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CASE REPORT

Follicular dendritic cell sarcoma associated with Castleman's disease presenting in the oral cavity

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Summary Follicular dendritic cell sarcoma (FDCS) is a rare intermediate grade malignant neoplasm of reticular dendritic origin. Castleman's disease (CD) represents a non-neoplastic lymphoproliferative disorder with various clinical and morphological features. FDCS has been reported to be associated with CD. In this article, we describe the first case of follicular dendritic cell sarcoma associated with Castleman's disease presenting in the oral cavity.

Introduction

Follicular dendritic cell sarcoma (FDCS) is a rare intermediate grade malignant neoplasm of reticular dendritic origin, first described by Monda et al. in 1986.[1] This entity frequently develops in the lymph nodes of the cervical, axillary, and supraclavicular region. The tonsil is one of the more common extranodal sites that has been involved.[2,3]

Castleman's disease (CD) represents a non-neoplastic lymphoproliferative disorder with various clinical and morphological features. Its clinicopathologic features depend on various etiologic factors such as Kaposi sarcoma herpesvirus, over secretion of IL-6, and follicular dendritic cell dysplasia. Clinically, solitary Castleman's disease presents as a single mass, often in the mediastinum or the pulmonary hilum, and occurs in a young population. Generalized involvement of nodes in several centers accompanied by systemic involvement is manifested in the multicentric type of Castleman's disease.[4]

Approximately 22 of the 55 cases of follicular dendritic cell sarcoma in the head and neck region that have been reported to date have been extranodal.[3,5] In the literature, 14 examples of FDCS have been reported to be associated with Castleman's disease.[6] In this article, we describe the first case of follicular dendritic cell sarcoma associated with Castleman's disease presenting in the oral cavity.

Case report

A 62-year-old Caucasian female was referred in December 2003 for assessment of a soft tissue swelling in the right maxillary buccal sulcus in the region of the missing 16. The patient wore a complete maxillary acrylic denture,

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and had experienced a sore denture hyperplasia with some mucosal ulceration which had previously resolved following adjustment of the denture by her dentist. Her past medical history was significant for osteoarthritis, one total knee replacement, and a transient ischaemic attack.

Clinical examination confirmed the presence of a large soft tissue mass measuring approximately 2.0 cm in greatest dimension, located in the reflection of the sulcus in the 16 region. The lesion could not be visualized intra-orally or extra-orally, but was easily palpable intra-orally. There was no evidence of ulceration, or epithelial hyperplasia.

An incisional biopsy was performed under local anesthesia and microscopic examination revealed a lymphoid nodule with a hyalinised connective tissue background. Molecular analysis showed a monoclonal B-cell population associated with a bcl-2 [t (14; 18)] chromosome translocation, suggestive of an inconclusive presentation of follicular lymphoma, although there were no constitutional symptoms of lymphoma.

Subsequently, a deeper biopsy was performed one month later on which extensive immunohistochemical and molecular studies were performed. Bcl-2 translocation was not detected, and TCR gene rearrangement studies showed polyclonal beta and gamma chains.

A working diagnosis of follicular dendritic cell sarcoma in association with extranodal Castleman's disease was made.

The patient was referred to the Oncology Clinic at the Princess Alexandra Hospital, Brisbane, Australia. A CT and MRI scan of the head and neck revealed a slightly lobulated but well-defined right infra-temporal fossa mass lesion marginating the anterior aspect of the right coronoid and mandibular ramus, with some erosion of the adjacent bone (Fig. 1). The cortex was absent on the posterior zygoma, but the tumour did not extend beyond its immediate margins.

There was no evidence of any involvement in the neck, chest, abdomen or pelvis. There was no lymphadenopathy or hepatosplenomegaly. A wide operative excision was performed which included a close normal margin, and this was followed with radiotherapy to the surgical site only. Nine months later the patient presented with a submandibular lymph node enlargement, which was confirmed on CT, and she underwent subsequent irradiation to the ipsilateral neck. The patient is now disease free.

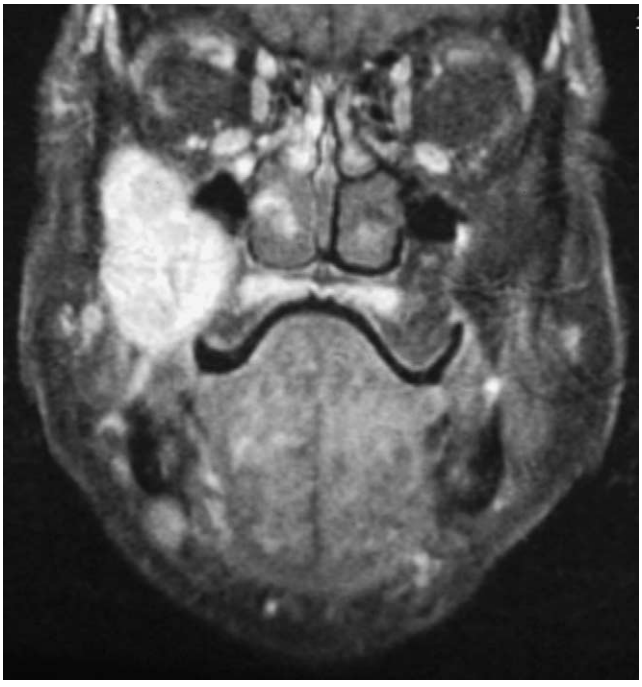

Figure 1 MRI showing large mass lesion in right infra-temporal fossa. The tumour appears to displace but not invade adjacent structures. The mass measured approximately 4.5 cm in the supero-inferior plane, 3.0 cm in the coronal plane, and 3.0 cm in the antero-posterior plane.

Histopathologic examination

Haematoxylin and eosin stained sections revealed two morphologically distinct areas. In one area, there were germinal centres with prominent mantle zones, many of which contained regressed follicles. Interfollicular sclerosis and lymphoid infiltrate were also evident (Fig. 2). The other distinctive area of abnormality was a monomorphic spindle cell proliferation containing admixed lymphocytes and scattered plasma cells (Fig. 3).

Immunohistochemistry

The described lymphoid area of proliferation showed follicles to be CD20+, CD79a+, CD3, CD43 and CD5 negative within the follicles, but were positive within interfollicular small regular lymphoid cells. The follicles showed weak positivity for CD10, but were negative for BCL-2. Kappa Lambda stains showed apparent polyclonal cytoplasmic immunoglobulin. BCL-6 was restricted to the germinal centres.

The spindle proliferation was strongly positive for CD21 (FDC marker) (Fig. 4). The spindle cell proliferation was

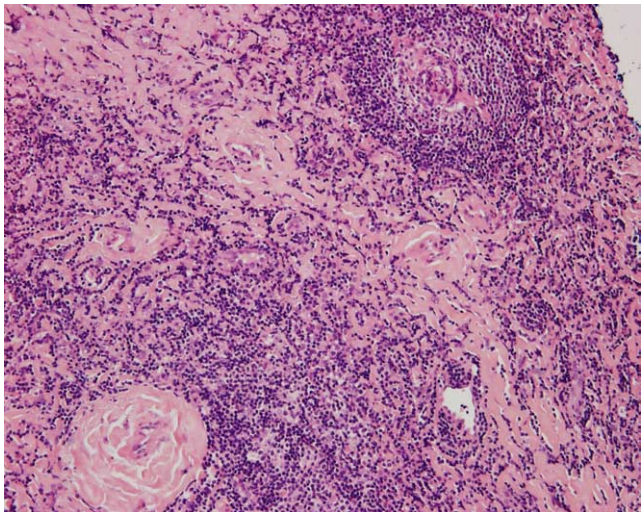

Figure 2 Castleman's disease showing germinal centres with prominent mantle zones, many of which contain regressed follicles. Interfollicular sclerosis and lymphoid infiltrate are also evident (H & E × 100).