Angiofibrosarcoma of the tongue - A rare histopathological presentation

Murad Ahmad, Kafil Akhtar, Mohd Naim

Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, India

Received April 03, 2015; Revised April 17, 2015; Accepted April 18, 2015; Published Online April 21, 2015

Case Report

Abstract

Angiosarcoma is a rare malignant neoplasm and very few cases have been reported in the literature. Skin and superficial soft tissue is the most common sites involved by angiosarcoma. Oral cavity comprises of less than 8.0% of head and neck angiosarcomas. Harter in 1927 first reported a case of angiosarcoma of the tongue in literature. Histopathology of angiosarcoma is widely variable, so the role of immunohistochemistry is very important to arrive at a correct diagnosis. Here, we report a case of a recurrent angiofibrosarcoma of the tongue in a 31 year old male, with variable histological features causing diagnostic difficulties.

Keywords: Angiosarcoma; Tongue; Histopathology; Immunohistochemistry

Introduction

Oral cancer is the sixth most prevalent cancer worldwide. The incidence of oral cancer is showing an increasing trend in developing countries like India. More than 90.0% of intraoral malignancies are of squamous cell carcinoma (SCC) of different grades and microscopic variants. Tongue is a frequent site of oral malignancy and is third most common site for SCC of oral cavity. Other infrequent oral malignancies reported in the literatures are salivary gland carcinomas, adenoid cystic carcinoma, lymphoma, and melanoma. Angiosarcoma (AS) is a rare oral malignant neoplasm and very few cases have been reported in the literatures. Here, we report a case of recurrent angiofibrosarcoma of the tongue with variable histological features, causing diagnostic difficulties

Case presentation

A 34 year old male presented with complaints of ulcer on lateral aspect of the tongue for the last 2 months. History revealed that the patient was operated for tongue malignancy at the same site three years back and was diagnosed as sarcomatoid squamous cell carcinoma with vascular malformation. The ulcer recurred after two years and was re-operated at a second health center with the histopathologic diagnosis of pleomorphic sarcoma, suggestive of liposarcoma of tongue with hyperplastic squamous epithelium.

Local examination showed an ulcer of approximately 1cm diameter, with scar mark on the left lateral border of the tongue. Local excision was done and tissue submitted for histopathologic diagnosis. Haematoxylin and Eosin stained tissue sections showed proliferation of blood vessels of variable size, lined by anaplastic endothelial cells with plenty of fibroblastic cell proliferation. (Figure 1) Foci of tumor giant cells and areas of pyogenic necrosis was also seen.

The overlying squamous epithelium showed marked acanthosis with elongated rete pegs and severe dysplasia of the basal lamina presenting as elongated navicular cells. Sub-epithelial tissue showed mixed inflammatory cell infiltrate, comprising mainly of lymphocytes and neutrophils. Epithelial findings suggested dysplasia of the tumor overlying squamous epithelium, pathognomonic of an early marjolin’s change. The underlying muscles showed mild chronic inflammatory infiltration. Reticulin and Van Geison stains demonstrated reticulin and collagen fibre synthesis (Figures 2 & 3).

FIG. 1: Photomicrograph shows proliferation of blood vessels of variable size, lined by anaplastic endothelial cells. H&E(120X).
AHMAD et al.: Angiofibrosarcoma of the tongue

FIG. 2: Photomicrograph shows reticulin fibre production around the tumor cells. Reticulin stain (500X)

FIG. 3: Photomicrograph shows collagen production by the proliferating fibroblastic tumor cells. Van Geison stain (120X).

Immuno-histochemical stain smooth muscle actin (SMA) showed strong positivity of tumor cells. (Figure 4)

FIG. 4: Photomicrograph shows tumor cells positive for SMA. SMA (500X).

The histopathological and immune-histo-chemical findings were diagnostic of Angio-fibrosarcoma of the tongue. Our patient is symptom free for the last 6 months on follow up.

Discussion

Angiosarcoma is malignant lesion of vascular endothelial origin characterized by rapidly proliferating, extensively infiltrating anaplastic cells lining irregular blood-filled spaces. This neoplasm contributes less than 1.0% of all the soft tissue sarcomas. Malignant Hemangioendothelioma, Angioblastoma, Hemangiosarcoma are terms used for angiosarcoma. Radiation, trauma and lymphedema are related to the cause of angiosarcoma.

Skin and superficial soft tissue is the most common site involved by angiosarcoma. Head and neck region is rarely involved. Oral cavity comprises of less than 8.0% of head and neck angiosarcomas. Most cases involve the mandible, but other less frequent sites of involvement are maxilla, parotid glands, lips, tongue, floor of mouth, cheek, palate and antrum. Harter in 1927 reported for the first time, a case of angiosarcoma of tongue. Very few cases of angiosarcoma of tongue have been reported in Indian literatures. Rao et al. in 1986 reported a case of primary angiosarcoma of tongue. Chadha et al. in 2012 reported another case of primary angiosarcoma of tongue.

Histopathology of angiosarcoma is markedly variable, so the role of immunohistochemistry is very important to arrive at a correct diagnosis. Various patterns have been identified in angiosarcoma, such as well-formed blood vessels, solid pattern or cells in sheath, and cells showing poor differentiation or undifferentiated type or a mixed pattern. Low grade, differentiated angiosarcomas mimic the normal vascular endothelial cells but poorly differentiated tumors may resemble a carcinoma. Vascular anastomosing channels are lined by atypical endothelium, which can be a single row of cells, but cellular proliferation often results in a thick endothelium. The highly cellular tumors show marked multiplicity of endothelial cells into the vascular lumen, leading to a solid growth pattern. As described by Campagnacci et al., low grade tumors show areas of well-formed capillaries with large flattened cells, containing nuclei of moderate size and demonstrating infrequent mitoses. Higher grade lesions have areas of hemorrhage, disordered architecture, and large cells with hyperchromatic, pleomorphic nuclei and frequent mitoses.

Tongue is commonly involved by hemangioma, hemangioendothelioma, hemangiopericytoma, epithelioid sarcoma, kaposi's sarcoma, malignant melanoma, anaplastic carcinoma and pyogenic granuloma. Therefore these lesions should be differentiated from Angiosarcoma. Angiosarcomas can be differentiated from the benign mimics by vascular endothelial atypia, atypical mitotic activity and infiltrative growth pattern. The lack of hyaline globules in the tumor cell cytoplasm with spindled cells having well-defined cell borders and isolated foci of plasma cells distinguishes angiosarcoma from Kaposi's sarcoma. It is very difficult to separate angiosarcoma from the epithelioid subtype of hemangioendothelioma and the low grade variants. A very distinctive growth pattern of cords to sheets of epithelioid cells with a distinctive hyaline stroma helps to differentiate the latter lesion.

Immuno-histo-chemical staining can be used to confirm the diagnosis. To differentiate angiosarcoma from the mimickers, markers such as Pan Cytokeratin, S-100, Human melanoma antibody (HMB-45) can be used. The vascular im-
mune-markers separate angiosarcoma from carcinoma and melanoma, although keratin may be observed in epithelioid endothelial tumors. CD31 is the best vascular marker for endothelium especially in the poorly differentiated angiosarcoma. Other marker which also show positivity are Ulex europaeus, Factor VIII, CD 34, Vimentin and Smooth muscle actin.12,13

Conflict of interest

The authors declare that they have no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

References