A Rare Case of “Rosai–Dorfman Disease”

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Abstract Sinus histiocytosis with massive lymphadenopathy (SHML) is a benign proliferating histiocytic disorder, predominantly of lymphnodes with extranodal involvement also seen. We present a case of 35 years old female with history of multiple swellings in neck since 1 month duration. On examination patient had painless bilateral cervical lymphadenopathy. No other ENT manifestations noted. Lymphnode biopsy revealed SHML. Abdominal scan and chest X-ray was done which was normal. This case report highlights the clinical, histological aspects of SHML, Rosai–Dorfman disease.

Keywords Rosai–Dorfman disease ·
Cervical lymphadenopathy · Sinus histiocytosis

Introduction

SHML introduced in 1969 by Rosai and Dorfman [1] is a distinct benign pseudolymphomatous entity with characteristic histologic features [2, 3]. Most common site for lymphadenopathy is neck. Mediastinal, axillary, inguinal and retroperitoneal nodes may be affected [2]. The lymphadenopathy when present, tends to be painless and bilateral. Extranodal tissue involvement occurs in 25–40 % of cases and may involve bone, testes, orbit, eyelid, upper respiratory tract, salivary glands, skin, lungs, adnexa,

kidney, central nervous system, peritoneum, thyroid, small intestine and joints.

Extranodal involvement appears to be more common in patients with immunologic abnormalities which may themselves contribute to poorer prognosis [5].

The disease is characterized in majority of cases by painless bilateral lymphnode enlargement in the neck, often associated with fever, leucocytosis and polyclonal hypergammaglobulinemia. It presents mainly in the 1st or 2nd decades of life but any age group can be affected especially in extranodal form. SHML should be kept in mind for differential diagnosis of neck masses especially in childhood.

Although the geographic distribution is widespread, there has been a high incidence of the disease amongst black African races.

Immunohistochemically, the sinus histiocytes are strongly positive for S-100 protein. Immunophenotypic profile suggests, origin from macrophage histiocyte cell types [8]. The aetiology of the disease include a possible viral infection (Epstein barr virus) and/or an undefined immunological alteration (Table 1).

Evaluation of the disease is somewhat variable from complete spontaneous remission in some cases, to a protracted clinical course for years in others with the possibility of involvement of vital organs and death.

Case Report

History

Thirty-five years old female patient presented to ENT opd on February 2011 with history of bilateral multiple bilateral

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Fig. 1 Pre operative photograph showing massive bilateral cervical lymphadenopathy. Note that burnt scars are seen as the patient consulted traditional healer

painless swellings in neck of one month duration (Fig. 1). Patient also complained of fever of moderate degree and dry cough since 15 days. Examination of neck revealed on left side revealed enlarged lymphnodes, level 1b; of 5×5 cm, level 5; 3×3 cm, on right side level 1B; of 5×3 cm, level 2; 5×7 cm level 5; of 4×3 cm, level 5; 3×3 cm All the lymphnodes are found to be firm, discrete, non tender and mobile.

Examination of ear, nose, oral cavity and oropharynx and larynx revealed normal findings. General physical examination was done and no other group of lymphnodes was found to be enlarged.

Investigations

Blood examination revealed Hb-13.1 g/dl, RBC- $4.76 \times 10^6/\text{mm}^3$, TLC 9,300 cell/cu mm, ESR-52 mm/h (raised), differential count revealed N76 %, L17 %, E04 %, M3 % Peripheral blood picture revealed normocytic hypochromic anaemia with relative neutrophilia.

FNAC showed polymorphous population of lymphocytes, histiocytes, plasmacells, plump histiocytes and few

atypical giant cells against background of fibrinous tissue. USG neck showed bilateral discrete lymphoechoic areas suggestive of lymphoma. Chest X-ray and abdominal scan was normal.

Biopsy of right post triangle lymphnode was done.

Histopathology

Gross appearance was a well circumscribed soft tissue mass of 1.5×1.5 cm with cut section grey white homogenous. Microscopy revealed dilated sinuses filled with many histiocytes. Some of the histiocytes having engulfed lymphocytes (emperipolesis) with plasma cell infiltration within the dilated sinus (Fig. 2a, b).

Histopathological diagnosis of SHML was made. Patient is advised T. Wysolone- 60 mg for 2 weeks in divided doses and is on follow up.

Discussion

SHML is regarded as a benign, pseudolymphomatous disorder of unknown cause [2]. Clinically, the disease is characterized by an often protracted although benign course [1–3]. spontaneous remissions and relapses have also been noted [10]. Recent reports document that infiltrates of SHML can cause death [2–7]. Foucar et al. [9, 10] attribute fatalities associated with SHML to related immunologic disorders. Histiocytes expressed S-100 protein by immunohistochemistry, S-100 protein is found in cytoplasm of many cells of neuroectodermal origin as well as subtypes from the macrophage/histiocyte family which can be demonstrated by immunoperoxidase staining of antibodies directed against S-100 protein. The presence of S-100 protein positive histiocytes that demonstrate emperipolesis is regarded as diagnostic of SHML.

The differential diagnosis of SHML includes Wegener's granulomatosis, midline malignant reticulosis, eosinophil granuloma, Hodgkins disease and fibro inflammatory lesions. Although the extranodal involvement occurs in 40 % of cases painless bilateral cervical Lymphadenopathy is the commonest presentation. Treatment may not be required in majority of patients with SHML since the disease does not usually threaten life or organ function and is often self limited and subject to spontaneous regression.

Conclusion

This paper highlights the clinical, histologic aspects of SHML, Rosai–Dorfman's disease and emphasizes on the

Table 1 Commonest sites of SHML in the head and neck area

Nasal cavity and paranasal sinuses
Orbit
Parotid and submandibular gland
Larynx
Nasopharynx
Temporal bone
Infra temporal fossa
Meninges

Fig. 2 **a** Section showing H&E stain under $\times 10$ Magnification—thick capsule with dilated sinuses filled with proliferating histiocytes. Plasma cell infiltration seen in dilated sinuses. **b** Section showing H&E stain under $\times 40$ Magnification—characteristic emperipolesis is seen

point that Rosai–Dorfman’s disease should be considered as one of the differential diagnosis when a patient comes with painless bilateral Lymphadenopathy of neck. Also highlights the commonest presentation of SHML. As the etiology still remains obscure and there is no pre set treatment guidelines, further work is needed.

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