Clinical Image

Cardiac Involvement of Rosai-Dorfman Disease: Causing Aesthetic Problem in a Young Woman

Running Title: Rosai-Dorfman Disease

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A 32- year-old woman was referred to our ENT clinics for the evaluation of multiple symmetrical swellings in neck. In the patient's family history, there was cross-cousin marriage in between her grandparents. The patient and her sister had a severe hearing loss and type 1 diabetes mellitus since their childhood. Also, she had symmetric hyperpigmented and erythema nodosum-like lesions on the anterior surface of the lower extremities which would suggest the patient had an immune system disorder (Figure 1a). The head and neck examination revealed bilateral symmetrical firm, painless and fairly mobile periparotid and submandibulary masses (Figure 1b). Magnetic resonance imaging (MRI) neck scans revealed conglomerated lymph nodes in size of 45x27 mm on the right, 37x22 mm on the left parotid region and also solid lymph node in size of 26x18 mm on the right and 21 x 18 on the left submandibular region. The patient's blood sample tests CRP and ESR were high, while capillary protein electrophoresis showed polyclonal hypergammaglobulinemia. Anti-thyroglobulin, anti-thyroid peroxidase, and anti-nuclear antigen were also positive. The patient's Cardiac MRI was reported that there was a uniform contoured mass lesion which is adherent to the interventricular septum with a protrusion through the right ventricular lumen, in the size of 10x15 mm. Compared with the myocardium, it had a hyperintense appearance in the T2 weighted series and a homogenously enhancing mass lesion after IV contrast material injection (Figure 2). Besides, similar mass lesions were observed in the anterior mediastinum. The masses on the neck did not cause any problems other than cosmetic appearance, so excisional biopsy from the parotid and submandibular regions was performed for diagnosis and cosmetic reconstruction. The histopathological evaluation showed that normal layout of the lymph node was impaired due to the marked enlargement of the lymph sinuses also there are numerous lymphocytes, plasma cells and large vesicular nuclei histiocytes within. Most of these histiocytes have intact lymphocytes and plasma cells in their cytoplasm, which is significant for the diagnosis of Rosai Dorfman Disease (RDD), called as "emperipolesis" or "lymphocytophagocytosis" (Figure 3a & 3b). During 24 months follow-up period, the size of the mediastinal and intracardiac masses did not change and the skin lesions were not activated. For this report the informed written consent was taken from the patient.

Sinus histiocytosis with massive lymphadenopathy, known as Rosai-Dorfman disease (RDD), is a rare, non-malignant histiocytic proliferative disorder. Altough the proliferation of the histiocytes presents in the pathogenesis, the etiology is unknown and classically it presents with bilateral, massive, painless cervical lymphadenopathy, but approximately 40% of cases have extranodal involvement. Extranodal RDD mostly involves the skin, nasal cavity and paranasal sinuses, orbita, upper respiratory tract, and bone (1,2). Cardiac involvement is very rare and occurs in

less than 0.1% of cases (3). The lesion defined in the heart and mediastinum had a signal intensity similar as seen in Daruwalla et al.'s case (4). Although the prognosis for RDD is usually good, fatal consequences had to be take into account with the involvement of vital organs such as the heart and mediastinum. Therefore multidisciplinary researches should be conducted thoroughly.

References

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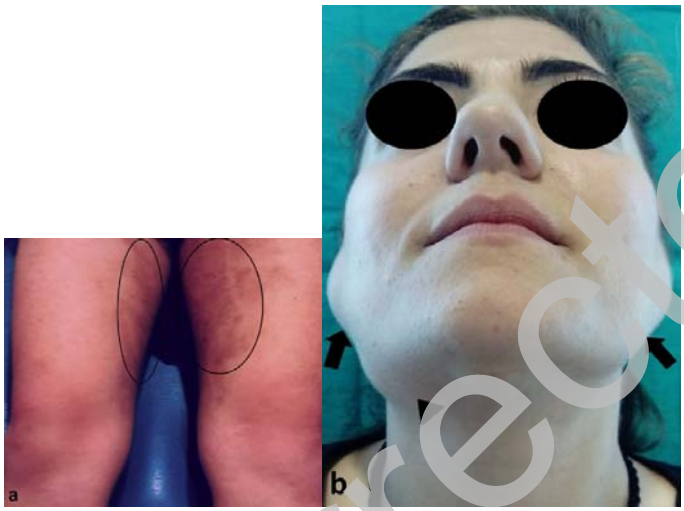

FIG. 1. a) The colored skin lesions on her lower extremities. b) The preoperative view of the patient with the right submandibular (black arrow head) and bilateral parotid region masses (black arrows).

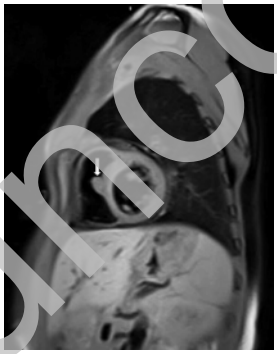

FIG. 2. Sagittal section with T2 sequences of Cardiac MR scan indicates a 10x15 mm mass in the cardiac interventricular septum (white arrow).

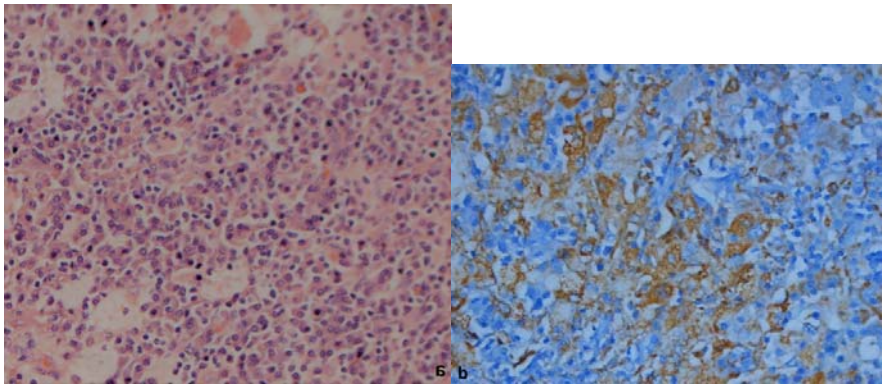

FIG.3. a) Hematoxylin and eosin staining of the specimen which revealed diffuse aggregats of histiocytoid cells with neutrophils, plasma cells and lymphocytes (HEx100) b)Emperipolesis of lymphocytes in histiocytes with positivity for immunstain S-100 (x400).