Extensive maxillofacial plasmablastic lymphoma in an immunocompetent patient

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Accepted 20 July 2014

DESCRIPTION

A 46-year-old man presented to a dentist reporting maxillary pain on the left side which had persisted for a few weeks. The pain along with an irregular appearance of the gingival mucosa were attributed to dental pathology and a molar was extracted. The initially limited gingival lesion evolved into a rapidly growing palatal tumour and recurrent episodes of unilateral epistaxis occurred. A CT scan and a biopsy were performed and the patient was referred to our department several weeks after the first symptoms had appeared.

Inspection of the oral cavity revealed a purple, fleshy, well-delimited exophytic tumour occupying the entire hard palate on the left and deforming the maxillary gingiva (figure 1). The left nasal cavity was also filled with a solid mass. A CT scan and MRI confirmed the presence of a voluminous

Figure 2 An MRI T2-weighted coronal view through the maxillary sinus, revealing a destructive tumour extending from the oral cavity to the ethmoid and the orbit through the maxilla and the nasal cavity.

Figure 1 The tumour was well delimited, with an exophytic 'fleshy' appearance, occupying the hard palate (A) and extending to the maxillary gingiva (B) on the left side.

tumour destroying the left maxilla and extending to the ethmoid, the orbit, the nasal cavity, the masticator space and the oral cavity (figure 2). A PET-CT scan revealed pulmonary and bone involvement along with multiple lymphadenopathies (cervical, mediastinal, subdiaphragmatic). Histology and immunohistochemistry (CD20-/+, CD79a-/+, VS38c+, CD10+, MUM1+, κ -, λ -, BCL2-, BCL6-, MiB1 90%) confirmed the diagnosis of EBV+ plasmablastic lymphoma (PBL). Bone marrow biopsy and HIV testing were negative. Despite aggressive chemotherapy the patient died 8 months later.

Lymphomas are rare among sinonasal, maxillary and palatal tumours, the differential diagnosis including both malignant and benign lesions (such as squamous cell carcinoma, accessory salivary gland tumours, adenocarcinoma, sarcomas, odontogenic tumours). PBL was first described by Delecluse *et al* in 1997.¹ It is a rare and aggressive B-cell lymphoma, usually associated with Epstein-Barr virus (EBV) infection and mainly occurring in immunodeficient, most often HIV-infected, patients²; the oral cavity is a common location although extraoral forms exist and have a worse prognosis. Treatment is not standardised; it is generally based on multiagent chemotherapy with or without radiation therapy and stem cell transplantation. The overall prognosis remains poor with 1-year survival of less than 50%.²

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To cite: Vourexakis Z,
Dulguerov P. *BMJ Case Rep*
Published online: [please
include Day Month Year]
doi:10.1136/bcr-2014-
204042

Learning points

- ▶ Patients reporting unilateral nasal symptoms (epistaxis, new-onset discharge and progressive obstruction), unexplained maxillary pain or hypoaesthesia should be referred to an otolaryngologist for nasal endoscopy.
- ▶ In most cases, biopsies of suspect lesions of the oral and nasal cavity are easy to perform, even on an outpatient basis; being essential for accurate diagnosis and an appropriate treatment, it is important to realise them in a timely fashion.
- ▶ Plasmablastic lymphoma is a rare B-cell lymphoma with poor prognosis, most frequently occurring in the oral cavity and mainly affecting immunocompromised patients.

Contributors Both authors have contributed to the treatment of the patient and the writing of this manuscript.

Competing interests None.

Patient consent Not obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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