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Case Report

Dermatopathic-Lymphadenitis – Two Case Reports

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Abstract

Dermatopathic lymphadenitis is a relatively rare non-neoplastic lymphnode lesion usually secondary to generalized dermatitis and primarily involves the axillary group of lymph-nodes. The lesion represents a T-cell response to skin antigens processed and by interdigitating dendritic cells.

Dermatopathic lymphadenitis represents a diagostic challenge in routine practice because of its close resemblance to various lesions, namely Langerhan's histiocytosis in children and mycosis fungoides, Hodgkin's lymphoma and monomorphic leukemia in adults with non-specific clinical features and in the absence of any associated clinical skin disorders.

Keywords

Dermatopathic lymphadenitis, Mycosis-fungoides

Introduction

Dermatopathic lymphadenopathy is a particular type of paracortical hyperplasia with predominance of dendritic cells. This disorder is associated with a variety of skin disorders, but rarely can occur without any skin involvement, as in our case.

Case Report 1 : A 38-year old male presented with fever and gradual, pinless posterior cervical lymph node enlargement since last 2 months. There was no associated chill and rigor or evening-rise of temperature. On examination, mild hepatomegaly was detected and a clinical diagnosis of tuberculous lymphadenitis was made. Routine blood examination showed normocytic, normochromic blood picture with eosinophilia and absolute eosinophil count (AEC) showing 11,000 cells/cumm. FNAC showed features of reactive lymphadenitis. Excision biopsy was done and material sent for histopathological study. Grossly the specimen consists of grey brown soft tissue bits measuring 4 x 1 x 0.5 cm. Cut section was grey-brown. Microscopy showed features suggestive of dermatopathic lymphadenitis with advice for immunohistochemistry studies for confirmation.

Case Report 2 : A 35-year old female presented with fever and gradual painless enlargement of posterior cervical lymph nodes. Fever was associated with cough and headache and a clinical diagnosis of respiratory tract infection was made with a course of antibiotics administered by the local doctor. However, the fever did not subside and the lymph nodes became enlarged and painful. Subsequently the patient was referred to our hospital for further investigation and treatment. Peripheral blood smear showed relative lymphocytosis and FNAC was suggestive of reactive lymphadenitis with advice for biopsy for confirmation of the diagnosis. The biopsy was done and sent for histopathological study. Grossly, the specimen consists of grey brown soft tissue bits measuring 3 x 2 x 1 cm. Cut section was grey-brown. Microscopy showed features suggestive of dermatopathic lymphadenitis with advice for immunohistochemistry studies for confirmation.

Discussion

Dermatopathic lymphadenitis was first described by Pauterir and Woring in the year 1932[1]. It is a reactive condition where lymph node enlargement is associated with

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a variety of skin disorder ranging from chronic dermatosis, generalised exfoliative dermatitis, psoriasis to T-cell lymphoma of skin such as Sezary- syndrome and Mycosis Fungoides[2]. Rarely dermatopathic lymphadenitis are seen in the absence of clinical skin disorders[3]. Persons of 5th to 7th decade of age are mainly involved.

A study conducted by Dowsen and Cooper revealed that of 906 consecutive lymph node biopsies dermatopathic lymphadenitis occurred in 40 cases, thus accounting for only 4.8% of the cases[4]. Axillary and inguinal lymph nodes are predominantly involved, although generalized superficial lymphadenopathy can also occur. Males are mainly affected with a M:F ratio of 3:1[4]. The duration of associated skin disorders can be variable ranging from 6 months to 6 years[5]. However, Dawson and Cooper study showed that 12% of the cases had no prior skin diseases. Eosinophilia is present in 35% of the cases with pruritus being present in many cases[5]. Of the 40 biopsies only 9 (22.5%) cases of dermatopathic lymphadenitis are associated with mycosis fungoides[3]. Two cases presented as breast masses due to involvement of the external mammary lymph nodes[6].

Mostly melanin pigment is present but sometimes haemosiderin laden macrophages are also seen. Lipid can be revealed in frozen sections by Oil red O and sudan black stain. Melanin being a reducing substance the macrophages show Schmorl-Masson Fontana positivity. Due to the presence of both lipid and melanin it is also termed as 'lipomelanic reticulosis'[2]. This term has been discarded because it may be mistaken for a neoplsatic process.

Grossly, the lymph nodes are enlarged and firm in consistency. The cut-section is bulging with occasional brownish pigmentation, mimicking malignant melanoma. Microscopically, the affected lymph nodes show follicular-hyperplasia with sinus histiocytosis. The paracortex is pale because of the proliferation of the interdigitating reticulum cells along with variable number of immunoblasts, eosinophils and plasma cells. Under higher magnification the reticulum cells appear to have folded nuclei, finely dispersed nuclear chromatin and a prominent nucleolus. Scattered amongst the reticulum cells are small lymphocytes. The ultrastructural features are best seen under electron microscopy which highlight the irregular nuclei along with abundant cytoplasm forming complex interdigitations along with many organelles such as golgi

Due to strong association of dermatopathic lymphadenitis with Sezary syndrome it is important to know the exact origin of the T-cells[7]. The T-cells may represent : 1) reactive local proliferation of T-cells, 2) drainage of T- lymphocytes from the respective skin lesions, 3) circulation of increased number of T-cells through the paracortex, 4) early nodal proliferation of neoplastic T- cells.

One of the important diagnostic clues in establishing whether there is an associated T- cell lymphoma is to look for not only significant number of small irregular T-lymphocytes arranged in clusters but also to look for the presence of larger blast forms of convoluted T-cells[8]. Nodes affected by dermatopathic lymphadentis may be confused with Hodgkin's disease, mycosis fungoides, monocytic leukemia and Langerhan's histiocytosis[9]. Mycosis fungoides will show characteristic cells with deeply indented "cerebriform nuclei" replacing areas of nodal parenchyma and Hodgkin's disease will have numerous plasma cells, eosinophils, Reed-Sternberg cells along with neoplastic mononuclear cells-Hodgkin's cells.

Immunohistochemistry helps us in ruling out mycosis fungoides, as nodes involved by mycosis fungoides show loss of CD-7 or CD-62L expression along with loss of pan T-cells markers such as CD-5, CD-3 and CD-2[10].

In our study both the cases showed certain unique features, namely, 1) Posterior cervical, lymphnode involvement in contrast to the usual axillary involvement, 2) No associated clinical skin disorders, 3) Presence of non-specific clinical symptoms giving rise to diagnostic confusion, and 4) Relatively younger age of the patients involved. The first case of our study was associated with peripheral eosinophilia. The second case showed features of respiratory tract infection. FNAC was done for both the cases and showed features of reactive lymphadenitis.

Our study shows that although FNAC is indicated to confirm dermatopathic lymphadenitis, but the results are inconclusive. Excision biopsy is the best way for confirmation of the diagnosis along with immunohistochemical studies to rule out mycosis fungoides/ Sezary syndrome and malignant melanoma. Special stains done by us including Fontana silver stains showed large macrophages containing abundant melanin pigment. Immunohistochemical studies done by us showed S- 100 positivity for both the interdigitating dendritic