http://www.ijp.iranpath.org/

Bilateral Cervical Lymphadenopathy - Need to Think Beyond Tuberculosis

Rupali Malik[1], Nisha Rana[2]

1. Department of Internal Medicine, VMMC and Safdarjang Hospital, New Delhi-110029, India

2. Department of Pathology, VMMC and Safdarjang Hospital, New Delhi-110029, India

Corresponding author and reprints: Dr Rupali Malik, MD, Department of Internal Medicine, VMMC and Safdarjang Hospital, New Delhi-110029, India, Email: drvickyster@gmail.com, Phone Number: 09711143397

Received: 2015 Jan 14 - Accepted: 2016 Apr 07

Dear Editor-in-Chief

Rosai-Dorfman Disease (RDD) or sinus histiocytosis with massive lymhadenopathy (SHML) is a rare, benign proliferative disorder of histiocytes characterized by painless bilateral cervical lymph node enlargement though extranodal disease involving skin, respiratory system, central nervous system (CNS) etc may been in > two third of cases (1-3). With lack of widespread awareness about this entity, misdiagnosis of RDD as more common ones such as tuberculosis is a common phenomenon. Therefore its revision would seem worthwhile with this report.

A nineteen year old boy presented with multiple cervical and submandibular lymphadenopathy of three months duration preceded by low grade fever of 7 days. There was no history of nodal pain, persistent fever, weight loss, night sweats, headache, seizures, cough, difficulty breathing, koch's contact or any symptoms related to ear, nose or throat. Clinical examination showed multiple, enlarged, non matted, non-tender, discrete, mobile bilateral, cervical and submandibular lymph nodes ranging in size from 1.5 x 1 cm to 2 x 1 cm (Fig. 1). Investigations showed hemoglobin 14 gm/dl, total leucocyte count 12,000/cm with neutrophilia (79%), platelet count 2.16 lacs/mm and erythrocyte sedimentation rate of 40 mm. Other investigations included negative retroviral serology, negative mantoux test and normal ultrasonography of abdomen/x-rays of chest and nasal sinuses.

Fine needle aspiration cytology (FNAC) of cervical lymph nodes revealed presence of numerous histiocytes throughout the smears with abundant pale cytoplasm, single to multilobated or multiple nuclei with fine chromatin and inconspicuous to prominent nucleoli but no nuclear atypia or nuclear grooving (Fig. 1). These cells (histiocytes) were characterised by presence of lymphocytes and neutrophils in the cytoplasm suggestive of emperipolesis. Plasma cells, lymphocytes, and eosinophils could be seen in the surroundings. A diagnosis of Rosai- Dorfman disease was subsequently considered based on the histopathology. Later, excisional cervical lymph node biopsy was done that revealed fibrous thickening of lymph node capsule and prominent dilatation of lymph sinuses resulting in partial architecture effacement. The sinuses were packed with numerous histiocytes having abundant pale cytoplasm and phagocytosed lymphocytes (emperipolesis), plasma cells and macrophages (Fig. 1). The patient was kept under conservative management only with decrease in size of lymph nodes at the end of 6 months without any clinical deterioration.

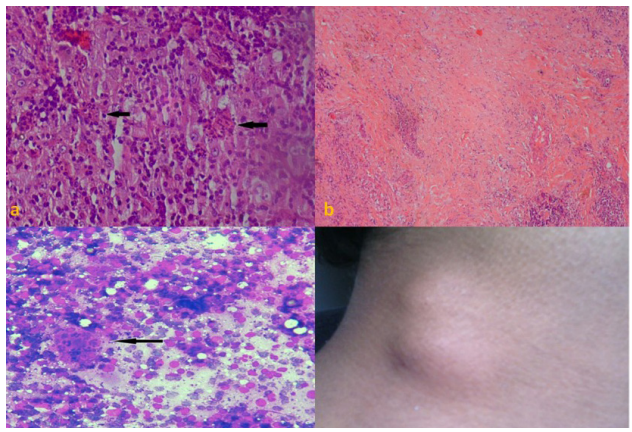

Fig. 1
A. Lymph Node Biopsy showing presence of numerous histiocytes with neutrophilic emperipolesis (arrows) {HE, 200X}. B. Areas in the Lymph node showing fibrosis (HE,200X). C. Cytology smear display presence of reactive lymphoid cells many histiocytes with neutrophilic emperipolesis (arrows). D. Clinical photograph showing right posterior cervical lymphadenopathy.

In a case with cervical lymphadenopathy, as in ours, tubercular lymphadenitis is usually the first entity that should be excluded. The possibility of Rosai-Dorfman disease, considering its rarity, was not considered in our case until FNAC was performed. The cytological features usually reveal numerous large histiocytes with phagocytosed lymphocytes (emperipolesis) as seen in our case (2, 4). Emperipolesis is the presence of intact cells (e.g. lymphocytes) located within cytoplasmic vacuoles in the cytoplasm of histiocytes (5). On electron microscopy, histiocytes are devoid of Birbeck granules and immunostaining reveals positivity for S100 and CD33/CD68 antigens but not for CD1a (4). The prognosis is excellent in most cases with spontaneous regression after conservative management only.

In conclusion, both clinicians and pathologists should be aware of entity of RDD in making a differential diagnosis of cervical lymphadenopathy even in tropical countries with widespread tuberculous infection.

Acknowledgments

The authors declare no conflict of interest.

References

1. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy: A newly recognised benign clinicopathological entity. Arch Pathol 1969;87:63-70

2. Chandrashekhara SH, Manjunatha YC, Muzumder S, Bahl A, Das P, Suri V et al. Multicentric sinus histicytosis (Rosai-Dorfman Disease): Computed tomography, magnetic resonance imaging findings. Indian J Med Paediatr Oncol 2011; 32:174-6

3. Foucar E, Rosai J, Dorfman R. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman Disease): Review of the entity. Semin Diagn Pathol 1990; 7:19-73.

4. Jani PA, Banjan D. A case of Sinus Histiocytosis with Massive Lymphadenopathy (Rosai- Dorfman Syndrome) from Western India. Mcgill J Med 2008 11(2):156-59.

5. Foucar E, Rosai J, Dorfman RE. Sinus Histiocytosis with massive lymphadenopathy: an analysis of 14 deaths occurring in a patient registry. Cancer 1984; 54: 1834-40.

How to cite this article:
Malik R, Rana N. Bilateral Cervical Lymphadenopathy- Need to Think Beyond Tuberculosis. Iran J Pathol. 2016; 11(3):298-300.