

Multilocular Thymic Cyst in a Patient with Diffuse Infiltrative Lymphocytosis Syndrome

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ABSTRACT

Diffuse Infiltrative Lymphocytosis Syndrome (DILS) is a multisystem disorder characterised by bilateral salivary gland enlargement and/or xerostomia in patients with Human Immunodeficiency Virus (HIV) infection. A 16-year-old young male patient, presented with bilateral painless parotid swelling since five years. On evaluation, patient was found to have oral thrush and bilateral cervical lymphadenopathy. He turned out to be positive for HIV with a cluster of differentiation 4 (CD4) count of 237/mcl. On Positron Emission Tomography- Contrast Tomography (PET-CT), a multiloculated thymic cyst (13×9×6 cm) was noted. He was diagnosed to have DILS with a large multiloculated thymic cyst. The patient was managed with tenofovir, lamivudine and efavirenz along with cotrimoxazole prophylaxis and fluconazole and was followed upto nine months for the regress of the conditions. The case highlights the rare association between DILS and thymic cyst.

Keywords: Human immunodeficiency virus, Parotid, Thymus multisystem disorders

CASE REPORT

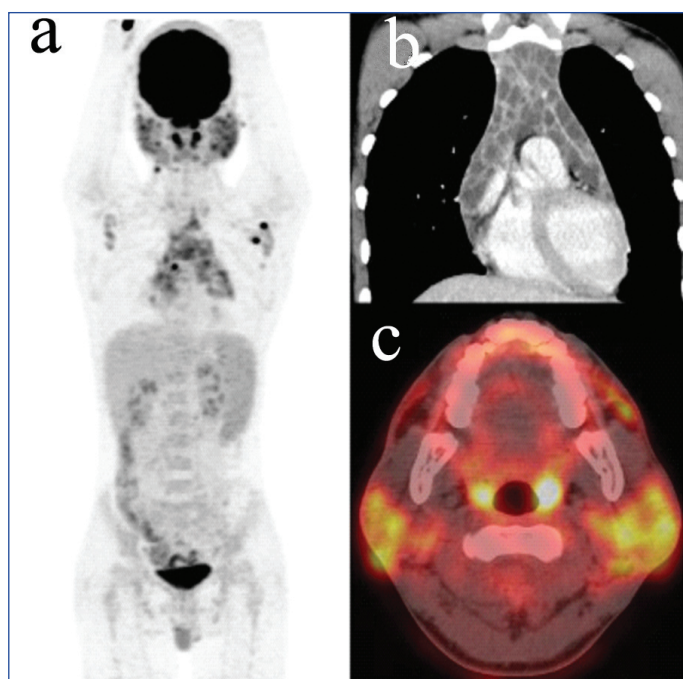
A 16-year-old male patient presented with bilateral painless parotid and submandibular swelling, slowly increasing in size, since five years. He did not have any history of fever, loss of appetite or loss of weight. His father died early in his childhood and his mother had a history of prolonged illness after which she moved out of the house. He was currently living with his aunt. He had no history of any high-risk behaviour.

On examination, he was found to have whitish patches involving the oral mucosa and palate (candidiasis), enlarged parotid glands of both sides and bilateral cervical lymphadenopathy. The parotid gland openings were not visualised.

His routine haematological and biochemical parameters were normal except for relative lymphocytosis. Ultrasonography of the neck revealed cervical lymphadenitis with cystic enlargement of bilateral parotid and submandibular glands. He was found to be positive for HIV-1 by serology. His CD4 count was found to be 237/mcl (6%). HIV was initially suspected as he had lymphadenopathy with oral thrush. Also, since both his parents had a history of prolonged illness, vertical transmission was suspected. A biopsy of the left supraclavicular lymph node was done which showed lymphocytic infiltration without any granulomas and was negative for bacterial, fungal (for endemic mycoses) and mycobacterial cultures. Auto-immune profile and Schirmer's test were negative. On Positron Emission Tomography-Contrast Tomography (PET-CT), a multiloculated thymic cyst (13×9×6 cm) was noted [Table/Fig-1].

The patient was diagnosed with DILS based on the clinical, radiological and histopathological findings. He was initiated on tenofovir, lamivudine and efavirenz along with cotrimoxazole prophylaxis and fluconazole (for oral candidiasis). No active intervention was planned for the thymic cyst as it was asymptomatic and is known to regress spontaneously.

His oral candidiasis improved within a week. Patient did not report any fever on follow-up after three months. His neck swellings had significantly reduced on follow-up after nine months. Repeat imaging could not be performed because of financial constraints. The aunt was counselled about the importance of adherence to medications. The patient was asked to follow-up in the local antiretroviral therapy centre.



[Table/Fig-1]: (a) Whole body PET showing metabolically active cysts in thymus and parotid along with right cervical, right internal mammary and left axillary lymphadenopathy; (b) Coronal section of thorax showing a multiloculated cystic lesion in anterior mediastinum suggestive of a thymic cyst; (c) Transverse section showing metabolically active cysts in the parotid gland.

DISCUSSION

The Diffuse Infiltrative Lymphocytosis Syndrome (DILS) is a multisystem disorder characterised by bilateral salivary gland enlargement and/or xerostomia for at least six months in individuals with HIV infection [1]. The diagnosis requires histologic confirmation of lymphocytic infiltration without any evidence of granuloma or neoplasia [1]. DILS can be associated with the involvement of other organs such as lungs, liver, kidney etc. Multilocular thymic cyst as a part of DILS is an extremely rare entity [1-5]. Parotid enlargement and lymphocytic interstitial pneumonia were concurrently present in some of the reported cases [4]. These cysts are usually detected as an incidental finding on radiology [4]. They are located in the anterior mediastinum and can grow up to a maximum of 17-20 centimetres [3]. Their histopathology resembles that of benign lymphoepithelial cysts [6].

Since the patient had HIV, bilateral parotid enlargement and bilateral cervical lymphadenopathy, the following differential diagnoses were kept: tuberculosis, lymphoma, Sjogren's syndrome and sarcoidosis [7-10]. However, the biopsy of the lymph node showed lymphocytic infiltration without any granulomas, which decreased the possibility of tuberculosis. The negative auto-immune profile and Schirmer's test pointed against Sjogren's syndrome [11-13]. To evaluate for sarcoidosis and lymphoma, a PET-CT was done, which showed a multiloculated thymic cyst. Based on the clinical presentation, lymphocytic infiltration and the thymic cyst, a diagnosis of DILS was made.

A thymic cyst is an important differential diagnosis of patients presenting with an anterior mediastinal mass. Thymic cysts may be congenital or acquired. While congenital thymic cysts are usually small and unilocular, acquired cysts are large and multilocular [6]. Similar to this patient, acquired thymic cysts as a part of DILS have been described in young patients with HIV with a slight male preponderance [14]. The clinical course of this DILS has been described as relatively slow, and most cases show an eventual reduction in thymic size. In the series by Kontny HU et al., the size of cyst reduced in five of the eight patients without any surgery while two of the patients showed complete resolution [14]. In asymptomatic HIV positive patients with characteristic cysts and no features suggestive of malignancy, active intervention may not be required. Surgical intervention may be warranted in those with large symptomatic masses [14].

CONCLUSION(S)

The report presented a newly diagnosed HIV patient with bilateral parotid and submandibular swelling. The clinical presentation was initially thought to be due to opportunistic infection or malignancy, but the patient was later diagnosed with DILS. The PET-CT showed a multilocular thymic cyst which has shown to be a rare manifestation of the same syndrome.

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