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"CARCINOSARCOMA OF ANTERIOR MAXILLA: A MISDIAGNOSIS OR MISSED DIAGNOSIS? "

Dental Science

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ABSTRACT

Carcinosarcoma is an exceedingly rare biphasic tumor affecting upper aerodigestive tract, reveals both carcinomatous and sarcomatous elements dominating later one. In head and neck most common site of involvement is larynx (vocal cord) and hypopharynx with a very rare occurrence in oral cavity. It has male predilection with alcohol abuse, cigarette smoking, poor oral hygiene and previous radiotherapy as predisposing factors. Aggressive behaviour, high recurrence rate and metastasis of tumor makes diagnosis critical. Owing to its rarity, histological and immunohistochemical features simulates with other benign and malignant spindle cell lesions, poses diagnostic challenge. Here we are presenting a rare case of 32 years old female, who was diagnosed histologically with Glomus tumor on incisional biopsy and myoepithelioma on resection but recurrence occurred within 60 days posing a malignancy with a final diagnosis of carcinosarcoma confirmed by immunohistochemistry.

KEYWORDS

INTRODUCTION:

Carcinosarcoma is a rare tumour expressing biphasic features (mesenchymal and epithelial) with distinguished hallmarks of malignancy. Virchow first coined the term "Carcinosarcoma" instead of its previous names, pseudosarcoma, spindle cell carcinoma and sarcomatoid carcinoma. It is a true malignant mixed tumor.1 The carcinomatous component varies, but can be composed of adenocarcinoma,squamous cell carcinoma,or undifferentiated carcinoma.They may also show specific salivary carcinoma phenotypes,including salivary duct carcinoma or adenoid cystic carcinoma.Sarcomatous elements can also be variable, including chondrosarcoma, osteosarcoma, fibrosarcoma, and malignant fibrous histiocytoma. Most frequently, the sarcomatous component dominates, though the two elements can be found in an intermixed pattern.2 Theses tumours are characterized by primitive mesenchymal blastic differentiation into multiple cell lines although true sarcoma remains the main cell line with high mitotic index, cellular pleomorphism, dysplasia and lack of transition zone.3 The WHO labelled this entity as spindle cell carcinoma and classified it under malignant epithelial tumors of squamous cell carcinoma. (Parikh N,Desai N.Spindle cell carcinoma of the oral cavity : A case report of a rare entity and review of literature. J academy Adv Dental Research 2011;2:31-36.). In head and neck most common site of involvement is larynx (vocal cord) and hypo pharynx with a very rare occurrence in oral cavity. It also occurs in the salivary gland, comprising 0.04% to 0.16%of all salivary gland tumors and 0.4% of all malignant salivary gland neoplasms.4 In oral cavity they might be situated within the hard and soft palates, cheeks, lips, tongue and tonsillar crypts with very few cases documented in the literature.5 They are characterized by high rates of recurrence, metastatic spread, and mortality.6

The case presented here is of 32 yrs old female patient diagnosed with a Carcinosarcoma of epithelial cell origin and discusses the importance of Diagnosis and treatment of this rare clinico-pathologic entity. Initially, three different diagnoses were made in dilemma to reach out the final diagnosis, Which was confirmed by immune-histochemistry.

CASE REPORT:

A 32 year old female patient came to our Department of Oral & Maxillofacial surgery with chief complaint of swelling in mid-palatine region since 5-6 months and pain in the same region since1-2 months which was gradual in onset,dull-aching and continuous in nature, radiating to right preauricular and post-auricular region, aggravates on mastication and relieves on its own.No H/O pus discharge/ trauma/bleeding/parasthesia or numbness .H/O single episode off fever one month back. H/O burning sensation at the site of swelling.No H/O weight loss or change in voice or dysphagia. Patient gave history of tobacco chewing 3-4 times a day since 10-11 years.Clinically a roughly oval,well defined polypoidal mass of size 4x3 cms approx was seen in hard palate extending mesiodistally from 23 to16 and supero-inferiorly from the hard palate to 2cm below incisal level.Surface of mass was ulcerated and covered with yellowish white slough. On palpation, mass was non tender,soft to firm inconsistency and bleeds on provocation.Panoramic radiography revealed ill-defined alveo crestal bone loss from premolar to premolar region in maxilla.

Occlusal radiograph of maxilla was taken which also revealed alveo crestal bone loss.

An incisional biopsy was performed in which sheets of small round cells with vesicular to hyperchromatic nuclei and granular cytoplasm was seen.

At places tumor cell showed pleomorphic oval to spindle cell population er chromatism.The tumor cells were intermixed with fibro vascular connective tissue stroma.A diagnosis of Glomus tumor was made.A computed tomographic(CT)scan revealed multiple calcified granulomas in the temporo-parietal region with strongly enhancing lobulated mass of size 30 x 35 mm attached to the hard palate causing focal destruction of the Hard palate suggestive of Carcinoma?

Ultrasonography (USG) report suggested of a large soft tissue mass lesion in the hard palate predominantly on the right side with moderate vascularity and with significant arterio-venous shunting and the blood supply was from right internal maxillary artery and facial artery which

were dilated and tortuous.Embolization of bilateral maxillary arteries,right facial and upper labial branches were done using gelsponge.

As the glomus tumor is benign only surgical excision of lesion along with Extraction of upper right 12, 13 ,14 was done under general anesthesia. The growth was excised smoothly and total mass came out in toto.

The histopathological features of resected mass showed proliferation of spindle cells in the form of fascicles and numerous plasma cytoid cells suggesting diagnosis of "myoepithelioma".

However, patient reported back in the span of 45 days with recurrence of Similar type of growth at the same site of size 2 x 2.5 cm approximately.

again patient was subjected for surgical resection under general anesthesia .As the tumor recurred so fast ,considering it to behaving malignant potential a subtotal maxillectomy was done from 16 to 28 followed by palatal obturator.

The histopathological features of resected mass revealed a malignant tumor with dual population of cells .Proliferation of spindled and epithelioid cells was seen beneath ulcerated squamous mucosa.The tumor cell shows vesicular nuclei with prominent nucleoli and eosinophilic cytoplasm. numerous plasma cytoid cells were seen. examined tissue were subjected to immunohistochemistry to eliminate the diagnostic dialemma. The tumor cells expressed positivity for pan-cytokeratin ,high molecular weight cyto keratin [marker for epithelial origin],Vimentin [marker for mesenchymal origin],Epithelial Membrane Antigen(EMA).The tumor cells were stained negative for Smooth muscle actin(SMA) [marker for muscle origin],p63,S-100 protein [marker for neural origin and salivary gland neoplasms], Human melanoma black (HMB-45),Melan-A [marker for melanocytes], MUM-1[marker for Follicular lymphoma],CD30 & ERG[markers for endothelial origin].

As tumor cells expressed both cytokeratin as well as vimentin and its Biphasic pattern of both epithelioid and spindle cells in histopathology Made a final diagnosis of Carcinosarcoma of squamous cell origin which is exceptionally unusal.

DISCUSSION:

Carcino sarcoma is a poorly differentiated variant of squamous cell carcinoma (E.B. StelowandS. E.Mills,"Squamouscell carcinomavaria ntsoftheupperaerodigestivetract,"

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Pathology,vol.124,supplement,pp.S96–S109,2005.) with a more aggressive behaviour (S.B.

Minami,S.Shinden,andT.Yamashita,"Spindlecellcarcinomaofthepalat inetonsil:reportofadiagnosticpitfalland literaturereview,"
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Modern
Pathology,vol.15,no.3,pp.229–254,2002.).It is a biphasic tumour composed of both malignant epithelial and mesenchymal components. The initial description of this entity was given by Virchow in 1864 7 and it accounts for less than 1 % of all tumors of oral cavity.8The tumor is prevalent in the age ranges from 47 to 88 years,with maximum number of patients in the fifth and sixth decades with a median age of 65.7 years and there is male predilection.(Parikh N,Desai N.Spindle cell

carcinoma of the oral cavity:A case report of a rare entity and review of literature.J academy Adv Dental Research 2011;2:31-36.). In contrast, present case reported in 32 years and is female patient.Duration of symptoms ranges from 20 days to 2 years with less than 1 year In 95 % of patients. The predisposing risk factors are tobacco chewing (68%), smoking (21%) alcohol (12%) and exposure to radiation in rare instances.9 Present case came with a complaint from last six month and had a history of tobacco chewing which some what relates etiology.

In the oral cavity the most common presentation varies from exophytic Polypoid mass with a nulcerated surface to a frankly infiltrative ulcer, Mostly involving alveolar ridge, lip, tongue, buccal mucosa and only few

Cases occurs in maxilla. (Parikh N,Desai N. Spindle cell carcinoma of the oral cavity:

A case report of a rare entity and review of literature. J academy Adv Dental Research 2011 ;2:31-36.).Reported cases hows polypoid growth with ulcerated surface At unusual site i.e.palate.

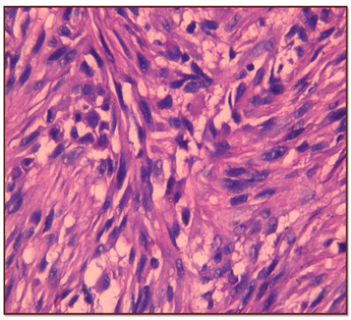
Clinically they tend to present at an advanced stage,with most tumors Being T3 or T4 at presentation and pursuing an aggressive course.10 Leventon et al. Have proposed depth of invasion by the tumor to be the Most important prognostic criterion, with deeper infiltration indicating a
Poorer survival.11Present case is also T4 at the time of presentation.
The histogenesis of carcinosarcoma is still controversial. No definitive Evidence exists to show whether this tumor arises from two distinct Carcinoma and sarcoma clones or from a single malignant clone that is Capable of both epithelial and mesenchymal differentiation. Sometimes the transition between carcinomatous and sarcomatous components can be observed in a single patient, and some of the spindle cells are immunoreactive to the epithelial markers. This suggests that carcinosarcomas may develop from a totipotent cell that is capable of both epithelial and mesenchymal differentiation.12 It is the tumor appropriately described in the epithelial-mesenchymal transition (EMT) theory.Through EMT,the malignant epithelial cells that trans differentiate into myofibroblasts lose their malignant phenotype,but they still retain the desmoplastic stroma that is essential for tumor proliferation and metastasis.In general, carcinosarcomas ,which very likely exemplify EMT, are highly infiltrative and metastasizing,and their behaviour is similar to that of poorly differentiated carcinomas.13

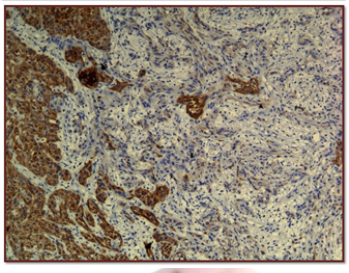
They may show either monophasic or biphasic cells Histopathologically & proliferation of both spindle cells and epitheliod cells14 while,myoepitheliomas show spindle cell proliferation in fasciculated pattern.Immuno histochemically myoepitheliomas will express positivity for S100,Vimentin,SMA,p63 and diffusely cyto keratin while carcinosarcoma will not show positivity for any of the markers except vimentin and it will show strong positivity for cytokeratin.15 Present case expressed positivity for both cytokeratin and vimentin.So,a diagnosis of carcinosarcoma was made in collaboration with histopathological features.Histopathologically, both spindle cells and epithelioid cells with nuclear and cellular pleomorphism with significant mitoses were seen in present case. Carcinosarcomas are exceedingly rare,and therefore a well established therapeutic approach is lacking.Current recommendations for the treatment of this entity include radical surgical resection,lymphnode dissection for palpable lymphadenopathy, and radiation therapy.16 Numerous authors attribute the majority of failed carcinosarcoma treatments to the use of radiation as the primary therapy,and, for cases in which surgery is the primary treatment,to incomplete resection.17 In our case,a misdiagnosis of Glomus tumor was made at the time of Incisional biopsy.So an incomplete resection was planned.USG report revealed hypervascularity of tumor and hence,the patient underwent preoperative endovascular therapeutic embolization.On resection,it came out in toto.Bone resorption was seen in maxilla.On histopathological examination of resected mass a diagnosis of myoepithelioma was made,as proliferation of spindle cells infasciculated pattern along with numerous plasmacytoid cells were seen.But the tumor recurred within a span of one and half month which made us suspect the malignancy.Immuno histochemistry was done which lead us to the final diagnosis of Carcinosarcoma.Prognosis is related to location, tumor size, depth of invasion,stage of disease.A tumor of the oral cavity and oropharynx is potentially aggressive and seems to recur easily and to metastasize easily.18 The incidence of metastases was 36% and the 2-year survival rate was 55 % in tumors involving the oral cavity.19

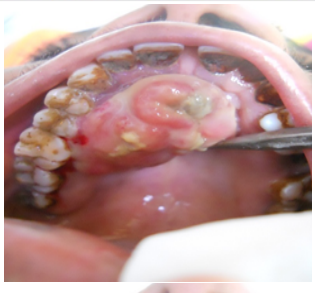
CONCLUSION

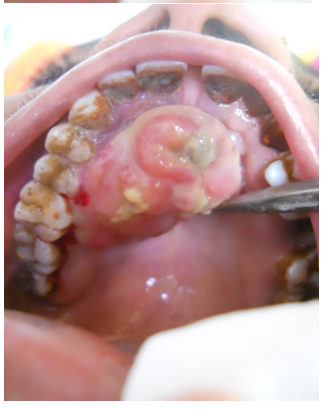
To conclude,such sporadic and complex entities pose diagnostic dilemmas, diagnosis of carcinosarcomas in the head and neck region is challenging because of overlapping histo pathological features with other spindle cell tumors.Understanding their clinical,morphological and immunohistochemical features with a systematic approach is critical for their accurate diagnosis which aids in correct patient management.

Moreover,each aspect of patient management merits full consideration, Decisively bearing in mind the risk-benefitration of the case in hand.









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