Rosai-Dorfman Disease Presenting with Multiple Contiguous Lymphadenopathy: A case report

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INTRODUCTION

Sinus histiocytosis with massive lymphadenopathy (SHML) also known as Rosai-Dorfman disease (RDD), is a rare entity of lymph nodes and was first described by Rosai and Dorfman in 1969(1). The disease affects predominantly the younger age group. Painless lymphadenopathy is the most frequent presenting symptom and involves the cervical lymph node in up to 90% of patients, which is typically bilateral and self-limiting. Extra-nodal involvement is documented in 43% of patients. It is a benign proliferative histiocytic disorder morphologically characterized by 'emperipolesis' (1).

CASE PRESENTATION

A 47-year-old man presented with an 8-month history of progressive multiple lymphadenopathies including bilateral cervical neck, bilateral axillary and bilateral inguinal lymph nodes without, fever and weight loss.

Computed tomography (CT) of the cranium, neck, chest, and whole abdomen demonstrated negative intracranial findings, multiple minimally enhancing enlarged lymph nodes diffusely scattered in the cervical region (IA, IB, IIA, IIB, III, IV, VA, VB, and VI) with short axis diameter of 1.9cm in the right and 1.8 cm in the left, bilateral axillary (0.6 to 1.6 cm in widest short axis diameter), abdominal para-aortic space (0.5 to 1.5 cm in widest short axis diameter) and bilateral inguinal areas with size ranging from 0.5 to 1.5 cm in widest short axis diameter

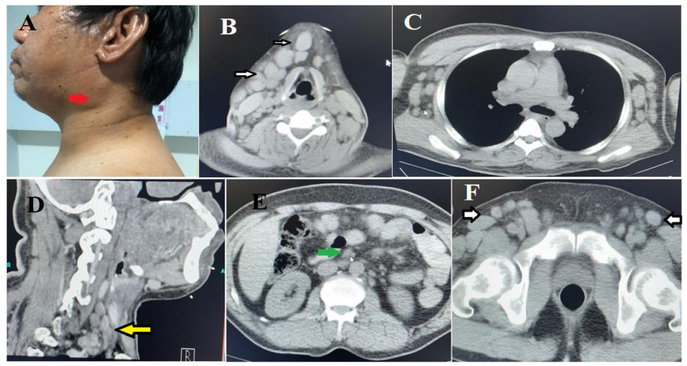

Figure 1. Clinical feature and Computed tomography. (A) Multiple cervical lymphadenopathies on the right side, (B, D) CT scan, Submental and Medial jugular lymphadenopathy, (C) Bilateral axillary lymphadenopathy, (E) Intra-abdominal para-aortic lymphadenopathy, (F) Bilateral inguinal lymphadenopathy.

Histopathological analysis revealed histiocytes containing phagocytosed cells (emperipolesis) and few eosinophils were observed. The less affected parts of the lymph node displayed prominent reactive inflammatory cells (Figure 2).

The immunohistochemical staining was positive for CD3, CD4, CD19, CD20, CD 45, S100, CD68 and negative for CD1a, thus compatible with RDD (Figure 2C & 2D).

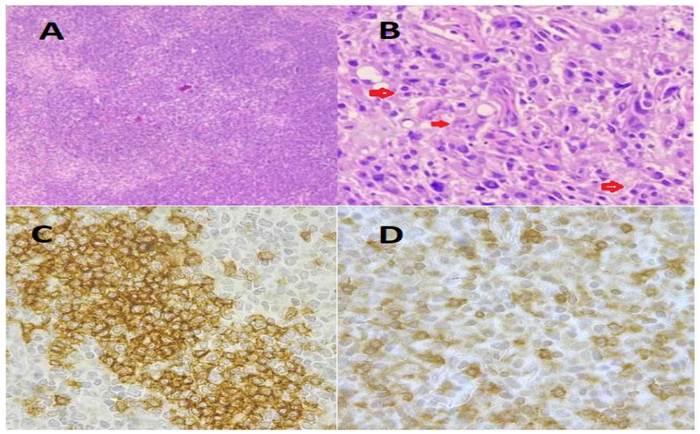

Figure 2. (A) Low-power HE stain showing atypical cells arranged in sheets infiltrating the lymphoid tissue stroma, (B) High-power (400x) HE stain showing characteristic cytomorphology of the histiocytic cells, with abundant finely granular pale acidophilic cytoplasm (lymphocytes within the cytoplasm, or emperipolesis (red arrows), characteristic of Rosai-Dorfman disease), (C) Positive S100 immunostain, (D) Positive CD68 immunostain

The patient has no evidence of airway compromise, extranodal involvement nor distant metastasis. Hence, a conservative approach (watch and wait) was adopted

DISCUSSION

The diagnosis of RDD is based on the clinical history and confirmed by histopathological examination.

The most important histological finding in RDD is emperipolesis, defined as the presence of intact lymphocytes inside other cells, in this case, histiocytes. The most useful marker of histiocytes in RDD is the expression of protein S100 and CD1a negativity (3,5).

Earlier reports suggest a possible link in the pathogenesis of RDD and lymphoma. Furthermore, RDD could be a middle step between a truly benign process and malignancy because, not only other cases of lymphoma transformed after RDD have been reported, but also extra nodal cancer following extra nodal presentation of the disease.

CONCLUSION

RDD is infrequently suspected clinically. Although many clues point to its involvement in the process of carcinogenesis, more clinical and molecular evidence are necessary to tie the link between RDD and cancer. Awareness of clinical profile and consideration in the differential diagnosis in the evaluation of histiocytic and lymphocytic pathologies is essential for proper interpretation, diagnosis, management, and surveillance

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