CASE REPORT

Isolated diffuse type B-cell lymphoma of the palate: an unusual entity

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Phoebe Roche,
phoebeeroche@rcsi.ie**SUMMARY**

Lymphomas frequently occur as extranodal lesions in the head and neck, but are rarely seen in the palate. We present a case of isolated diffuse type B-cell lymphoma of the palate, which occurred in a 28-year-old man. The patient had no history of immune compromise, and he presented to the emergency department with a 7-month history of a painful, non-healing ulcerative mass in the hard and soft palate. Positron emission tomography facilitated pretreatment assessment of the extent and activity of the lesion. Histopathological and immunohistochemical analyses of biopsied tissue confirmed a diagnosis of diffuse type B-cell lymphoma. The clinical findings and therapeutic challenges in this heterogeneous group of malignancies are discussed.

BACKGROUND

Non-Hodgkin's lymphomas (NHLs) typically originate in the lymphoid system. Approximately 90% of NHLs are of B-cell origin and when compared with Hodgkin's disease, NHLs have been found to be both less predictable, and significantly more likely to spread to extranodal sites.¹

Between 24% and 48% of all extranodal NHLs occur in the head and neck region. Of these, less than 5% occur within the oral cavity² and case reports of lymphoma presenting with isolated hard palate lesions are extremely rare.³ Although most extranodal lymphomas in the head and neck are of B-cell origin, natural killer/T-cell variant lymphoma is reported as the most common subtype that presents in the palate.⁴

Lymphadenopathy is the most common manifestation of all types of lymphoma. B symptoms including fevers, night sweats, weight loss and fatigue are also often reported. Less frequently, symptoms, which arise as a result of mass effect, for example dysphagia, may occur.

CASE PRESENTATION

A 28-year-old man presented to the emergency department with a 7-month history of slow-growing painful ulceration of the hard and soft palate. The patient denied any history of difficulty in swallowing or bleeding from the lesion.

He had no medical history and there was no significant family history of malignant disease. He was a former heavy smoker of 60 cigarettes per day and when further questioned, he admitted to 7 kg unintentional weight loss—despite maintaining a normal diet, and recent drenching night sweats.

Intraoral examination revealed a 3×4 cm oval exophytic lesion over the hard and soft palate, with

areas of central necrosis. His neck examination was normal, and fibre optic nasal endoscopy showed a degree of fullness in the right tongue base (figure 1). No obvious lesion was seen in the oropharynx and there was no evidence of disease in the nasopharynx.

The initial haematological work up included a full blood count, erythrocyte sedimentation rate and a negative HIV test.

INVESTIGATIONS

An urgent positron emission tomography (PET) scan was requested and a representative biopsy was performed under local anaesthetic. Staging PET scans confirmed the presence of avid FDG (SUV max >42.8) uptake in the palate extending from the lesion to the piriform sinus submucosally, with appearances highly suggestive of a malignant process. The microscopic evaluation of the tumour cells obtained at biopsy confirmed the presence of atypical lymphoid cells with cytoplasmic clearing and inconspicuous nucleoli.

Immunohistochemical staining was positive for the B-cell markers CD45 CD20 BCL CD3 CD10 CD5 and MUM1 transcription factor. MIB proliferation marker staining gave a proliferation index of 90%. The findings were consistent with a diagnosis of non-Hodgkin's diffuse type B-cell lymphoma (figures 2 and 3).

Lesion at presentation

Figure 1 Oval exophytic lesion involving the hard and soft palate.

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Figure 2 H&E stained specimen showing (left to right) lymphocytosis, apoptosis, vacuolisation with muscle, CD 20 stained tissue—B-cell marker and A13 stained tissue—keratin marker—out ruled squamous cell carcinoma.

DIFFERENTIAL DIAGNOSIS

The initial differential diagnoses included squamous cell carcinoma, salivary gland tumour, paranasal sinus malignancy, fungal infection, Mycobacterial infection and extranodal presentation of Rosai-Dorfman disease.

TREATMENT

The patient was promptly referred for early chemotherapy. He received a total of seven cycles of rituximab, cyclophosphamide, adriamycin and vincristine (figures 4 and 5).

OUTCOME AND FOLLOW-UP

On cessation of treatment, the lesion showed complete resolution. The patient remained stable with normal complete

blood count results at latest review 3 months postcompletion of treatment.

DISCUSSION

Malignant lymphomas account for almost 5% of all head and neck malignant disease. The prevalence, with which they occur, is increasing, and increasing numbers of extranodal lymphomas are now being seen in routine clinical practice. It is estimated that approximately 20 510 deaths occur due to Hodgkin's disease and NHLs per annum in the USA alone⁵ and the rising incidence in the past two decades has been characterised by a marked increase in the occurrence of extranodal lymphoma.⁶ Tumours are often described as being either indolent or aggressive⁷ and the survival rates reported are affected accordingly.

Figure 3 (Left) Coronal positron emission tomography I image showing avid FDG uptake in palate. (Right) Sagittal view showing sub mucosal spread to the tongue base.

Regimen: RITUXIMAB plus CHOP

- Pre medication:
 - Paracetamol 1g PO pre Rituximab (Day 1)
 - Dexamethasone 8mg IV pre Rituximab (Day 1)
 - Chlorpheniramine 10mg IV pre Rituximab (Day 1)
 - Ondantsetron 8mg IV pre CHOP

Drug	Dose	Route of Admin	Infusion Fluid	Day	Doses per cycle
Rituximab	375 mg/m squared	IV	500ml NaCl 0.9%	1	1
Cyclophosphamide	750 mg/m squared	IV	N/A	1	1
Doxirubicin	50 mg/m squared	IV	N/A	1	1
Vincristine	1.4 mg/m squared	IV	N/A	1	1
Prednisolone EC	60mg OD	PO	N/A	1 - 5 then taper	5 then taper
Ranitidine	150 mg BD	PO	N/A	With all steroid doses	Per steroid doses

Figure 4 Treatment regimen.

Aggressive tumours have a shorter natural history and a more varied response to treatment, in contrast to indolent lymphomas, which are associated with better outcomes and reported survival rates of up to 10 years.

The rarity with which isolated extranodal diffuse B-cell lymphoma of the palate occurs, made this case noteworthy. Patients with uncommon disease in this anatomically complex region can often prove exceptionally challenging to diagnose and to manage. With any extranodal lymphoma, it is important

to note that there is often an associated inflammatory response in the surrounding tissue. Early recognition of lesions such as this one and the taking of representative biopsies are extremely important because, in the early stages, the disease may be confined entirely to the palate and therefore respond well to irradiation, whereas disseminated disease necessitates chemotherapy. As demonstrated, microscopy supported by thorough immuno-histochemistry staining is an essential component of accurate final diagnosis.

In the last 10 years, the use of FDG PET in the visualisation of metabolically active tumour cells has become critically important in the assessment of patients with cancer. There is a growing consensus that PET is a superior modality of imaging for the staging, diagnosis and post-treatment surveillance of lymphoma to gallium scintigraphy⁸ MRI and CT. With clear advantages over these other modalities, FDG PET/CT is now the imaging modality of choice in Hodgkin's disease and most NHLs (figure 6).⁹ In this instance, it provided an excellent assessment of the degree of submucosal spread of the lesion, which was essential for the evaluation of the disease stage.

Post treatment

Figure 5 Complete resolution of lesion by cycle 7.

Learning points

- ▶ The oral cavity is an anatomically complex region and lesions can prove exceptionally challenging to diagnose.
- ▶ Isolated extranodal diffuse B-cell lymphoma of the palate is extremely rare.
- ▶ FDG positron emission tomography is the imaging modality of choice for diagnosis, staging and surveillance.
- ▶ Early recognition of lesions and taking of representative biopsies are extremely important.

Contributors PON was involved in the diagnosis and management of the patient throughout. EK was involved in the case. HR had responsibility of the patient. All four co-authors contributed fully to the writing of this paper.

Published Cases of Palate NHL

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Figure 6 Published cases of palate non-Hodgkin's lymphomas.

Patient consent Obtained.

Competing interests None.

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