Rosai-Dorfman Disease of the Pineal Region: A Case Report

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Citation

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Abstract

Rosai-Dorfman disease (RDD) is rare, especially in the central nervous system (CNS). We report a 4-year-old boy with a neoplasm in the pineal region, verified as RDD, with no concurrent clinical lymph node involvement. Emperipolesis was also identified in this case. Fibrosis was additionally found at extranodal sites. Positive results for the S100 protein and negative findings of CD1a were important for diagnosis.

INTRODUCTION

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is an idiopathic histiocytic disease of lymph node and extranodal sites. The most common extranodal sites include the skin, orbit, and upper respiratory tract; involvement of the central nervous system, especially in the absence of nodal disease, is rare.

CASE REPORT

A 4-year-old boy presented with a history of esotropia of the right eye for 1 year and cohesion of the left eye for 20 days. Projectile vomiting was also evident. Physical examination revealed that ocular fixation and Parinaud syndrome appeared in both eyes. Enlarged lymph nodes were not palpated. Magnetic resonance imaging results showed an isointense lesion in the pineal region (Fig. 1), which blocked the interventricular foramen and compressed the third ventricle. A mass with integrated capsule as found in the pineal region during surgery, consisting of damaged, grayish-white tissue with a solid surface. Microscopically, the lesion was comprised of regions of dark and bright with fibers arranged in a storiform pattern and some pinealocytes arranged in a nested pattern (Fig. 2). Histiocytes were mixed with lymphocytes and plasma cells in the fibrous background. Typical histiocytes showed indistinct cellular margins with abundant cytoplasm. The nuclei were round or oval. Occasional histiocytes were multinucleated. Unequivocal emperipolesis could be identified in hematoxylin - eosin stained section(fig - 3). The histiocytes were strongly positive for S100 (fig - 4) and CD68 protein, and not reactive to CD1a. Staining for CD3 and CD20 showed a mixed population of T and B lymphocytes in the background. Staining for CK、EMA、CD117、NF presented negative. Staining for NSE and Syn illustrated positive in the clumps of pineal cells. The fraction of Ki67-positive cells is 10%.The histopathological diagnosis was Rosai-Dorfman disease of the pineal region.

Figure 1

Fig 1 Sagittal T1- weighted MRI showing an isointense lesion in the pineal region.

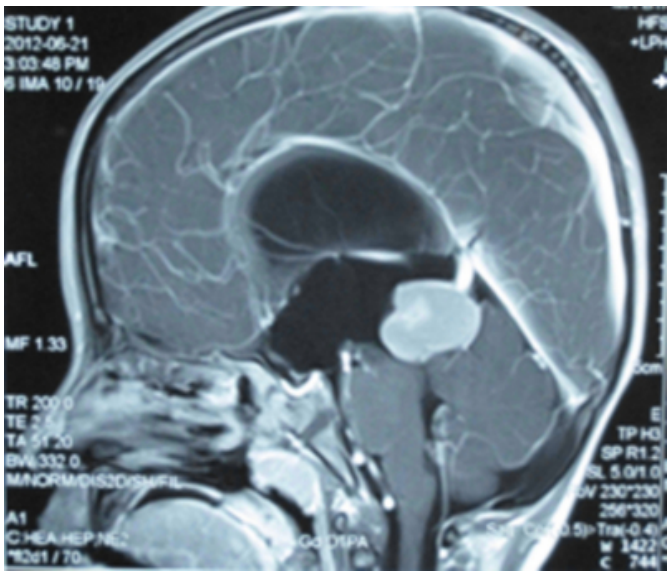


Figure 2

Fig 2 shows a clump of pineal cells in the lesion (H&E x 100).

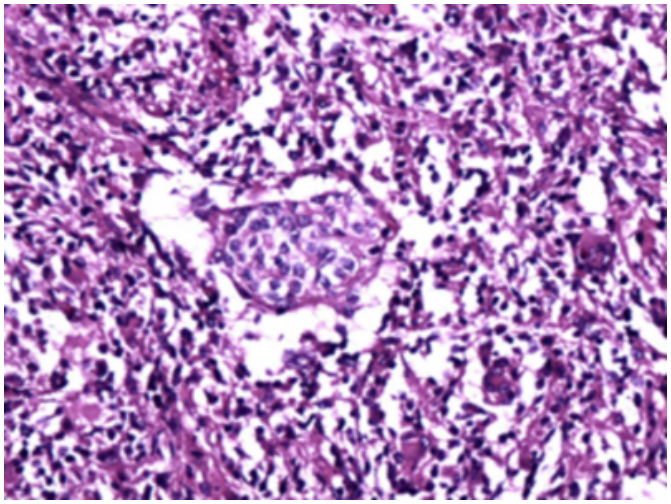


Figure 3

Fig 3 The histiocytes have phagocytosed several lymphocytes in the background of increased fibers and inflammatory cells (H&E x 400).

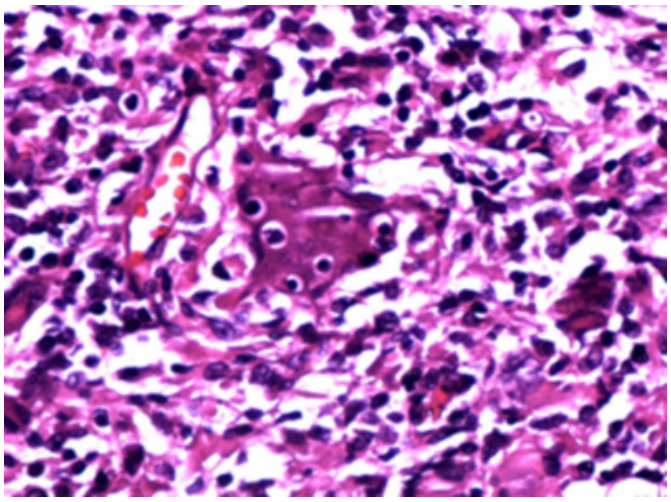
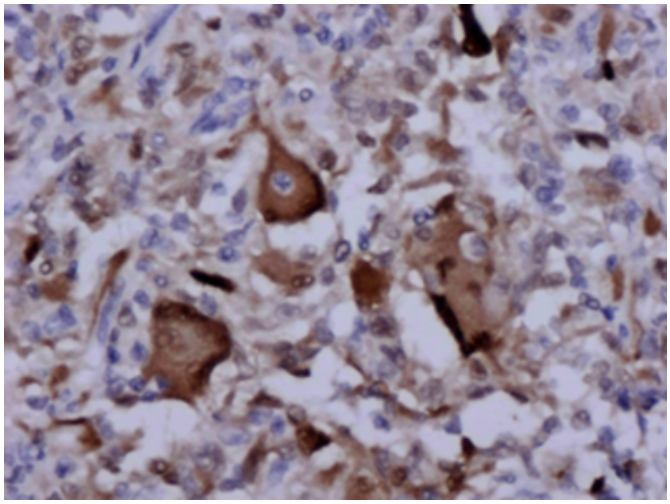


Figure 4

Fig 4 IHC shows histiocytes strongly positive for S100 protein.



DISCUSSION

Rosai-Dorfman disease was first described in 1969 as a systemic non-neoplastic histioproliferative disorder[1]. RDD in the CNS is extremely rare, accounting for < 5% of extranodal cases of RDD[2]. It is a condition of unknown cause that predominantly affects children and young adults and shows a predilection for males[3]. RDD-CNS usually have headaches, seizures, visual symptoms and focal deficits, and the disease course is not usually acute[4-7].

Histology and immunochemistry is necessary for the diagnosis. On microscopic examination, the polymorphic infiltrate consists of histiocytes, lymphocytes, and plasma cells. Emperipolesis, signifying the phagocytosis of lymphocytes, may be more difficult to identify in extranodal sites such as the CNS [8] . Emperipolesis is present in only 70% of cases[9]. The phagocytized cells were often lymphocytes, but plasm cell and erythrocytes could also be occasionally seen. Fibrosis , another feature commonly seen in extranodal sites , often obscured the histiocytic proliferation[2]. It is important for diagnosis that the histiocytes were positive for S100 and CD68 protein but negative for CD1a.

The histological differential diagnosis includes Langerhans cell histiocytosis, inflammatory myofibroblastic tumor, and Lymphoplasmacytic meningioma. It can be differentiated ultrastructurally from Langerhans cell histiocytosis by the absence of Birbeck granules and negativity for CD1a. Inflammatory myofibroblastic tumor (IMT) is composed of myofibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and

eosinophils. Negativity for S100 staining could make a distinction between them. Lymphoplasmacytic meningioma elicits a pronounced chronic inflammatory reaction, which may be confused with the infiltrates of RDD-CNS. Recognition of typical meningioma histology coupled with confirmatory immunostaining for epithelial membrane antigen readily differentiates these two processes.

In summary, the diagnosis of Rosai-Dorfman disease should be considered in any atypical chronic inflammatory lesion with a histiocytic component involving multiple organ systems, especially in the CNS. Immunohistochemical stains for S-100 protein should be performed in such cases.

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