CASE STUDY

Follicular Dendritic Cell Sarcoma of the Tonsil

A 80-year-old caucasian man presented in the otorhinolaryngology emergency with a 3-week long history of sore throat. This patient reported heavy smoking in the past 20 years. He denied fever, dysphagia or weight loss. On clinical examination a 2 cm exophytic violaceous mass involving the upper pole of the left tonsil was found. The mucous membrane covering the tonsil was normal, without ulcerations. No other masses or lymphadenopathies were noted.

Computed tomography of the head and neck confirmed a tumour with 1.9 × 1.8 cm in the left tonsil (Fig. 1). After tonsillectomy was performed, histological examination was compatible with a Follicular Dendritic Cell Sarcoma (FDCS), as the tumour cells expressed CD23, CD21 and Vimentine on immunohistochemical study (Fig. 2). This tumour was classified as T1 N0 M0 according to TNM Classification System. Positron emission tomography carried out after the tonsillectomy did not show any residual fixation in the left tonsil area or the lymph nodes. Because the margins of resection were positive, partial pharyngectomy was performed. The new margins were tumour free.

The patient died 1 week after, at home, of cardiac arrest.

Discussion

Follicular dendritic cells are non-lymphoid, non-phagocytic elements that fall under the generic designation of accessory cells of the lymphoid system. Their primary role is to capture and present antigens and immune complexes to B-cells. The FDCS was first described in 1986 by Monda et al. and is most frequently found within the axillary, mediastinal and cervical lymph nodes, but extranodal involvement has been seen in approximately 30% of cases: tonsils, oral cavity and abdominal organs are the most frequent. In the head and neck, the tonsil is the most frequently affected structure, even though less than 20 cases have been described.

The aetiology of FDCS is poorly understood. To date, Castleman disease and Epstein–Barr infection are the only predisposing factors that have been identified.

Figure 1: Computed tomographic scan showing a mass in the left tonsil (arrow).
Follicular dendritic cell sarcoma of the tonsil

Follicular dendritic cell sarcoma (FDCS) are most common in adults of 30–50 years of age (average 47 years old), with an equal gender distribution.1,2,7,9

Most patients with FDCS present to the doctor with a painless, slow-growing and well-circumscribed mass.1,4,8 When affecting the tonsils, it frequently presents as a lobulated, well-demarcated and grey-white mass,2 like in the case presented (Fig. 1).

Histological examination and immunohistochemical assays are mandatory for diagnosis. Histological examination commonly reveals eosinophilic oval or spindle cells arranged in sheets and fascicles,1 which typically stain positively with CD21, CD23, CD35, CD31, Ki-M4, Ki-M4p, S-100 protein and Vimentine immunohistochemical markers.1-8 As most of these markers are not routinely used in poorly differentiated tumours, FDCS may be misdiagnosed in more than 50% of cases.1,6

The differential diagnosis of FDCS includes undifferentiated carcinomas, squamous cell carcinoma and large cell lymphoma, among others.2-10

FDGS is considered a low- or intermediate-grade malignant neoplasm.1,4,6-8 Metastasis may occur in 25% of the cases1 (usually affecting the lungs, liver or lymph nodes) and local recurrences are common (40%)4; locoregional recurrence has been reported up to 15 years after initial treatment.4,6 To date, no deaths have been reported with FDCS of the tonsil.6

The prognosis is aggravated by tumour size (>5 cm–6 cm) and the presence of nuclear pleomorphism, numerous mitoses (>5 per 10 high-power field), coagulative necrosis or significant cellular atypias on histological examination.1,4-8

In the current case none of these prognostic factors were reported – the patient died, probably, due to conditions unrelated to the tumour.

Complete surgical excision is the gold-standard treatment of tonsils’s FDCS1,6,8,9; some authors advocate neck dissection in case of evidence of ganglionar metastasis.3 The benefit in survival of postoperative radiation or chemotherapy is unknown.7

Conclusion

Follicular dendritic cell sarcoma is a recently recognized condition that must be considered in the differential diagnosis of unilateral tonsillar swelling, due to its potentially aggressive behaviour with local recurrences and metastasis.

References