Case Report-III

Rosai-Dorfman Disease : A Case Report With Review of Literature

GULL MOHD BHAT, SURENDER KUMAR, ATUL SHARMA

ABSTRACT

Sinus histiocytosis with massive lymphadenopathy (Rosai Dorfman disease) is an uncommon benign condition, often confused with lymphoma. Patients usually have massive enlargement of cervical lymph nodes. Clinical course is variable. We describe here one such young male patient with tonsillar enlargement. Pertinent literature is reviewed.

INTRODUCTION

Castleman's disease, dermopathic lympha-denitis, mucocutaneous lymph node syndrome (Kawasaki's disease), histiocytic necrotising lymphadenopathy (kikuchi's disease), vascular transformation of lymph nodes and inflammatory pseudotumour of the lymph node are among the rare causes of lymph node enlargement. Sinus histiocytosis with massive lymphadenopathy (SHML) is an important addition to this list. An oncologist comes across a case every now and then with a clinical picture hard to tell from malignant lymphomas. Histopathology usually comes as a surprise. At the same time it is a relief to the physician and the patient, as this disease has essentially a benign course.

Case Report: -

MSA, a 10 years young boy, resident of Bihar was apparently alright about 3-4 months when he noticed a right neck mass, insidious in onset and slowly increasing in size. Similar painless swellings appeared on the other side of the neck, both maxilla and epitroclear areas. The upper neck swellings progressed to attain the size of a walnut. There was accom-panying history of intermittent low grade fever, lasting for a couple of days unaccompanied by chills but usually associated with cough and blood tinged expectoration. He had a single episode of epistaxis; (on follow up visit had repeat episodes of nasal bleed) and had been transfused a unit of blood during this period. There has been no accompanying history of weight loss, night sweats or pruritis. His past, personal, and family history has been normal. Clinical examination showed that his weight was 25 kgs, height 129 cms and had finger clubbing, bilateral tonsillar hypertrophy, a bull neck with multiple, generalized, non-tender, firm, and matted (in cervical region) lymphadenopathy. In other areas lymph nodes varied from 1 to 2 cms in size and (during follow up developed lymph node which increased in size in armpits). His abdominal examination revealed moderate non-tender and soft / firm hepatosplenomegaly (5 & 2 cms below costal margins) with no free fluid. Chest, cardiovascular and central nervous system examination was unremarkable.

Investigations showed a hemoglobin of 8.2 WBC - 8500/cmm, defferential - N 63, E7, L30, platelets 180000/cmm. Peripheral blood film showed microcytic, hypochromic red blood cells. ESR 58 mm/hr. Biochemical profile revealed normal renal and liver functions, with serum alkaline phosphates 225 units and LDH-64 units/ dl. Chest x-ray showed bilateral Hilar and Paratracheal lymphadenopathy.Chest imaging studies revealed mediastinal lymphadenopathy. USG scan of the abdomen

Dept. of Medical Oncology, IRCH
All India Institute of Medical Sciences, New Delhi.
Correspondence to : **GULL MOHD BHAT**
Email : drgull8125@yahoo.com

revealed many peripancreatic and periportal lymphadenopathy. FNAC of left cervical node showed sheets of sinus histiocytes, some of which had evidence of emperopolesis and there was presence of histiocytes and necrosis on the right side.

Lymph node biopsy showed markedly dilated sinuses containing sheets of histiocytes with eosinophilic cytoplasm and small nuclei; some of the histiocytes show small lymphocytes in cytoplasm (emperipolesis). Scanty lymphoid tissue is seen in between. There was no evidence of malignancy or granulomas.These features were consistent with a diagnosis of sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman Disease) (Fig. 1.A & B).

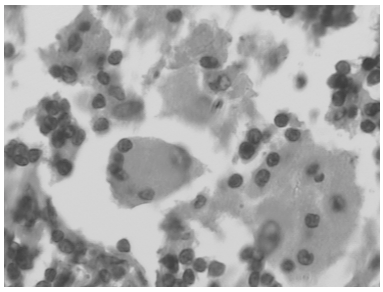

Fig 1. a Low power view (lymph node) showing massive distension of the sinuses by the histocytic infilterate.

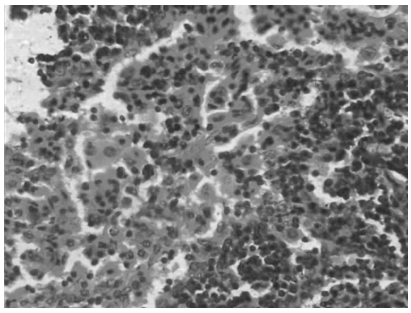

Fig 1. b High power view showing lymphophagocytosis (omperipolesis) by the sinus histiocytes.

Bone marrow exami-nation revealed no evidence of neoplastc infiltration, granulomas or parasitic infections. Patient continued to be symptomatic and had high ESR. Lymph node aspiration was repeated. It reconfirmed the diagnosis. Repeat bone marrow aspiration/biopsy was unremarkable. The child was given a course of antibiotics and planned for brief low dose courses of steroids.

DISCUSSION:

This condition was first described by Robb-Smith in 1947 in children and was termed as giant cell sinus reticulosis . Sinus histiocytosis with massive lymphadenopathy (SHML) has been recognized as a distinct clinicopathological entity, though first given this name by Rosai and Dorfman in 1969,1972. It has morphological features of greatly exaggerated reactive process. There is usually no demonstrable etiological agent. The lymph nodes may attain enormous size and may remain persistently enlarged for a prolonged period. The disease is not confined to a particular race or geographical area, although African Negroes are more commonly involved than whites. Characterized by massive, painless and generally bilateral cervical lymph node enlargement with relatively minor involvement of other groups of nodes in most instances. About 25% of cases have involvement of extra nodal sites, especially the upper respiratory tract, salivary glands, orbit and testis and lesions at other sites and skin may appear before the appearance of lymph nodes.[1] Patients with extensive lung and liver involvement may not have as innocent course.[2] There may be osseous involvement in pediatric age group. Other rare sites included solitary involvement of talus, triquetrum,sclera, conjunctiva, thymus, pleura, nasopharynx, genitourinary and central nervous system. These patients usually have moderate pyrexia with mild anemia, neutrophilia, a raised ESR and polyclonal gammapathy. Autoimmune hemolysis can also occur. Suggestion has been made that it may be an unusual reaction to a klebsiella infection.[3] Association to subsequent malignancies is also described.[4] An infant presenting with hepatosplenomegaly, bicytopenia considered as congenital form of Rosai dorfman desease has been described.[5] The diagnosis is usually made on characteristic histopathological and cytological features. HUMARA assay shows the disease to be monclonal.The cells have histochemical and phenotypic features of macrophages.The cell contain large amount of esterases and acid phosphates and show positivity for CD 68 and alpha-1 antitrypsin.They also exhibit some of the phenotypic features of dendritic cells such as S100, cathepsin E, fascin and at times CD 1a.[6,7] The natural history is that of a regression and

resurgence followed eventually by complete resolution. No form of therapy has been found to be of any benefit. In the Rosai and Dorfman series of 34 patients, one died of renal amyloidosis, another after cytotoxic therapy. The latter form of treatment is contraindicated in this disease. Surgical option may be reserved for compressive symptoms.[8,9] Course in our patient was more protracted / progressive with no abatement of lymph node masses as with about 6 months follow up; rather additional lymph node enlargement appeared in both axill noticed in recent follow up. Dominant nasopharyngeal involvement probably caused recurrent epistaxis.

REFERENCES :

1. *Foucar Eliot and Rosai Juan and Dorfman RF: Sinus histiocytosis and massive lymphadeno-pathy. Ear,nose,throat manifestations., Arch Otolaryngol., 1978,104:687-693.*

2. *Foulard E, Rosa J, Dorman RF. Sinus histocytosis with massive lymphadenopathy (Rosia- Dorfman disease): Review of the entity: Semin Diagn Patho 1990;7:19.*

3. *Lampert F. and Lennert K: Sinus histiocytosis with massive lymphadenopathy. Fifeteen new cases. Cancer, 1976,37:783-789.*

4. *Rat zinger J ,Ziegler Beholding Wet al. Sinus histiocytosis with massive lymphadenopathy. Arch. 2003;443(6):797-800.*

5. *Iwabuchi H, Kakihara T,Tanaka A et al. Congenital Rosai- Dorfman disease without lymphadenopathy. Pediatr Pathol Mol MED 2003 Sep-Oct;22(5) :399-403*

6. *Eisen RN , Buckley PJ, Rosai J: Immunophenotypic characterization of sinus histiocytosis with massive lymphadenopathy (Rosai- Dorfman disease) : Semin Diagn Patho 1990;7:74.*

7. *Jaffe R,Vaughan De.,Langhoff E:Fascin and differential diagnosis of childhood histiocytic lesions. Pediatrol & Dev Pathol 1998;1:216.*

8. *Dodson KM, Powers CN, Reiter ER, Rosai Dorfman disease presenting as synchronous nasal and intra-cranial mass Am J Otolaryngol 2003Nov-Dec;24(6),USA.*

9. *Raveenthiran V,Dhanalakashmi M,Hayavadana Rao,Vushwanath Rosai-Dorfman disease:report of a 3-year-old girl with critical review of treatment options. Eur J Pediatr Surg. Oct; 2003;13(5):350-4.*