid, orbit, bone, salivary gland and CNS. Although extranodal involvement has been reported in diverse sites, primary isolated intracranial RDD without lymphadenopathy is extremely rare (less than 5% of RDD) [2]. In the available literatures, approximately 75% of CNS RDD is intracranial, whereas less involve the spine [3]. Occasionally, simultaneous involvement of the brain and spinal cord had also been described [3]. Patients with intracranial involvement usually present with headache and seizures. They can also present with dysphasia, cranial nerve deficit, hemiparesis, and endocrine dysfunction mainly depending on the location and size of the lesion. The precise etiology, pathogenesis, and natural history of RDD are still unknown. Although researches revealed some underlying association between RDD and bacteria, virus and immune dysfunction, a definitive agent has never been isolated [4,5].

The MRI characteristics of intracranial dura-based RDD are very similar to that of meningioma, thus it is the most easily to be diagnosed as meningioma according to MRI. On MRI, although the image is uncharacteristic, the typical appearance is a dura-based homogeneously enhancing mass, which is associated with peritumoral edema and having a dural tail, iso-hypointense on T1-weighted images and isointensity or very low signal intensity on T2-weighted images [6]. These MRI features may help us to differentiate RDD lesions from the meningiomas. Our case presented as a meningioma-like, extra-parenchymal, dural-based mass, which also had very low signal intensity areas on T2-weighted and FLAIR images on MRI. The lesion was smaller, but the peritumoral edema was remarkable.

The histopathologic differential diagnosis includes lymphoma, plasma cell granuloma, Langerhans cell histiocytosis (LCH) and plasma cell rich meningioma. In our case, considering the lack of atypical lymphocytes, lymphoma was unlikely. Because of the presence of emperipolesis, plasma cell granuloma could be excluded. LCH was also unlikely, because Langerhans histiocytes with their folded nuclei and eosinophil infiltrates were absent, and the CD1a was negative in the tumor. Radiologically, contrast enhancement and perilesional edema are common to both lymphoplasmacyte-rich meningiomas and RDD, but S100 and EMA are positivity in the plasma cell rich meningioma [7]. In view of these possibilities, histological and immunohistochemical examinations are essential for a definitive diagnosis of RDD. In addition, the presence of emperipolesis is a hallmark of the disease, even if this may be less marked in intracranial disease [8]. In our case, the findings of emperipolesis, positivity for CD68 and S100 and negativity for CD1a were coherent with the diagnosis of RDD.

There are no specific treatment guidelines for intracranial RDD. Although a variety of treatment methods have been used, including steroid therapy, chemotherapeutic agents and radiation, surgical resection appeared to be the most appropriate choice [3,9,10]. When surgical excision is not possible and recurrence or residual after surgery, adjuvant therapy such as local low dose radiotherapy and steroids can be used.

Prognosis of the disease is optimistic, and no death was reported. Our patient, who had a single operation for the total resection of the lesion and did not receive any further therapies, was followed up over a period of 5 years and had no recurrent.

4. Conclusions

In summary, we provide detailed information for intracranial RDD. Despite intracranial RDD is uncommon, more attention should be given.

Funding

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Conflict of interest

None declared.

Disclosure

The authors have no personal, financial, or institutional interests in any of the drugs, materials, or devices described in this article.

Informed consent

The patient was asked to sign an informed consent for the consent study. The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association for experiments involving humans.

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