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Anaplastic Large Cell Lymphoma of the Tongue

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Abstract

Presentation

A 66-year-old lady presented with a 7-month history of oral phase dysphagia and a 1-month history of a mass increasing in size on her left tongue with enlarged lymph nodes in the right neck.

Diagnosis

Biopsy showed this to be an anaplastic lymphoma (ALK) kinase negative anaplastic large cell lymphoma (ALCL). Radiological investigations showed no disseminated spread of disease.

Treatment

Her case was referred to the haematology service and she was treated successfully.

Conclusion

Primary oral cavity ALCL is a rare pathology with only 16 cases reported in the English language literature. However, it has a favourable prognosis with a 77% 5-year survival. This case reminds us to be cognisant of different pathologies when assessing patients in head and neck clinics and represents another rare systemic ALK-negative ALCL case involving the oral cavity.

Introduction

More than 95% of Head and neck cancer (HNC) are squamous cell carcinomas which are associated with cigarette smoking, oncogenic viruses with excessive alcohol consumption a strongly related co-factor [1]. Lymphomas are uncommon causing 2-5% of oral malignancies [2]. An exceedingly rare lymphoma of the oral cavity is presented.

Case Report

A 66-year-old lady presented with a 7 month history of oral phase dysphagia and a 1 month history of a mass increasing in size on her tongue. Her past medical history was significant for a treated oligodendroma, hyperlipidaemia and osteoporosis. Clinically she had a large exophytic mass occupying 80% of the anterior left tongue with preserved movement and had nodes palpable in the right level 2 area. CT neck identified a large 5 cm tumour in the left oral cavity with nodal disease in the right submental and submandibular area. PET CT showed no distant disease. Full blood count, renal, liver function and LDH were normal.

Biopsy showed squamous mucosa with underlying stroma infiltrated by a poorly differentiated malignant tumour. Immunohistochemical studies were positive for lymphoid marker CD45. Tumour cells were CD3 positive, CD30 positive, ALK 1 negative, CD4 positive, CD8 negative, CD56 negative and EBV negative. The immunohistochemistry confirmed a diagnosis of ALK negative ALCL (see figure 1).

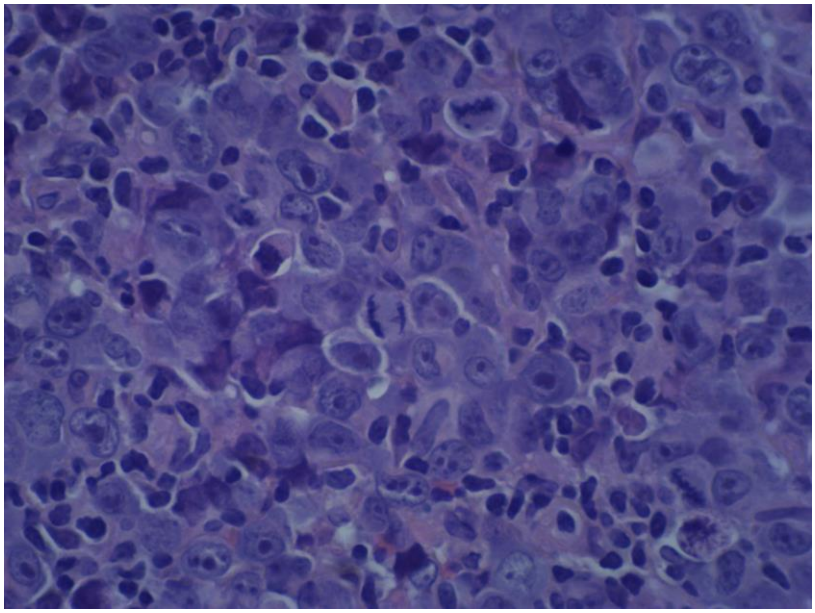

Figure 1: Left anterior tongue incisional biopsy. H&E 20X Magnification. Sheets of large pleomorphic cells, with the characteristic 'horseshoe shaped' nuclei with prominent and often numerous nucleoli. The cells have abundant eosinophilic cytoplasm. Numerous mitoses are also present.

Her care was referred to the haematology service and she was treated successfully with cyclophosphamide, vincristine sulphate and prednisolone. At the time of writing she remains disease free.

Discussion

Lymphomas are a heterogenous neoplasm of lymphoid tissue subdivided into Hodgkin and non-Hodgkin. Hodgkin disease is characterised by Reed-Sternberg cells [3]. Non Hodgkins Lymphoma (NHL) consists of malignant neoplasms of lymphoid tissues derived from B and T lymphocytes.

While primarily originating in lymph nodes, extra nodal NHL accounts for 20-40% of cases [4]. 50% of lymphomas occur in the head and neck, particularly in cervical lymph nodes with Waldeyer's Ring the most common site.

First described in 1985, anaplastic large cell lymphoma (ALCL) originates from T or NK cells and accounts for 2% of NHL [5]. Like other NHL subtypes ALCL primarily involves nodal areas but extra nodal sites can occur. They are characterised by large blastic tumour cells with anaplastic morphology, sinusoidal infiltration of lymph nodes, pseudocohesive appearance with expression of cytokine receptor CD30 [6]. Tumours cells are atypical and larger than usual lymphoma cells with more abundant tumour cells. ALCL are further classified based on their expression of the anaplastic lymphoma kinase (ALK) gene. The ALK expression is caused by a t(2;5) translocation abnormality[7]. ALK +ve disease represent 50-80% of cases and occur in younger patients carrying a better prognosis. Although an aggressive tumour, it responds well to chemotherapy with 77% of patients having a 5 year survival rate [4].

Primary ALCL in the oral cavity is rare; only 16 cases have been reported. Previous publications have occurred in patients at a mean age of 45 years. Most cases have been reported in the gingiva, 2 cases in the hard palate, soft palate and lip. Spontaneous remission has been described [8 9]. While lymphoma is relatively rare in the oral region for the head and neck surgeon, it is the most frequent non epithelial malignant tumour in the region [10]. Although lymphomas are regarded as aggressive disease processes ALCL has a 77% 5 year survival rate[4]. With this in mind it should be considered when assessing patients presenting with oral masses or ulceration, particularly with repeated recurrence and spontaneous regression. While difficult to distinguish from oral inflammatory disorders and other neoplastic diseases it highlights the needs for a high index of suspicion. It is essential to diagnose this neoplasm to facilitate appropriate therapies and a favorable prognosis.

This case reminds us to be cognisant of different pathologies when assessing patients in head and neck clinics and represents another rare systemic ALK-negative ALCL case involving the oral cavity.

Declaration of Conflicts of Interest:
The authors declare that there are no conflicts of interest regarding the publication of this paper.

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