European Annals of Otorhinolaryngology, Head and Neck diseases 136 (2019) 331–333

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What is your diagnosis?

# An unusual palatine tonsil lesion

C. Fabre[a,*], H. Gil[a,b], C.A. Righini[a,b,c]

[a] Service Hospitalo -Universitaire d'ORL et Chirurgie cervico-faciale, CHU de Grenoble - Alpes (CHUGA), France
[b] Faculté de médecine, Universités des Alpes, Grenoble, France
[c] UGA/UMR/CNRS 5309/Inserm 1209, Institut Albert-Bonniot, Grenoble, France

## 1. Case report

A 56-year-old woman with no particular medical history and no smoking or drinking risk factors, attended the emergency department for left supraclavicular oedema present for three days, which had become worse despite treatment with clarithromycin. Clinical interview revealed moderate dysphagia to solids and the appearance of bilateral cervical lymph nodes two weeks previously in a context of nasopharyngitis. She presented a good general state of health with no weight loss. The patient also reported abdominal pain for one month, for which colonoscopy and abdominal CT scan had been requested, but not yet performed at the time of management. Physical examination revealed:

- left supraclavicular induration ascending to the jugular region;
- bilateral, non-inflammatory, painless, mobile, and indurated superior and middle jugular lymph nodes measuring about one centimetre.

Serology (EBV, CMV, HIV, toxoplasmosis) was not contributive.

The patient was referred to the ENT department for diagnostic lymphadenectomy. Physical examination revealed an exophytic granulating lesion of the inferior pole of the right palatine tonsil. Contrast-enhanced neck, chest and abdomen CT scan revealed:

- heterogeneous bilateral cervical lymphadenopathy, with a necrotic centre and a left supraclavicular lymph node conglomerate associated with internal jugular vein thrombosis;
- suspicion of an appendicular tumour (Fig. 1A).

$^{18}$FDG PET-CT (Fig. 1B) and diagnostic tonsillectomy were performed. Histological examination confirmed the presence of a tonsillar tumour (Fig. 1 C).

* Corresponding author at: CHU Grenoble Alpes, avenue Maquis-du-Grésivaudan, 38700 La Tronche, Grenoble, France.
*E-mail address:* cfabre2@chu-grenoble.fr (C. Fabre).

https://doi.org/10.1016/j.anorl.2019.05.014

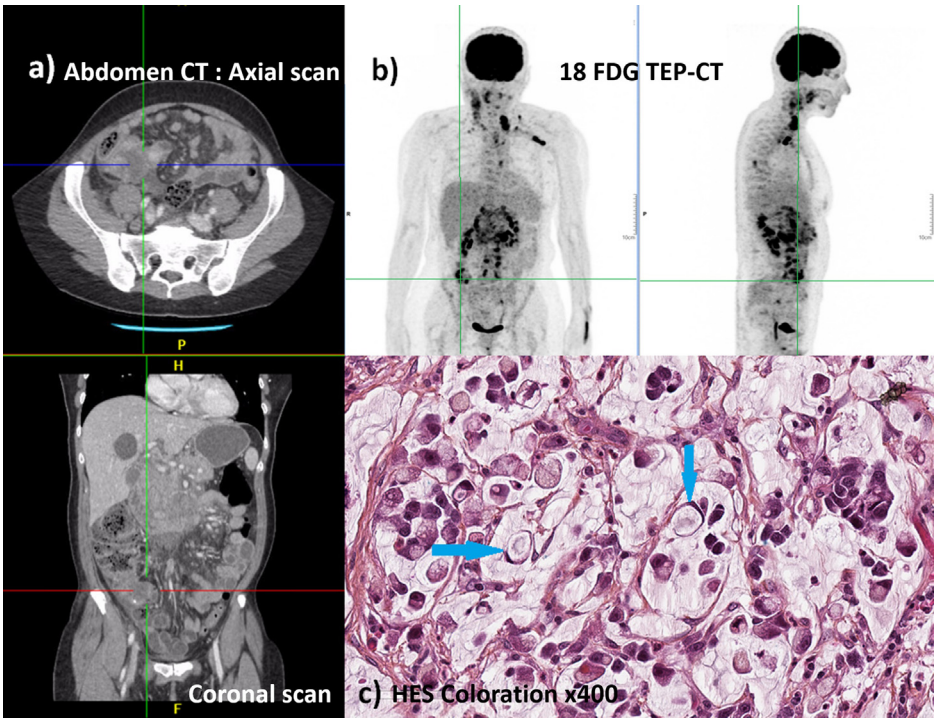


**Fig. 1.** Staging of the case reported here: Metabolic (B) and radiological (A) imaging of the tonsillar tumour. Histological appearance after fixation and staining (C).

**What is your diagnosis?**

## 2. Answer

This tumour was a tonsillar metastasis from a mucinous signet-ring cell adenocarcinoma of intestinal origin, probably arising from the appendix ($^{18}$FDG PET-CT and abdominal CT). Tumour cells were non-cohesive and were floating in mucoid material. These zones of mucus dissected the lamina propria, resulting in an alveolar-like architecture. Tumour cells comprised hypertrophic nucleolated nuclei with the presence of mitotic figures, and some cells had a "signet-ring" appearance with a nucleus displaced towards the periphery of the cell by intracytoplasmic mucin (blue arrows on the histological section).

Tumour morphology and immunophenotyping looking for proteins of the DNA mismatch repair (MMR) system failed to specify the exact tissue of origin or the presence of mutations or genetic instability of the MMR system. Colonoscopy visualized an infiltrated and ulcerated appearance at the centre of the caecal mucosa over the zone of implantation of the appendix. Biopsies of the periphery of the appendicular orifice did not reveal any signs of malignant proliferation. However, according to the gastroenterologist, these biopsies were too superficial and should have been repeated, but this examination could not be performed due to rapid disease progression. Upper GI endoscopy was normal.

Due to the presence of multiple metastases and disease progression, 5-fluorouracil, oxaliplatin and bevacizumab chemotherapy was initiated, but was complicated by hepatorenal insufficiency requiring discontinuation of treatment. Palliative care was therefore decided and the patient died one month later.

Metastases of the palatine tonsils are rare, as less than 100 cases have been reported in the literature. In more than 90% of cases, the primary tumour is a melanoma (cutaneous or mucosal), lung cancer or kidney cancer [1].

Intestinal signet-ring cell adenocarcinoma is characterized by intracytoplasmic mucin accumulation that displaces the nucleus towards the periphery of the cell, as in the case reported here. The majority of primary gastrointestinal tumours of this type are gastric tumours, but they can also arise throughout the gastrointestinal tract [2]. The appendicular origin of this type of carcinoma is extremely rare and remains controversial [3,4]. This aggressive cancer is often only discovered at the metastatic stage due to its highly lymphophilic nature, consequently resulting in a very poor prognosis [2]. Tonsillar metastasis as the first sign of the disease, leading to the discovery of an intestinal primary cancer, has been reported only once in the literature in a case of caecal mucinous signet-ring cell adenocarcinoma [5]. In the presence of a tumour of the palatine tonsil, although rare, a tonsillar metastasis from a primary tumour situated outside of the upper aerodigestive tract constitutes the main differential diagnosis, while the two most common histological types of primary tumours of the palatine tonsil are squamous cell carcinoma and lymphoma. A very cautious diagnostic approach is therefore essential, particularly in the absence of smoking and drinking. In this case, the presence of symptoms and physical signs suggesting metastases from a primary tumour situated outside of the upper aerodigestive tract must be systematically investigated.

## Disclosure of interest

The authors declare that they have no competing interest.

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