Localised extranodal non-Hodgkin’s lymphoma of the tonsil: report of a rare case

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Abstract

Introduction
Non-Hodgkin's lymphoma of the Waldeyer’s ring is a relatively rare entity and the palatine tonsil is the most frequently involved site. Although, the exact aetiology remains unclear, a number of predisposing factors have been identified, including human immunodeficiency virus and Epstein–Barr infection. We report a case of localised extranodal non-Hodgkin’s lymphoma of the tonsil.

Case report
A 64-year-old woman presented with a sore throat. On physical examination, an approximately 2 × 1 cm smooth non-tender mass was observed in the left palatine tonsil. Serology was negative for human immunodeficiency virus and Epstein–Barr virus. Computer tomography scan revealed a non-enhancing left tonsillar mass but no signs of neck lymphadenopathy. The patient underwent bilateral tonsillectomy. Histological examination confirmed a diagnosis of NHL diffuse large cell type of B phenotype. The stroma was densely infiltrated by medium-sized lymphoma cells and the neoplastic cells were positive for CD3, Cyclin D1, CD1a, HMB45, CKAE1 and CKAE3 (Figure 3). Bone marrow biopsy did not reveal lymphomatous involvement (stage 1 according to tumour, node, metastasis classification). The patient received chemotherapy based on CHOP (cyclophosphamide, doxorubicin, vincristine and prednisolone) protocol combined to Rituximab. During follow-up, she remains disease-free 30 months after diagnosis.

Conclusion
Non-Hodgkin’s lymphoma rarely involves tonsils with the diffuse large B-cell type being common at this location. A combined treatment consisting of chemotherapy and radiotherapy leads to a satisfactory outcome in patients with this uncommon neoplasm, which tends to present at an early stage and to have a favourable prognosis.

Introduction
Non-Hodgkin’s lymphoma (NHL) of the oral cavity and oropharynx account for 13% of all primary extranodal NHL with approximately 70% of these occurring in the tonsils. The palatine tonsil is the most frequently involved site followed by palate, gingiva and tongue. Most lymphomas found in the palatine tonsils are the B-cell type, and of these, diffuse large B-cell lymphoma (DLBCL) represents most of the cases, reaching as much as 80% in some of the groups studied. Although, the exact aetiology remains unclear, a number of predisposing factors have been identified, including human immunodeficiency virus and Epstein–Barr infection. This study reports a rare case of localised extranodal NHL of the tonsil.

Case report
We report a case of a 64-year-old woman who presented with a sore throat during the last four months. On physical examination, an approximately 2 × 1 cm smooth non-tender mass was observed in the left palatine tonsil. The remainder of the physical examination was normal. Laboratory studies for tumour markers and serology tests for human immunodeficiency virus and Epstein–Barr virus were negative as well. Computer tomography scan revealed a non-enhancing left tonsillar mass but no signs of neck lymphadenopathy. Past history did not appear to be contributory regarding the aetiology. The patient underwent bilateral tonsillectomy. Histological examination confirmed a diagnosis of NHL diffuse large cell type of B phenotype. The stroma was densely infiltrated by medium-to large-sized lymphoma cells and the surface stratified squamous epithelium was ulcerated (Figures 1 and 2). Immunohistochemically, the neoplastic cells were positive for CD19, CD20, CD10, CD79a, CD22, Bcl-2, Bcl-6 and negative for CD57, CD56, CD2, EBV, CD4, CD5, CD7, CD8, S-100p, CD3, Cyclin D1, CD1a, HMB45, CKA1 and CKA3 (Figure 3). Bone marrow biopsy did not reveal lymphomatous involvement (stage 1 according to tumour,
node, metastasis classification). The patient received chemotherapy with a CHOP regimen (cyclophosphamide, doxorubicin, vincristine and prednisolone) combined to Rituximab. During follow-up, she remains disease-free 30 months after diagnosis.

Discussion
NHL represents a small percentage of oral malignancies and palatine tonsil is the most frequently involved site. This lymphoma has a peak incidence in the 6th and 7th decades of life in published series and the sex incidence is slightly male predominant. Clinical signs and symptoms are not specific and occur as a result of asymmetrical tonsillar enlargement. They may include a sensation of fullness in the throat, sore throat, dysphagia, odynophagia, otalgia, cervical adenopathy, tonsillar swelling or snoring. Systemic symptoms, such as fever, weight loss and night sweats are uncommon and may develop in patients with advanced disease.

Most tonsillar lymphomas reported in the literature are of B-cell origin and the most common histologic type, ranging from 67% to 96%, has been reported to be DLBCL. In many series, it is reported that the majority of these patients have localised disease (stage I or II).

Patients with lesions that were clinically determined to be over 7 cm in size (bulky) had a significantly poorer outcome as compared with those with smaller tumours. Authors have reported 5-year survival rates of 65%–85% for patients with early stage disease and no present bulky mass.

Treatment includes chemotherapy alone, radiotherapy alone or a combination of both. The majority of patients received chemotherapy followed by radiotherapy.

Conclusion
NHL rarely involves tonsils with the diffuse large B-cell type being common at this location. A combined treatment consisting of chemotherapy and radiotherapy leads to a satisfactory outcome in patients with this uncommon neoplasm, which tends to present at an early stage and to have a favourable prognosis.

Consent
Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

References