

Parotid Lymphoepithelial Cyst in a HIV-Negative Individual: A Case Report

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Received: 7 July 2017

Revised: 2 October 2017

Accepted: 12 November 2017

Abstract

Benign lymphoepithelial cysts of the parotid are common in retropositive patients, but extremely rare in HIV-negative individuals. We present a 28-year-old man who presented with a painless, gradually increasing swelling in the left parotid region and was clinically diagnosed to have a pleomorphic adenoma of the left parotid gland. Preoperative blood investigations revealed that the patient was seronegative. He underwent a left superficial parotidectomy and the histopathology report indicated a benign lymphoepithelial cyst. The rarity of this lesion in a seronegative patient is the main reason for reporting this case.

Please cite this article as: Carnelio S, Chandramouli M, Rodrigues G. Parotid Lymphoepithelial Cyst in a HIV-Negative Individual: A Case Report. Iran J Med Sci. 2018;43(6):668-670.

Keywords • Parotid gland • Adenoma, pleomorphic • Epidermal cyst • HIV infections

Introduction

Pleomorphic adenomas are the commonest benign parotid tumours causing unilateral enlargement of the parotid gland. Various causes of parotid enlargement in seropositive (HIV) patients are diffuse infiltrative lymphocytosis syndrome, parotitis, intraparotid lymphadenopathy; benign lymphoepithelial cyst (BLEC), adenoid cystic carcinoma, Kaposi sarcoma, and lymphoma.¹ Though rare, BLEC should be considered in the differential diagnosis of parotid swelling and can often be the first indication of HIV infection.² They are slow growing tumours and are extremely rare in non-HIV individuals. This report presents such a rare entity with the treatment modalities and detailed literature review.

Case Presentation

A 28-year-old man with no comorbidities presented with complaints of a painless and gradually increasing swelling in the left parotid region. General examination was unremarkable. Local examination revealed a 5×4 cm painless, non-tender, mobile, and smooth left parotid enlargement. The deep lobe and facial nerve were normal. A clinical diagnosis of pleomorphic adenoma was made and planned for surgery. Blood investigations were normal and the patient was HIV-negative. A workup for Sjogren's syndrome was also negative. A contrast-enhanced computed tomography (CECT) scan revealed a cystic neoplasm of the left parotid gland (figure 1, arrows) and fine-needle aspiration cytology (FNAC) was inconclusive. The patient underwent a superficial parotidectomy (figure 2). Histopathological findings showed a tumour predominantly composed of lymphoid tissue with germinal centres, cystic spaces, and lymphoplasmacytic infiltration with

What's Known

- Benign lymphoepithelial cysts of the parotid are common in retropositive patients and their occurrence could mark seropositivity.

What's New

- Benign lymphoepithelial cysts of the parotid are extremely rare in HIV-negative individuals.
- A differential diagnosis of lymphoepithelial cysts should be considered in patients with parotid swellings and a proper diagnosis and management will offer cure to these patients.

acinar atrophy, which were consistent with BLEC (figure 3). The patient made an uneventful recovery and is asymptomatic with no recurrence on follow-up for 2 years.

A written informed consent was obtained from the patient to present this case report.

Discussion

BLECs usually occur in the tail of the parotid, as in our patient, with an incidence of 3-6% in adults and 1-10% in children. They can be solitary or multiple and represent cystic degeneration within the lymph nodes of the parotid gland.^{1,2} They are slow growing in nature, typically seen in HIV-positive adults. It is unusual to find them in non-HIV individuals. They often grow to be extremely large, resulting in physical deformity and gross facial asymmetry. Most salivary gland BLECs are seen in the parotid as it is the only gland to embryologically develop intraglandular lymphatic tissue. Though differential diagnosis of salivary gland disease (SGD) in seropositive patients would include diffuse infiltrative lymphocytosis syndrome (DILS), parotitis, intraparotid lymphadenopathy, adenoid cystic carcinoma, Kaposi sarcoma, and lymphoma; their varied

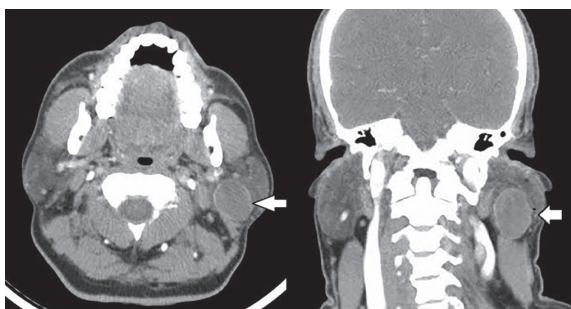


Figure 1: Transverse and coronal CECT showing cystic lesion of the left parotid (arrows).



Figure 2: Superficial parotidectomy showing the cystic lesion.

clinical, radiological, and microscopic features will differentiate them from BLECs.¹ Literature review has no mention of these entities in HIV-negative patients.

Mickulicz first described salivary gland lymphoepithelial lesion (LEL) in 1885. However, in 1985, Ryan et al. first identified this condition in HIV-positive individuals. Other terminologies for BLEC are benign lymphoepithelial lesion (BLEL), Sjogren-like lesion, cystic epithelial lesion, and HIV-SGD.^{1,2}

BLECs are believed to develop due to lymphoid hyperplasia of embryologically sequestered lymphatic tissue within the parotid gland.³ There are two theories as to the pathogenesis: (a) the obstructive theory, which states that lymphoid hyperplasia in the parotid leads to ductal obstruction and salivary dilatation, like a true cyst; (b) HIV-related reactive lymphoproliferation occurs in the intra-parotid lymph nodes resulting in trapping of parotid glandular epithelium causing cystic enlargement. The exact prevalence of BLECs in seronegative individuals is not known, but is thought to be very rare, with only 21 cases reported as of 1981, before the onset of the HIV pandemic.⁴

BLEC typically presents with bilateral parotid enlargement that generally consists of multiple cysts in one gland. These painless cysts grow slowly and are soft, often fluctuant.⁵ These cysts are not locally invasive and have virtually no malignant potential but can cause significant local disfiguration.^{3,5}

Imaging (CT/MRI) shows multiple thin-walled cysts, generally with cervical lymphadenopathy.⁴ However, the occurrence of unilateral lesions without lymphadenopathy is not uncommon, as in our patient. FNAC reveals clear, proteinaceous, and straw coloured fluid with a mixture of epithelial and lymphoid cells.^{1,3} Treatment options include observation, aspiration, sclerotherapy, radiation, and surgery.

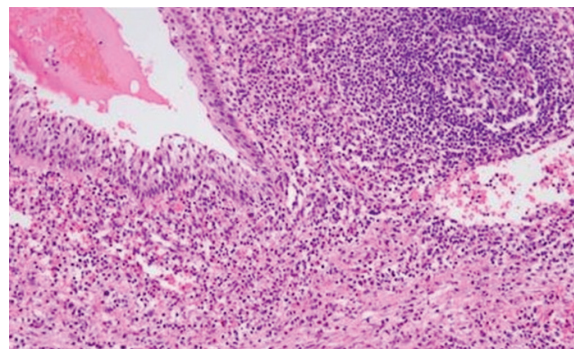


Figure 3: Photomicrograph showing tissue predominantly composed of lymphoid tissue with germinal centres, cystic spaces, and lymphoplasmacytic infiltration (Haematoxylin and Eosin, ×40).

The standard of care for HIV-associated BLEC is HAART, which alone causes resolution of the disease in many patients.⁵ Surgery is generally not recommended as the first-line of treatment in retroviral associated BLEC due to the bilateral and progressive nature of the disease, resulting in a significant risk of facial nerve injury (2.3-6%), and need for multiple surgical procedures post-superficial parotidectomy.³⁻⁵ A complete response is seen in virtually all patients' post-surgery; however, many patients develop cysts on the contralateral side.

Conclusion

Although BLECs are common in retropositive patients, they should be kept in mind in HIV-negative patients as well and should be considered as a differential diagnosis in benign parotid cystic neoplasms. A timely diagnosis and early surgical intervention will offer cure in these patients.

Conflict of Interest: None declared.

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