CASE REPORT

Pseudovascular adenoid squamous cell carcinoma of oral cavity: A mimicker of angiosarcoma

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ABSTRACT

Pseudovascular adenoid squamous cell carcinoma (PASCC) is an uncommon histological variant of squamous cell carcinoma that can mimic vascular neoplasms, particularly angiosarcoma, in its morphologic characteristics. PASCC has been reported in the head and neck, as well as in the other organs such as the breast, lungs, urinary bladder, vulva, and uterine cervix. Only two cases of PASCC arising from the upper aerodigestive tract have been reported so far. We report a case of PASCC of oral cavity in a 40-year-old man, which mimicked an angiosarcoma initially. Immunohistochemical analysis led to a conclusive diagnosis of PASCC.

Key words: Angiosarcoma, pseudo vascular, adenoid squamous cell carcinomaoral cavity

INTRODUCTION

Pseudovascular adenoid squamous cell carcinoma (PASCC) is an uncommon histological variant of squamous cell carcinoma (SCC), characterized by acantholysis of the tumor cells, leading to the formation of anastomosing spaces and channels, mimicking an angiosarcoma.[1] PASCC has been reported in the head and neck,[2] as well as in the other organs, such as the breast,[3] lungs,[4] urinary bladder,[5] vulva, and uterine cervix.[6] Only two cases of PASCC arising from the upper aerodigestive tract have been reported so far.[7]

CASE REPORT

A 40-year-old man presented to the Department of ENT with a fast-growing polypoid tumor on the right side floor of the mouth. He was a smoker but denied alcohol abuse. Biopsy showed features of moderately differentiated SCC. Wide excision of the tumor with right hemimandibulectomy and modified radical neck dissection was performed. On macroscopic examination, the right hemimandibulectomy specimen showed a polypoid mass measuring 6 × 6 × 3 cm at the right gingivo–buccal sulcus [Figure 1]. Surface of the growth showed necrotic areas. Cut section showed gray-brown and gray-white areas. Modified radical neck dissection showed 18 lymph nodes, largest measuring 1 × 1 cm, with gray-white to gray-brown areas.

On microscopic examination, the tumor was composed of vessel-like anastomosing channels, which were lined by a single layer of atypical epithelioid cells, along with dilated and congested blood vessels [Figures 2 and 3]. The lumina of these spaces contained pinkish material, few erythrocytes, and rare acantholytic tumor cells. There were no vascular/lymphatic emboli. Seven out of 18 lymph nodes from modified radical neck dissection showed tumor deposits of similar looking cells. A differential diagnosis of SCC and angiosarcoma was considered due to the presence of anastomosing channels of cells.

Immunohistochemical analysis showed strong positivity for cytokeratin in the tumor cells. Vimentin showed focal positivity, but the tumor cells were negative for CD 34 [Figure 4]. Hence, angiosarcoma was ruled out and a diagnosis of PASCC [$T_3N_2M_0$] was made.

DISCUSSION

The most common cancer in the upper aerodigestive tract is conventional SCC. Variants of SCC, which occur less frequently are verrucous carcinoma, spindle SCC, papillary SCC, basaloid SCC, adenoid SCC, lymphoepithelial carcinoma, and adenosquamous SCC. Their recognition is

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Figure 1: Excised exophytic polypoid growth on the gingivo buccal sulcus

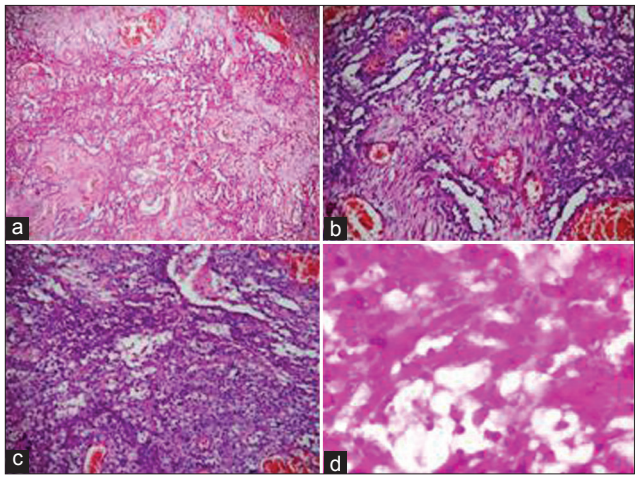

Figure 3: (a, b) Tumor showing areas of reactive fibrosis; (c) Tumor showing areas with acantholytic pattern (H and E, x100); (d) Lumina of pseudovascular spaces contain acantholytic tumor cells and erythrocytes (H and E, ×400)

important because most of them are true clinicopathological entities, with an important prognostic implication. Moreover, they may mimic other neoplasms, resulting in erroneous treatment.[7]

PASCC is a rare variant of adenoid SCC. The adenoid form of SCC is a neoplasm that is characterized by the fact that it presents a pseudoglandular pattern. The biological phenomenon that explains this histological pattern is acantholysis, which when massive may mimic a vascular proliferation and hence is known as pseudovascular adenoid SCC. Hence, PASCC is also termed as angiosarcoma-like SCC/pseudoangiosarcomatous SCC. Less than 30 cases of PASCC in various sites have been documented in international literature so far.[8]

PASCC has the clinical characteristics of SCC, but histologically may mimic an angiosarcoma. Interestingly in the oral cavity, both these entities show a comparable clinical appearance.[8] The peak incidence of angiosarcoma is in the 7th decade and that of PASCC is in the 6th decade.[8]

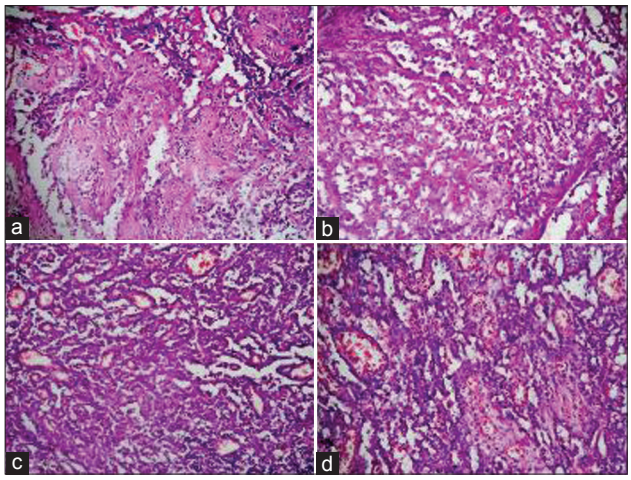

Figure 2: (a, b) Tumor with anastomosing vessel-like channels, lined by a single layer of malignant cells (c, d) showing tumor with dilated and congested blood vessels (H and E, ×100)

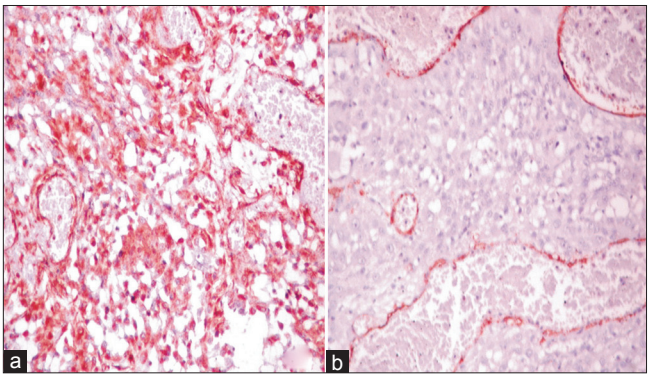

Figure 4: (a) Immunohistochemical analysis showed tumor cells strongly positivity for cytokeratin (b) Tumor cells are negative for CD34

Macroscopically, both these entities are seen as fast-growing eruptive lesions in the oral cavity and have a poor prognosis.[8]

Histologically, angiosarcoma consists of interanastomosing vascular spaces lined by malignant cells, which are spindle shaped or epithelioid in shape; whereas, in PASCC, massive acantholysis in the tumor gives the appearance of pseudovascular spaces. Occasionally, reactive fibrosis within the tumor, superimposed upon the acantholytic pattern, results in artifactual clefts lined by poorly differentiated malignant cells, creating an angiomatous appearance.[4] The histologic similarity of PASCC and angiosarcoma has been emphasized by several authors. Fanberg-Smith *et al.* in their study found that one-third of oral angiosarcomas were of the epithelioid type, which may be mistaken for other epithelioid tumors.[9] Immunohistochemistry is very helpful in differentiating both these entities, because the epidermoid differentiation in PASCC may be extremely masked by pseudovascular proliferation.

With the onset of cytological techniques, the correct identification of PASCC and its distinction from angiosarcoma is very important. Aspiration of PASCC yields individually

scattered and syncytial fragments of malignant epithelioid appearing cells with oval to spindle shapes, prominent nucleoli, and a finely vacuolated amphophilic cytoplasm.[4] Aspirates of angiosarcoma are typically cellular and associated with abundant blood. Malignant cells are seen singly and in clusters or arranged in tight aggregates, loose groups, and parallel bundles of streaming malignant cells resembling abortive capillaries. A careful search will reveal the association of malignant cells with vascular lumina, indicating their endothelial origin.[4] In the absence of these vasoformative structures, angiosarcoma may be misinterpreted as poorly differentiated carcinoma, PASCC, or other sarcomas.[4,10] Ancillary immunohistochemical studies and clinical history are essential in such cases to arrive at a correct diagnosis.

On immunohistochemistry, PASCC will usually express epithelial markers such as cytokeratin and epithelial membrane antigens, whereas angiosarcoma typically expresses vascular antigens – CD 31, CD 34, and von Willebrand factor, which are not expressed in adenoid SCC.[2] Cytokeratin may, however, be positive in some cases of angiosarcoma.[11] Recently, Fli-1 protein, a member of the ETS family of DNA binding transcription factors, has been highlighted as a new vascular differentiation marker, which is positive in angiosarcoma.[8] PASCC has been found to be negative for Fli-1 protein. Another new marker, Laminin-5, was found to be positive in the cytoplasm of PASCC cells and negative in angiosarcoma. Laminin-5 also acts as a tumor biological indicator of the unfavorable prognosis of PASCC.[8]

The exact pathogenesis of PASCC is not completely understood. Acantholysis is suggested to be the underlying pathogenetic mechanism, possibly as a consequence of changes in adhesion molecule expression by the tumor cells.[7] This can be observed as loss of expression of adhesion molecule E- cadherin, which is one of the major adhesion molecules on the epithelial cells. b- Catenin- E- cadherin complex, which is present on the cell membrane and which mediates cell-to-cell adhesion, is obviously disturbed in PASCC and this is responsible for forming of typical intercellular spaces.[12] Studies have shown that E- cadherin is expressed in most SCC in the head and neck, the expression being strong in well-differentiated cancers, but reduced in poorly differentiated tumors.[7]

PASCC is almost always accompanied by foci of conventional SCC, suggesting the correct diagnosis. The problem arises in biopsies, when the presence of a pseudovascular component only may be misinterpreted as an angiosarcoma. In the present case, biopsy was suggestive of a moderately differentiated SCC, whereas the tumor itself showed features of PASCC, thus simulating a vascular neoplasm. But a review of the biopsy, along with immunohistochemistry of the tumor proper, helped us to come to the correct diagnosis.

The etiology and prognosis of PASCC are probably similar to adenoid SCC than conventional SCC. Some authors believe that PASCC has a more aggressive behavior than conventional SCC. But the number of patients reported so far is too small to draw firm conclusions.[7]

In conclusion, although the prognostic importance of PASCC in the head and neck is still ill understood, its recognition is important because it may mimic angiosarcoma, which may result in erroneous treatment.

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