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Spindle Cell Sarcoma of the Tongue- An Unusual Presentation

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ABSTRACT

Sarcomas are rare malignant tumors of mesenchymal cells which occur primarily in the head and neck region in the adults. Occurrence of this tumour in the tongue has very rarely been reported and is associated usually with recurrence and metastasis, than complete and permanent cure after the first surgical intervention. We report here an unusual case of spindle cell sarcoma arising from the tongue in a 25 year-old male who presented with an ulcerated mass on the anterior right margin of the tongue.

Keywords: Sarcoma, Tongue, Immunohistochemistry.

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INTRODUCTION

Sarcomas are rare malignant tumors of mesenchymal cells and account for less than 1% of all malignancies.[1] Sarcomas occur primarily in the head and neck region in the adults, accounting for 5-15% of the cases.[2,3] Only a few cases of oral cavity sarcoma have been reported yet.[1] The commonest site in the head and neck region include the orbit, nasopharynx, paranasal sinuses, cheek, neck, middle ear and larynx.[1] Occurrence of this tumour in the tongue has very rarely been reported.[3,4] We report here an unusual case of spindle cell sarcoma arising from the tongue in a 25 year-old male who presented with a ulcerated mass on the anterior right margin of the tongue.

CASE SUMMARY

A 25-year-old man presented a painless ulcerated mass on the right margin of the tongue for the last 6 weeks. Clinical examination showed a firm, ulcerated mass of 15x10 mm in size. The remaining mucosa of the oral cavity showed no abnormality with no regional lymphadenopathy. The personal history of the patient was not significant, but there was a history of cigarette smoking for the last 10 years.

Clinical diagnosis was suggestive of a benign mesenchymal neoplasm. The fine needle aspiration biopsy was inconclusive. An excisional biopsy was performed and histologically, the tumor showed a fascicular or storiform pattern with pleomorphic cells infiltrating the striated muscle fibres of the tongue (Figure 1). The spindle-shaped tumor cells had ill-defined cell outline with pale cytoplasm and mild dyskaryosis with distinct nucleoli with a mitotic count of 3-4 per HPF. (Figure 2) An intense stromal mixed inflammatory reaction was seen. A diagnosis of spindle cell sarcoma was given.

Immunohistochemically, the tumor cells showed diffuse and strong positivity for vimentin (Figure 3) and smooth muscle actin and negative for S- 100 protein. Adjuvant chemotherapy was administered consisting of 6 cycles of gemcitabine, 800 mg/m^2 (day 1) and docetaxel 80 mg/m^2 (days 1 and 8). The patient is well after 6 months of follow up period.

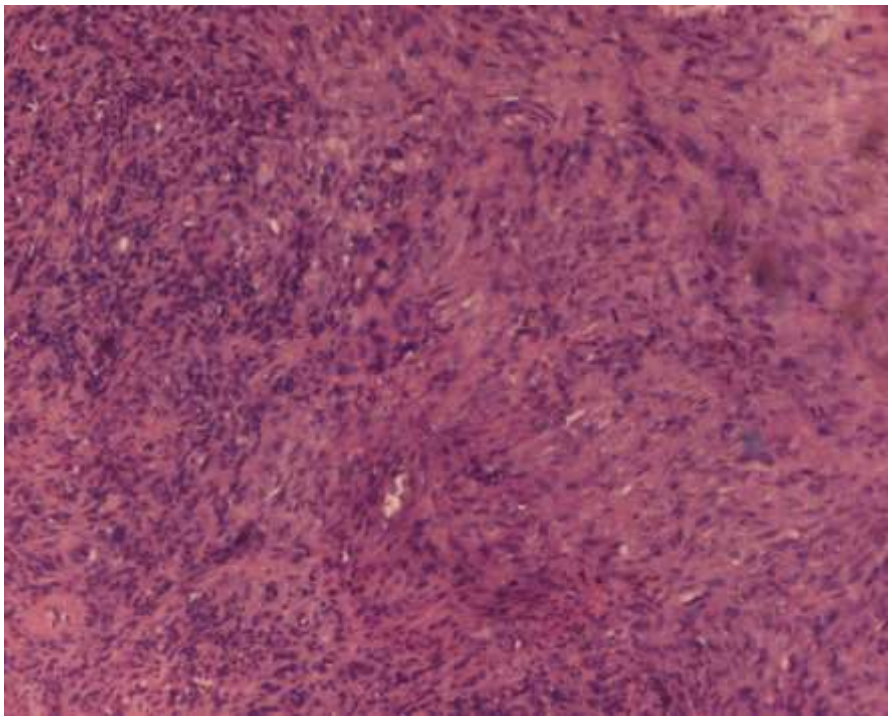

Figure 1: Microscopically the tumor showed a fascicular or storiform pattern with cells infiltrating the striated muscle fibres of a tongue. The spindle-shaped tumor cells had ill-defined cell borders, pale cytoplasm and mild dyskaryosis with distinct nucleoli with a mitotic count of 3-4 per HPF. Haematoxylin and Eosin x 10X.

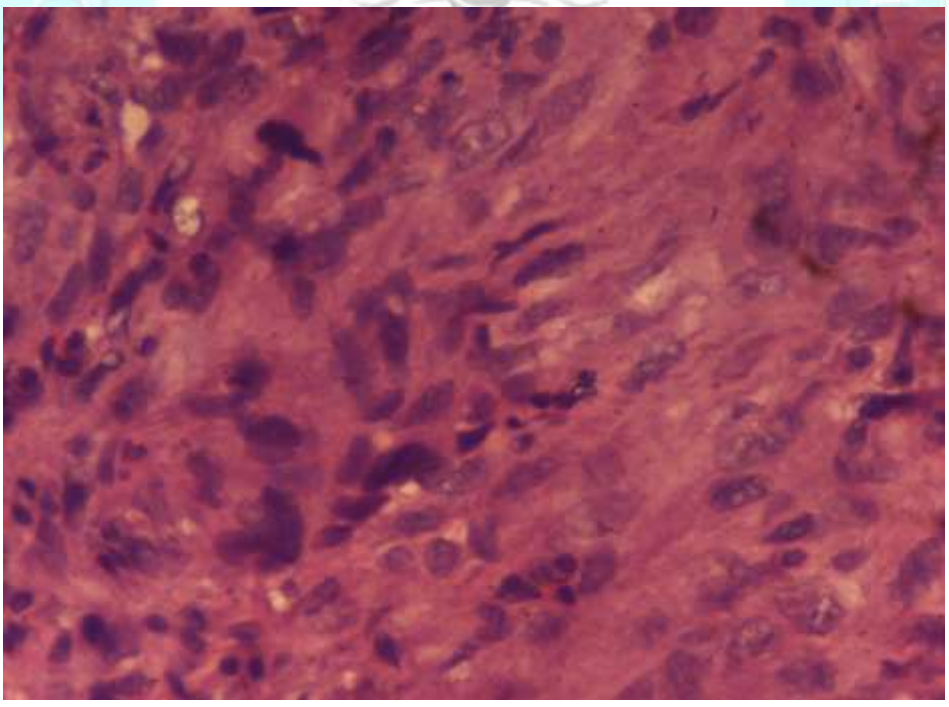

Figure 2: Microscopically the tumors cells show marked pleomorphism with hyperchromatic nucleus and indistinct eosinophilic cytoplasm. Haematoxylin and Eosin x 40X.

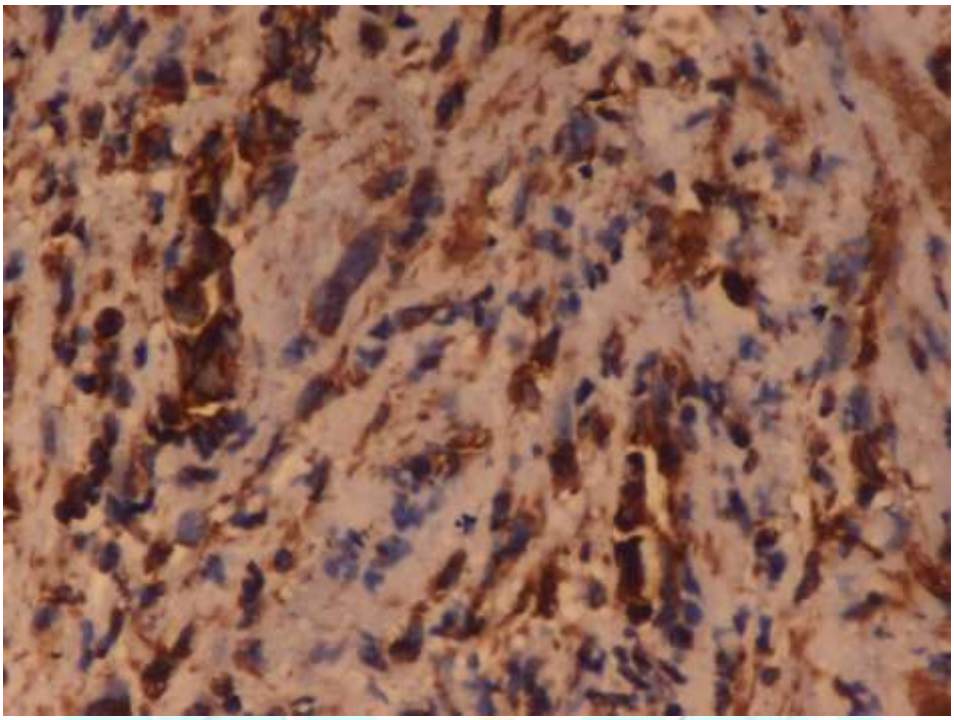

Figure 3: Immunohistochemically, the tumor cells showed diffuse and strong positivity for vimentin. IHC Vimentin x 40 X.

DISCUSSION

Sarcomas are relatively rare malignant tumors, arising from the connective tissue. They include tumors from bony elements (osteosarcoma and chondrosarcoma) and from soft tissue elements (soft tissue sarcomas). Approximately 80% of all sarcomas in the head and neck are soft tissue sarcomas and only 20% are osteosarcomas or chondrosarcomas.[2] Soft tissue sarcomas are named according to their tissue of origin; rhabdomyosarcoma, fibromyosarcoma, synovial sarcoma, liposarcoma, malignant fibrous histiocytoma (MFH), alveolar soft part sarcoma, hemangiopericytoma, neurogenic sarcoma, etc.[5]

The classification of soft tissue sarcomas is still evolving.[4] Andriana et al classified sarcomas according to the tissue of origin, the histological grade or the respective anatomical site in the head and neck region.[6] Recent advances in immunohistochemistry and molecular biology have led to the confirmation in the diagnosis of soft tissue sarcomas.[2] In a review of 281 sarcomas Duagard et al reclassified their cases using immunohistochemistry, and changed their original diagnosis in 57% of the cases.[5] Moreover, 7% of these cases were not found to be sarcomas. However, we should take into account that up to 10% of soft tissue sarcomas remain unclassifiable.[5]

Soft tissue sarcomas arising in the head and neck are very rare accounting for 10% of all sarcomas and approximately 1% of head and neck tumors.[5] Maxillofacial sarcomas constitute between 4-8% of all malignancies in the head and neck region.[6] There is a bimodal

distribution of presentation with an initial peak incidence between 2-5 years of age and a second surge at 10-19 years.[7]

The spindle cell variant was first described in 1992 and is associated with a good prognosis.[8] Clinical findings, diagnostic evaluation and therapy depend upon location of the primary tumour and age of the patient.[7] Current treatment regimen of soft tissue sarcomas is a multi-modal approach comprising of surgery and either radiotherapy or adjuvant chemotherapy.[9] The prognosis is related to the age of the patient, site of origin, widest diameter of the tumour, resectability, presence of metastases, number of metastatic sites or tissues involved, presence or absence of regional lymph node involvement and histopathological subtype.[9,10] In this case, the patient was managed with surgery and chemotherapy. The patient responded very well clinically after the initial phase of treatment.

Soft tissue sarcomas are tumors with extensive local invasion, resulting in a high rate of local recurrence, which depends on the histological type of the tumor. Liposarcoma, well-differentiated sarcoma, and MFH have an increased tendency in local recurrence, while fibrosarcoma, synovial sarcoma, and alveolar soft part sarcoma have an increased tendency in long delayed metastases. The occurrence of nodal metastases in soft tissue sarcomas is usually less than 5%, while the lung is the initial site of distal metastasis, in approximately 90% of patients.[7] Synovial sarcoma of the tongue is extremely rare, with tumor expression of cytokeratins AE1-3, and EMA as well as Bcl-2 with molecular analysis of positive SYT/SS2 fusion transcript.[10]

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