Egyptian Journal of Ear, Nose, Throat and Allied Sciences (2016) 17, 43–46

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Egyptian Journal of Ear, Nose, Throat and Allied Sciences

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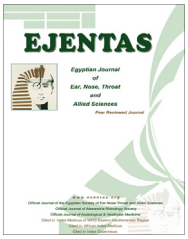

CASE REPORT

Rare case of Rosai Dorfman disease involving paranasal sinuses in paediatric patient: A case report

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Received 17 May 2015; accepted 24 November 2015

KEYWORDS

The Rosai–Dorfman disease;
Histiocytosis;
Cervical node;
Nasal cavity;
Emperipolesis

Abstract Rosai Dorfman disease (RDD) is a rare, benign disease of unknown aetiology. It typically presents with massive, painless cervical lymphadenopathy but may have a varied presentation. We report a paediatric case of RDD with initial isolated involvement of nasal cavity and PNS with subsequent involvement of cervical lymph nodes. Endoscopic biopsy confirmed the pathological diagnosis and he was managed successfully with medical therapy. At 20 month follow up there was no evidence of recurrence. The ideal protocol for the treatment is still debated. A long term follow-up is warranted to detect relapses. This case report is illustrated aiming at developing insights into management and diagnosis of such rare clinical entity in a paediatric patient.

1. Introduction

Rosai Dorfman syndrome is an immune mediated histoproliferative disease described by Rosai and Dorfman in 1969. Other theories like infectious origin have also been suggested however the immunological theory has been the most accepted one.[1]

It commonly affects children and young adults. It is marked by massive cervical lymphadenopathy with a low grade fever. Detailed investigative workup may show anaemia, leucocytosis and polyclonal hypergammaglobulinemia. The enlarged nodes show the proliferation of sinusoidal histiocytes. Approximately 43% of cases show extra nodal involvement wherein the possible sites are skin, central nervous system, ear, and orbit and rarely nose and paranasal sinuses.[2]

Usually the treatment of choice is medical therapy using a cocktail of steroid and cytotoxic drugs as in our case steroid and cyclophosphamide were administered. However in refractory cases or in cases where the adenopathy is causing pressure effects, treatment options like radiation and surgery can also be considered. This entity has an indolent course with a possibility of spontaneous remission even after long periods. Hence these patients require a long term follow up.[3]

2. Case report

An eight year old child presented to the outpatient department with a history of progressive left sided nasal block for the last 4 months with associated intermittent epistaxis for the last

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Peer review under responsibility of Egyptian Society of Ear, Nose, Throat and Allied Sciences.

http://dx.doi.org/10.1016/j.ejenta.2015.11.004

1 month. He also gave a history of bilateral rapidly progressive neck swelling for the last 1 month. He has no other known comorbidities.

Neck examination revealed multiple bilateral cervical lymphadenopathies. Level 2 lymph node on the left side and level 1b on right side were significantly enlarged measuring 5 cm × 5 cm and 1.5 cm × 1.5 cm, respectively. He had no other palpable lymph nodes or organomegaly. Haematological and immunological workups were irrelevant. A diagnostic nasal endoscopy revealed a friable mass involving the left lateral wall and inferior turbinate going up to the choana.

MRCT scan T2 images revealed a 3.4 × 2.7 × 2.4 cm sized T2 isointense lesion in the left nasal cavity (Fig. 1) The mass was involving the left inferior turbinate, lateral nasal wall with bowing of the medial wall of the maxillary sinus (Fig. 2) with posterior extension up to the nasopharynx (Fig. 3). Histopathological examination was consistent with extra nodal sinus Histiocytosis (Figs. 4 and 5) with massive lymphadenopathy (Rosai–Dorfman disease). He was treated successfully with oral steroid and low dose cyclophosphamide and at 20 month follow up no evidence of recurrence or residual disease was noted. Repeat nasal endoscopy showed no lesion in the nasal cavity and there were no palpable cervical lymph nodes.

3. Discussion

Rosai Dorfman syndrome, well known as Sinus Histiocytosis with Massive Lymphadenopathy (SHML), is a rare benign systemic histio-proliferative disease commonly involving cervical group lymph nodes.[1] It was first illustrated by Azoury and Reed and later by Rosai and Dorfman in 1969.[4]

Presently SHML is classified as one of the non-Juvenile Xanthogranulomas (non-JXG) under the sub-group of the non-Langerhans Cell Histiocytosis (non-LCH).[5]

It commonly affects a younger age group, usually in the first and second decades with a male-to-female preponderance of 1.4:1.[6]

Aetiology is still debated, however immune mediated theory or infections caused by Epstein Barr Virus, brucella or klebsiella have been the two most proposed theories.[2]

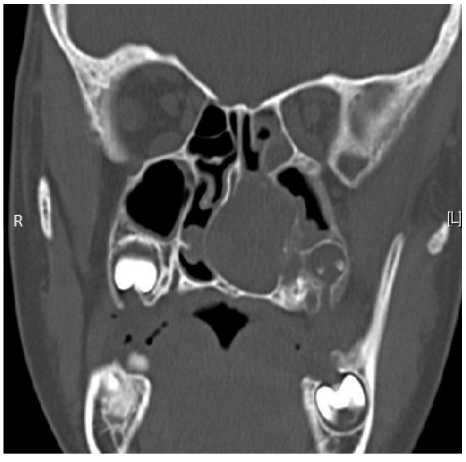

Figure 1 CT scan coronal view.

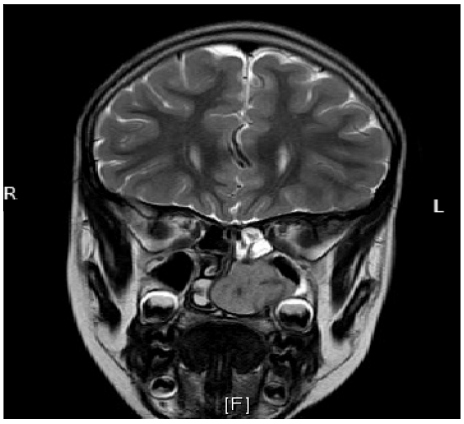

Figure 2 MRI T2 image in coronal view.

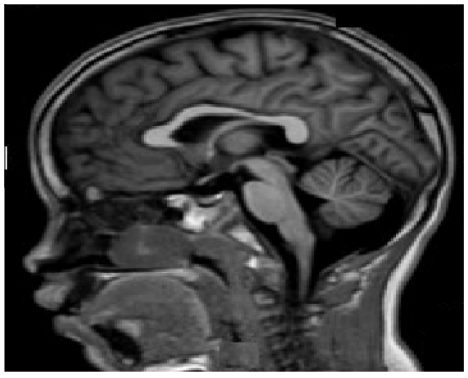

Figure 3 MRI T1 sagittal view.

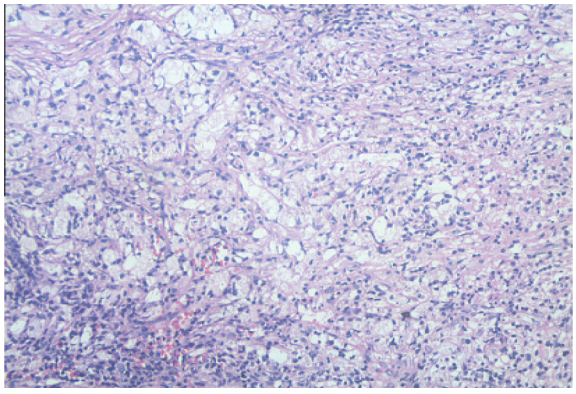

Figure 4 H&E stain – showing cells arranged in solid sheets, with round to oval, irregular, vesicular nuclei, prominent nucleoli and moderate amounts of eosinophilic to vacuolated cytoplasm. The background contains many plasma cells and lymphocytes.

Lymph nodes are usually involved but in about 43% of cases, however extra nodal involvement is also well known. Frequently involved extra nodal sites are skin, mucosa, spinal cord, pancreas, nasal cavity and major salivary glands.[7]

Those affected with this disease present with massive, painless, bilateral or unilateral cervical lymphadenopathy with other nodal sites such as axillary, inguinal, para aortic and mediastinal lymph nodes.[8]