Available online at www.sciencedirect.com
ScienceDirect
Pathology – Research and Practice 205 (2009) 149–153
PATHOLOGY RESEARCH AND PRACTICE
www.elsevier.de/prp

TEACHING CASES

Extranodal follicular dendritic cell sarcoma of the tonsil – Case report of an epithelioid cell variant with osteoclastic giant cells

Pradeep Vaideeswar[a,*], Sophia M. George[b], Shubhada V. Kane[b], Rachana A. Chaturvedi[a], Shobhana P. Pandit[a]

[a]*Departments of Pathology, Seth G.S. Medical College, Parel, Mumbai 400 012, India*
[b]*Tata Memorial Hospital, Mumbai, India*

Received 29 March 2008; received in revised form 9 July 2008; accepted 25 July 2008

Abstract

Follicular dendritic cell sarcomas are rare neoplasms arising from the accessory cells of the lymph nodes, the follicular dendritic cells. They commonly occur in the lymph nodes, but have also been reported at extranodal sites (especially the tonsil). At both sites, there is usually a proliferation of spindled to ovoid cells, mimicking a mesenchymal tumor. Herein, we report a tonsillar tumor in a 50-year-old man, which was composed exclusively of large polygonal cells and numerous osteoclastic giant cells that resembled a giant cell carcinoma. The true nature of the tumor was revealed after an array of immunohistochemical stains. The patient is well 4 years after tonsillectomy.

Keywords: Follicular dendritic cell sarcoma; Extranodal site; Tonsil; Epithelioid cell variant; Giant cell variant

Introduction

Tumors arising from the histiocytic and dendritic cells are among the rarest of the neoplastic proliferations of the lymphoid system, accounting for $<1\%$ of all tumors [16]. A neoplasm comprising proliferation of spindled to ovoid cells showing morphologic and phenotypic features of follicular dendritic cells (FDC), first described by Monda et al. [12], has been designated as FDC sarcoma/tumor owing to its variable cytologic grade and indeterminate behavior [16]. Although lymph nodes are the most favored sites of occurrence, an origin of the FDC tumors (FDCT) at many extranodal sites (especially the head and neck region) has been described; the tonsil is one such site [8]. Herein, we present a case of tonsillar FDC tumor that posed a diagnostic dilemma due to its atypical "epithelioid" histomorphology, the presence of many osteoclastic giant cells, and, more importantly, an absence of spindle-shaped or ovoid cells.

Case history

A 50-year-old male, a chronic tobacco-chewer, presented with a 2-month complaint of dysphagia. On examination of the oral cavity, a $2 \times 2\,cm^2$ smooth soft to firm swelling involving the left tonsil was noted. Computed tomographic scans from the base of the skull to thorax were unremarkable. There was no associated lymph node enlargement. A tonsillectomy was performed, and the specimen was submitted for histological examination. The postoperative course was uneventful.

*Corresponding author. Tel.: +91 22 24136051x2550, 91 22 32501327; fax: +91 22 24143435.
E-mail address: shreeprajai@yahoo.co.in (P. Vaideeswar).

0344-0338/$ - see front matter © 2008 Elsevier GmbH. All rights reserved.
doi:10.1016/j.prp.2008.07.006

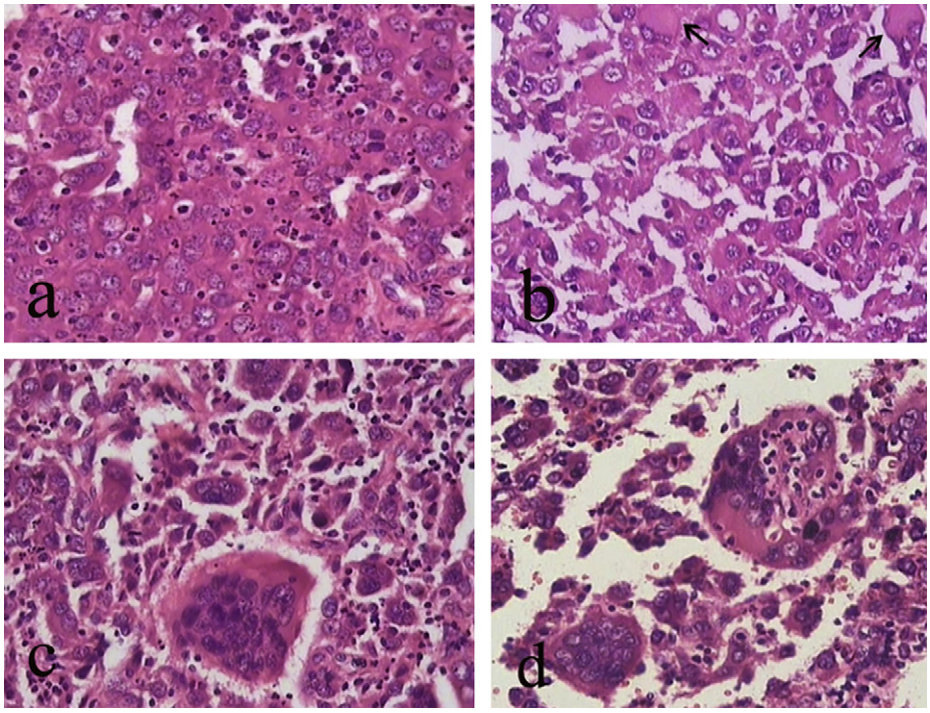

Fig. 1. (a) Large polygonal cells arranged in a syncytial pattern with few scattered lymphocytes, (b) less cohesive areas showing a spiky cytoplasmic outline. Note abundant eosinophilic cytoplasm (arrows), (c) presence of multinucleated giant cells occurring at regular intervals in the tumor, and (d) some giant cells also showed hemophagocytosis (H&E, ×400).

We received a specimen of the enlarged left tonsil, measuring 2.5 cm in diameter. The overlying mucosa appeared ulcerated. The cut surface was yellow-white, solid with faint lobulations. The histology showed replacement of the tonsil by a tumor composed mainly of sheets of large "epithelioid" cells arranged in compact and loose areas. A syncytial growth pattern was seen in compact areas, while a 'spiky outline' was observed in loose areas (Fig. 1a and b). A scant lymphocytic infiltrate was seen at the periphery and in between the cells. The cytoplasm was abundant, pale eosinophilic, and slightly granular; the nuclei were vesicular, some showing prominent nucleoli. Many multinucleated giant cells were also observed, exhibiting hemophagocytosis (Fig. 1c and d). No infiltration, necrosis, nuclear pleomorphism, or increased mitoses were noted. There was preservation and compression of the tonsillar parenchyma with formation of well-defined capsules in most places. The overlying epithelium was focally ulcerated, with candidial pseudohyphae and an actinomycotic granule.

With this histomorphology, differential diagnoses of neoplastic histiocytic proliferation, an undifferentiated carcinoma (lymphoepithelioma-like), poorly differentiated carcinoma, and melanoma were considered. Immunohistochemistry (IHC, Table 1) was done to resolve this dilemma. The tumor cells were positive or diffusely positive for vimentin, CD21 (Fig. 2), CD23, and CD 35, and focally positive for high-molecular weight cytokeratin (HMWCK, 34<beta>E12). A diagnosis of FDCT of the tonsil was made, and the patient was discharged with a close follow-up. The patient has been disease-free to date, after 4 years of operation.

Discussion

In the present report, we describe a rare case of a FDCT of the tonsil with unique histological features. The tumor, which was entirely subjected for processing, was composed exclusively of large epithelioid cells with an almost regular sprinkling of osteoclastic giant cells, many exhibiting hemophagocytosis.

The FDCs are an integral part of the lymph nodes, where their primary role is directed towards antigen presentation and antigen-dependent B-cell maturation. Hence, it is not surprising that the rare neoplastic proliferation of these cells occurs in the lymph nodes [12]. However, they also colonize the lymphoid tissues that are inherent or acquired in other organs, which thereby serve as extranodal locations of occurrence [14]. In the head and neck region, the tonsils are the most common extranodal sites, and, so far, there have been a total of 17 reported tonsillar tumors [8,5,13,15,6,3,20,19,17,10,9,4,2,11]; we are presenting the eighteenth (Table 2). However, Aydin et al. [2] and McDuffie