Mantle cell lymphoma: primary oral presentation

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ABSTRACT

Mantle-cell lymphoma is an uncommon lymphoid malignancy of B-cells. It is often aggressive and prognosis is poor. A 69-year-old gentleman with a history of ischaemic heart disease was referred from primary care with a painless right floor of mouth swelling that had been present for 1 month. He otherwise completely asymptomatic. Incisional biopsy of the lesion was undertaken and marker studies demonstrated mantle cell lymphoma. Positron emission tomography–computed tomography and bone marrow biopsy showed widespread but low volume involvement. The patient was referred to the haematology multidisciplinary team for further assessment and treatment.

KEYWORDS

Mantle cell lymphoma – Non-hodgkin lymphoma – Oral lymphoma

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Introduction

Mantle-cell lymphoma (MCL) is an uncommon lymphoid malignancy of B-cells of the mantle zone or primary lymphoid follicle. MCL is itself a relatively new diagnosis, having only been recognised by the World Health Organization in 2001.¹ It typically occurs in middle-aged to older adults, with a marked male predilection, and rarely presents primarily in the head and neck.² Prognosis of this often-aggressive lymphoma is poor.

Case history

A 69-year-old retired accountant was referred by his general dental practitioner with a right-sided floor-of-mouth swelling. Medically, he had ischaemic heart disease and suffered a myocardial infarction in 2002, with an implantable defibrillator being placed 2 years later. He had a 40 pack a year cigarette smoking history and rarely drank alcohol.

The mass had gradually increased in size over the previous month but otherwise it remained completely asymptomatic. The patient reported no other systemic symptoms such as weight loss or night sweats.

On examination, there was a 4cm × 2cm rubbery submucosal mass present in the right floor of mouth (Fig 1). The overlying mucosa appeared normal and saliva was freely expressed from Wharton's duct. There were no similar lesions, lymphadenopathy or hepatosplenomegaly on full clinical examination.

The patient underwent incisional biopsy of the lesion under local anaesthesia. The histopathology results demonstrated squamous mucosa with underlying mucous-type

salivary gland tissue. Much of the salivary gland had been replaced by a dense lymphoid infiltrate that consisted predominantly of mature lymphocytes. The appearances were suggestive of a lymphoepithelial sialadenitis, which, as it is uncommon in minor salivary glands, was sent for marker studies to exclude lymphoproliferative disorder. These marker studies demonstrated MCL phenotype.

The patient was referred to haematology, where he underwent staging investigations of the MCL in the form of positron emission tomography–computed tomography (PET-CT) and bone marrow biopsy. These investigations showed widespread but low volume involvement. There was a 2cm lymph node in the right upper neck. The multidisciplinary team came to the decision to treat conservatively and repeat the PET-CT scan in several months' time to assess disease progression and to plan further treatment if required.

Discussion

MCL is a rare form of non-Hodgkin lymphoma, with an incidence of 0.55 in 100,000.³ It is a well-defined lymphoid neoplasm that shows proliferation of B-lymphocytes. It is more common in men and its incidence increases with age, with the median age of diagnosis being 63 years.⁵ Extra-nodal involvement is common and it often presents at a late stage (III–IV) with leukaemic spread, bone marrow involvement, hepatosplenomegaly and lymphadenopathy.⁴ Response to conventional chemotherapy and overall survival are both poor. Factors that can help to predict poor outcome include the age of the patient, hyponatraemia, hypoalbuminaemia, splenomegaly and stage of disease. It presents most commonly in Waldeyer's ring and can also occasionally occur in

Figure 1 Lesion at initial presentation

salivary and lacrimal glands, as well as the skin, stomach and spleen.⁴

Lymphoma represents 2.2% of all malignant neoplasms of head and neck, 3.5% of malignant intraoral neoplasms, 5% of tumours of salivary glands and 2.5% of all lymphomas on

the head and neck; 85% of the lesions affect tonsils and palate. Extra-nodal MCL lesions rarely present primarily in the oral cavity, with only one case being previously reported in the floor of the mouth, and the majority of other lesions presenting in the hard palate.²

Treatment provided to these patients varies from combination chemotherapy to chemotherapy and radiotherapy to observation. Typically, MCL has an aggressive clinical course that is resistant to therapy, with a mean survival of 3 years.⁵ Recognition of MCL and its differentiation from other non-Hodgkin's lymphoma subtypes is important because of this poor prognosis and changing therapeutic regimens.

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