Unilateral tonsillar lymphoepithelioma with ipsilateral parapharyngeal space involvement: A case report

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Abstract

We report a case of unilateral tonsillar lymphoepithelioma with extension into the ipsilateral parapharyngeal space, and we review the clinical, histologic, and radiographic findings of the case. The patient presented with a tonsillar mass that was confirmed on biopsy to be lymphoepithelioma. Computed tomography demonstrated ipsilateral parapharyngeal space involvement. Association with Epstein-Barr virus was not assessed since it does not affect the treatment modality. We also review the literature and discuss the diagnosis and current treatment options.

Introduction

Lymphoepithelioma (LE) most commonly occurs in the nasopharynx. It is a nonkeratinizing (undifferentiated) squamous cell carcinoma, previously classified by the World Health Organization (WHO) as a WHO type 3 lesion.[1] Non-nasopharyngeal LE is rare and has been described as occurring in Waldeyer's ring, the palate, hypopharynx, larynx, trachea, salivary glands, and even the maxillary sinus.[1-8] Tonsillar LE is typically unilateral, and about three-quarters of patients have nodal involvement at diagnosis.[3,8] Here we report a case of unilateral tonsillar LE with direct extension into the ipsilateral parapharyngeal space.

Case report

A 66-year-old white man was referred by his primary care physician for an enlarging right tonsillar mass. The patient had noticed the mass after developing dysphagia to solids and ipsilateral otalgia, both of which had grown worse during the 4 months preceding his visit to his physician. He also reported to us that he had lost weight during the previous 4 months; when we saw him 4 weeks after our initial evaluation, we documented that he had lost another 12 kg. The patient is a smoker and drinker, but it appears that there is little association between LE and tobacco/alcohol use.[8]

Physical examination revealed a right tonsillar mass with ulceration inferiorly. He had localized tenderness on palpation, and flexible endoscopy revealed moderate displacement of the right soft palate.

Computed tomography (CT) of the neck revealed a heterogeneously enhancing mass in the right tonsillar fossa measuring 2 × 3 × 4.5 cm. The mass was contiguous with a node in the ipsilateral parapharyngeal space that measured 2 × 4.5 cm (figure 1). The oropharyngeal airway was compressed, as was the right internal jugular

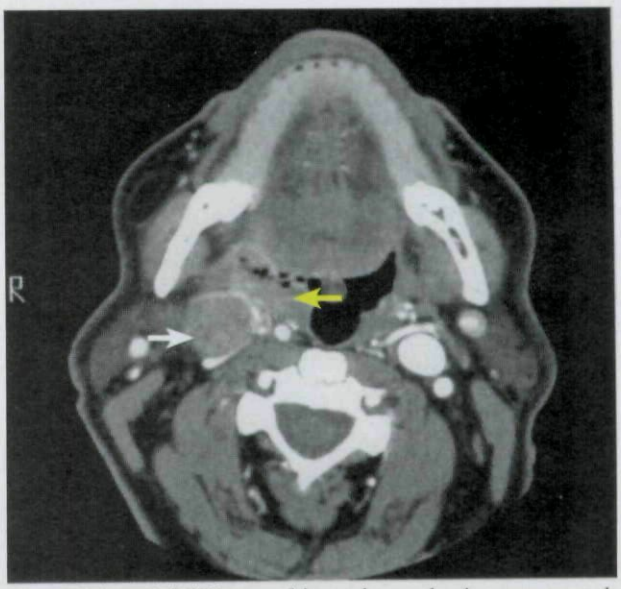

Figure 1. An axial CT image of the neck reveals a heterogeneously enhancing mass in the right tonsillar fossa measuring 2 × 3 × 4.5 cm (yellow arrow). This was contiguous with a node in the ipsilateral parapharyngeal space that measured 2 × 4.5 cm (white arrow) (H&E, original magnification ×100).

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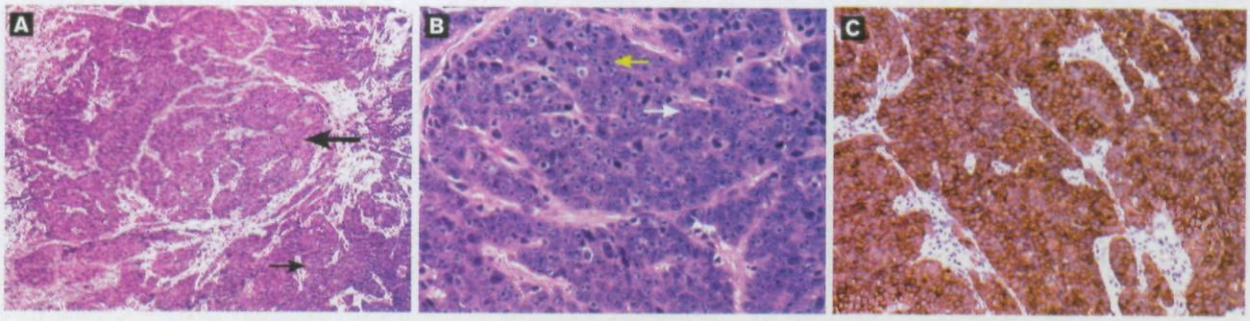

Figure 2. ***A:*** *The neoplastic cells (large arrow) form cohesive nests, or sheets. An inflammatory infiltrate rich in small, mature lymphocytes (small arrow) is noted at the periphery of the tumor cells (H&E, original magnification ×100).* ***B:*** *At higher magnification, the neoplastic cells (yellow arrow) have round vesicular nuclei, prominent nucleoli, indistinct cell borders, and increased mitoses (white arrow) (H&E, original magnification ×400).* ***C:*** *Immnunohistochemical studies show strong cytokeratin staining of the neoplastic cells and negative staining for leukocyte common antigen (original magnification ×200).*

vein. Magnetic resonance imaging of the neck with gadolinium was also performed to provide a baseline for future comparison, if needed; it showed tumor extension into the right pyriform sinus.

Biopsy specimens obtained 1 week after the patient's initial visit to our clinic established a diagnosis of LE (undifferentiated carcinoma). In the specimen shown in figure 2, A, the neoplastic cells can be seen forming cohesive nests, or sheets. A surrounding inflammatory infiltrate rich in small, mature lymphocytes is seen at the periphery of the tumor cells. At higher magnification, the neoplastic cells can be seen with round vesicular nuclei, prominent nucleoli, indistinct cell borders, and increased mitoses (figure 2, B). Immnunohistochemical studies showed strong cytokeratin staining of the neoplastic cells and negative staining for leukocyte common antigen (figure 2, C). These findings are consistent with a diagnosis of LE.

Subsequent metastatic workup (3 weeks later) included CT of the chest, abdomen, and pelvis, which revealed a nonspecific 5-mm nodule on the lower lobe of the patient's right lung.

The patient received chemoradiation therapy and had a complete response. He remained free of disease until he was lost to follow-up at 11 months.

Discussion

Our case was unique in that our patient with unilateral tonsillar LE had tumor extension into the ipsilateral parapharyngeal space. Non-nasopharyngeal LE is rare and is significantly different from nasopharyngeal LE. One difference is that non-nasopharyngeal LE tends to affect a younger population.[8] Unlike patients with nasopharyngeal LE, patients with non-nasopharyngeal LE typically present with neck masses.[3] About three-quarters of these patients will have cervical lymphadenopathy at the time of diagnosis,[8] which is often confirmed on CT. A histopathologic diagnosis is helpful, especially given the rarity of LE outside the nasopharynx.

Another difference between the two types of LE is that the association between Epstein-Barr virus (EBV) and non-nasopharyngeal LE is less common than the association between EBV and nasopharyngeal LE,[6,9] although Ahuja et al reported on 4 patients with palatal LE who all tested positive for EBV.[3] Although the association between EBV and non-nasopharyngeal LE remains uncertain, treatment is the same regardless and includes neoadjuvant chemotherapy followed by radiation therapy. This combination has been shown to improve disease-free survival when compared with radiation therapy alone.[8] Surgery is reserved for patients in whom chemoradiation has failed.

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