Case Report

Monophasic synovial sarcoma of the tongue in an elderly lady: A diagnostic dilemma

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ABSTRACT

Synovial sarcoma (SS) is a morphologically characteristic sarcoma of uncertain lineage, typically containing both epithelial and mesenchymal elements. The most common location is the extremities, and very few cases have been reported from the head and neck region. The monophasic spindle cell variant often poses a diagnostic challenge as it mimics poorly differentiated carcinoma or sarcoma. We report a case of monophasic SS of tongue in a 74-year-old female. On Immunohistochemistry, the tumor cells showed strong, diffuse positivity for vimentin, Bcl-2, CD99, and focal positivity for epithelial membrane antigen. Tumor cells were negative for pan cytokeratin, smooth muscle actin, S100, HMB-45, CD34. A high index of clinical suscipicion is required to reach an accurate diagnosis for institution of appropriate therapy.

Key words: Elderly, monophasic synovial sarcoma, tongue

INTRODUCTION

Synovial sarcoma (SS) is a mesenchymal tumor commonly affecting the extremities in young adults and represents approximately 10% of all soft tissue sarcomas.[1] Only about 5% of cases occur in the head and neck region, the most common anatomical site being hypopharynx and parapharyngeal spaces.[2,3] Location of the tumor in the oral cavity is rare, the tongue being an uncommon site as only about 12 cases have been previously reported in the literature.[4,5] The classical biphasic morphology of SS does not pose any diagnostic difficulty, however, the monophasic variant of SS can mimic any soft tissue sarcoma, poorly differentiated carcinoma or malignant peripheral nerve sheath tumor (MPNST) and is likely to be misdiagnosed when present in rare sites such as head and neck.[4] We report the clinical features, histopathology, differential diagnosis, and immunohistochemical (IHC) profile of a case of monophasic SS of tongue in an old lady, due to its rarity and uncommon age of presentation. It needs to be emphasized that a high index of clinical suspicion is required to diagnose this entity which can present in unusual locations so that appropriate treatment can be instituted.

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CASE REPORT

A 74-year-old lady presented to the ENT Department with an ulceroproliferative growth measuring 5 cm × 4 cm × 2 cm on the right border of the tongue since 6 months. There was h/o bleeding on food intake. On examination, deviation of the tongue to the left side along with speech impairment was seen [Figure 1a]. There was no h/o tobacco or alcohol intake. The patients' general condition was fine, and her routine hematological and biochemical parameters were normal. No other abnormality was detected in the oral cavity, nasopharynx, or laryngopharynx. There was no lymphadenopathy or hepatosplenomegaly.

Positron emission tomography-computed tomography scan done revealed a fluoro-deoxy-glucose avid heterogeneously enhancing ill-defined mass measuring 54.5 mm × 14.9 mm × 31.2 mm with an ulcerated contour and infiltrative margins was seen involving the right lateral border of oral tongue. Extension of the tumor into the posterior one-third of the tongue along with infiltration of the intrinsic muscles was seen. Genioglossus,

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geniohyoid muscles and the adjacent mandible was normal [Figure 1b]. A whole body scan done was negative for regional or distant metastasis. A wide local excision was done. The hemiglossectomy specimen revealed a grayish-white tumor involving the right lateral border of the tongue [Figure 1c and d]. Microscopy revealed a partially ulcerated stratified squamous epithelial lining which was unremarkable. The subepithelium showed a spindle cell tumor arranged in intersecting fascicles with a focal storiform pattern infiltrating the skeletal muscle fibers. The individual tumor cells were short, plump, spindly appearing to overlap with oval hyperchromatic nuclei and scant eosinophilic cytoplasm. At places, the tumor cells showed pleomorphic nuclei and prominent nucleoli, and a prominent vascular pattern [Figure 2a, b and d] Intervening stroma was scant and showed a moderate lymphocytic infiltrate. Mitoses were frequent. There was no evidence of necrosis. Epithelial or neural differentiation and rosettes were not evident. IHC revealed a strong and diffuse positivity in the tumor cells for vimentin, CD99, and Bcl-2, [Figure 3a,c and d] and focal positivity for epithelial membrane antigen (EMA) [Figure 2c]. The tumor was negative for pan cytokeratin (CK), smooth muscle actin (SMA), S100, HMB-45, CD34. Proliferative activity of the lesion was evaluated using Ki-67 (mouse monoclonal. Clone MIB 1, ready to use, Biogenex, USA) and was 18%. The histopathological diagnosis given was a high-grade malignant spindle cell tumor favoring an SS. No postoperative radiotherapy or chemotherapy was given. Patient is doing well as seen on her last follow-up visit 4 months after surgery.

DISCUSSION

Synovial sarcoma is a morphologically characteristic sarcoma of uncertain lineage with variable epithelial and mesenchymal differentiation and having a specific chromosomal translocation t (X; 18)(p11:q11).[6] SS is a misnomer as it has no origin or differentiation toward synovium. It is reported to occur from birth to 89 years, the median age of presentation being 32 years.[7] Though SS can occur at any site in the body, the most common location (80%) is the lower extremity. In the head and neck region, SS has been reported in a soft palate, tongue, tonsil, larynx, cervical esophagus, and trachea.[8]

The histological patterns of SS can be varied; the most common biphasic pattern consists of epithelial cells and spindle cells in varying proportions. Monophasic spindle cell SS often shows the presence of relatively uniform spindle cells arranged in distinct fascicles, variably interspersed by collagenous matrix and calcification. A hemangiopericytoma – like vascular pattern may be seen. Out of the 12 cases of SS of tongue reported till date,

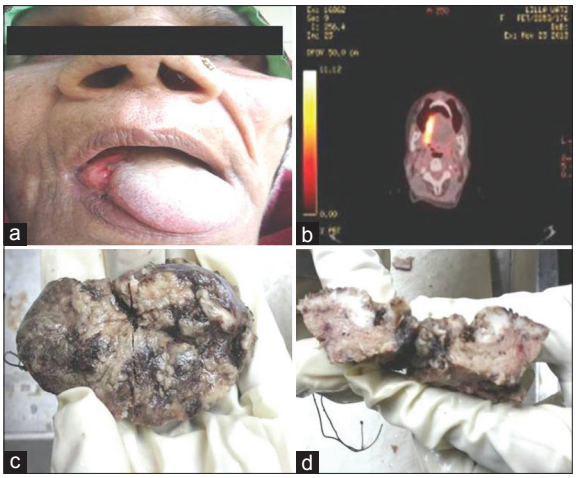

Figure 1: (a) Ulceroproliferative growth 5 cm × 4 cm × 2 cm on the right border of the tongue. (b) Positron emission tomography-computed tomography scan done revealed a fluoro-deoxy-glucose avid heterogeneously enhancing ill-defined mass. (c and d) Specimen of hemiglossectomy showing a greyish-white tumor on cut section measuring 5.5 cm × 1.5 cm × 3.5 cm involving the right lateral border of oral tongue

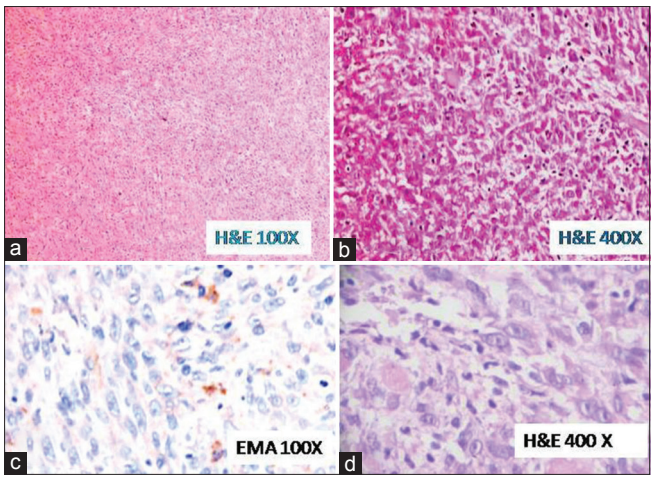

Figure 2: (a and b) Photomicrograph showing a malignant spindle cell tumour with a fascicular and storiform pattern. (c) Epithelial membrane antigen: Focal positivity in tumour cells. (d) Pleomorphic cells with prominent nucleoli seen at places

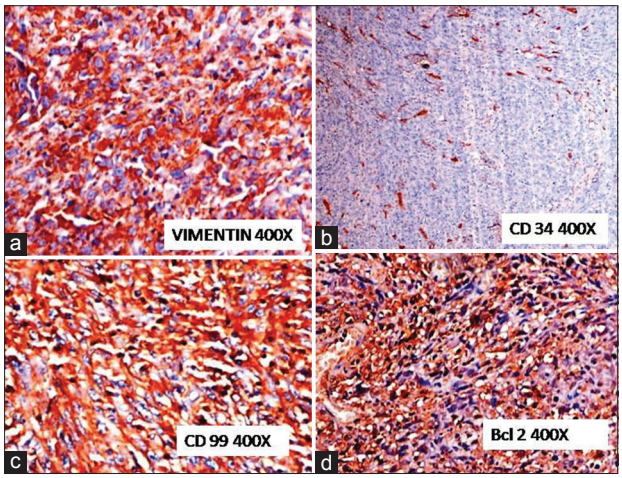

Figure 3: (a,c,d) IHC showing strong and diffuse positivity for vimentin, CD99 and Bcl-2 in tumour cells. (b) CD34 negative

11 cases had a classical biphasic pattern, and only one case had a monophasic spindle cell morphology.[4,5] The present case is only the second case with a monophasic histology of SS in tongue.

Our initial differential diagnosis included a spindle cell carcinoma, primitive neuroectodermal tumor (PNET), MPNST, melanoma, high-grade myofibroblastic sarcoma, and a spindle cell rhabdomyosarcoma. In our patient, the overlying epithelium was normal. Rosettes suggestive of PNET or features of MPNST like alternate cellular and myxoid fascicular pattern, verocay bodies or geographic areas of necrosis were not present. Most MPNSTs show positivity for S-100 protein which was negative in our patient. Tumor cells with prominent eosinophilic nucleoli made us suspect a melanoma, which was ruled out due to the absence of melanin pigment and negative HMB-45 staining. High-grade myofibroblastic sarcoma was considered since it occurs in head and neck region with cellular fascicles infiltrating muscle. However, no giant cells or SMA positivity was noted. Absence of rhabdomyoblasts ruled out a spindle cell rhabdomyosarcoma.

Synovial sarcoma is immunoreactive for CK and EMA and can be mistaken for carcinomas, 65% of SS show positivity for CD99 (MIC-2) and 40% to S-100 protein creating confusion with MPNST or PNET. Our patient was negative for S-100 and pan CK with EMA showing focal positivity. A strong and diffuse positivity for CD99 and Bcl-2 is diagnostic and was noted in our case.[9]

Studies show that SS are immunoreactive for TLE1 and calponin, and though helpful, have limited diagnostic value. These, however, could not be carried out in the present case. Molecular detection to demonstrate t (X: 18)(p11;q11) translocation can also be done.

Surgery with wide local excision is the recommended treatment for localized SS, nodal dissection or chemotherapy is not required, which highlights the importance of an accurate diagnosis. Postoperative radiotherapy may have a role in inoperable cases or those with positive surgical margins.

More recently, the development of individualized, antineoplastic immunotherapy against the protein produced by fusion of SYT-SSX gene may change the management of SS in the future.[10]

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