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Case Report

Sinonasal biphenotypic sarcoma of uvula: the first case report

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ABSTRACT

Biphenotypic sinonasal sarcoma is a rare entity and it is a new term included in literature recently. It affects sinonasal tract exclusively but we report the first case of biphenotypic sinonasal sarcoma of uvula. The patient is a 56-year-old male who had a mass in the oropharynx hanging from the uvula exclusively. Contrast enhanced computed tomography of oral cavity reported homogenously enhancing benign mass arising from uvula probably pleomorphic adenoma. It was excised in toto and sent for histopathological examination. The detailed histopathological analysis concluded the diagnosis as sinonasal biphenotypic sarcoma of uvula. Sinonasal biphenotypic sarcoma is exclusively found in sinonasal tract. Occurrence of this entity in uvula is first of its kind and needs special attention.

Keywords: Uvular mass, Biphenotypic sarcoma

INTRODUCTION

Biphenotypic sinonasal sarcoma is an uncommon low-grade sarcoma with neural and myogenic differentiation. First described in 2012 by Lewis et al, it is seen to exclusively affect the sinonasal area. Biphenotypic sinonasal sarcoma (BSNS) has a female predilection and affects individuals in their sixth decade and involves the superior nasal cavity and ethmoid sinus more commonly.[1]

Due to histological overlap with other tumors, it has previously been misdiagnosed as fibrosarcoma, leiomyosarcoma, malignant peripheral nerve sheath tumor, cellular schwannoma, or synovial sarcoma.[2-5]

Uvular enlargement is quite rare but can rapidly progress to upper airway obstruction and respiratory distress.[6] Tonsillitis, peritonsillar abscess, allergies are common causes of uvular enlargement. Primary squamous cell carcinoma of the uvula has been well documented.[7]

To the best of our knowledge, this is the first case of BSNS involving the uvula.

CASE REPORT

A 56-year-old male patient presented to ear nose and throat (ENT) outpatient department (OPD) with chief complaints of difficulty in swallowing since 1 month and change in voice since 14 days. Dysphagia was gradual in onset and progressive and more to solids than liquids. The patient noticed a heavier and throatier voice than before. There was no complaint of any difficulty in breathing or oro-nasal bleeding and no history of alcohol or tobacco intake. On oral cavity examination, an oval pinkish mass approximately 3 by 3 cm in dimension arising from uvula was seen hanging in the oropharynx (Figure 1). The mass was pedunculated with a stalk attached exclusively to the uvula. It was a fleshy lobulated mass with smooth surface moving up with deglutition when its free lower edge was seen obscuring the view of endolarynx. On 70-degree endoscopy the scope was negotiated bypassing the swelling and it was found that the mass had no other attachment other than uvula. The endo larynx was visualized and found to be normal. The patient underwent a contrast enhanced computed tomography (CECT) of oral cavity and neck which reported a well-defined lobulated homogenously enhancing soft tissue pedunculated mass

lesion originating from uvula filling oropharynx (Figure 2). A provisional diagnosis of a benign lesion most likely a pleomorphic adenoma was made. Owing to the clinicoradiological benign appearance of the mass, surgical excision with grossly safe appearing margin of 1 cm was done per orally at uvular base under general anesthesia (Figure 3). Patient was allowed clear fluids after 8 hours of surgery and cold soft diet from day 2 of surgery. The specimen was subjected to histopathological examination.

Histopathological examination

The postoperative specimen was globular, grey-white soft tissue measuring 3.5×2.5×2 cm with smooth external surface, which on cutting open showed a solid lesion with intermittent areas of hemorrhage. Histopathological examination revealed a tumor comprising of sheets of long, spindled out cells with focal fasciculation and whorls. The cells had pale eosinophilic to clear cytoplasm, ovoid to round nuclei, high nuclear to cytoplasmic ratio, prominent nucleoli and frequent mitosis of 1-2/high power field. Epithelioid appearance was appreciated in some foci. Focal areas of necrosis and hemorrhage were also seen along with surface ulceration. A panel of immunohistochemical stains was performed to exclude carcinoma and melanoma; and to subtype the sarcoma. The tumour cells showed immunoexpression for S100, smooth muscle actin (SMA), h caldesmon and focal desmin. The cells were negative for cytokeratin (CK), epithelial membrane antigen (EMA), CD34, HMB45 and myogenin. Nuclear expression of beta catenin, however, was not seen. Based on co-expression of S100, SMA, h-caldesmon and focal desmin, and spindle-ovoid cell morphology, a diagnosis of biphenotypic sarcoma was suggested (Figure 4). However, cytogenetics and molecular analysis could not be performed on account of lack of resources.

Table 1: Differential diagnosis of sinonasal biphenotypic sarcoma.

Diagnosis	Morphology on H&E	Immunohistochemistry	Treatment
Monophasic synovial sarcoma	Hypercellular sheets of monotonous spindle cells with frequent mitosis	CK (+), EMA (+), vimentin (+), S100 (-)	Wide local excision with radiotherapy
Spindle cell carcinoma	Fascicles of highly pleomorphic cells (variable epithelial differentiation) with high mitotic index in collagenous matrix. Storiform pattern can be seen	CK (+) and vimentin (+)	Surgery with or without radiotherapy
Spindle cell melanoma	Spindle shaped melanocytes with variable pleomorphism and atypia and high mitotic index	Vimentin (+), S-100 (+), HMB-45 (+)	Wide surgical excision with adjuvant targeted therapy/ chemo/ radiotherapy
Leiomyosarcoma	Fascicles of high grade spindle cells with cigar shaped nuclei and eosinophilic fibrillary cytoplasm	SMA (+), Desmin (+), S100 (-)	Wide local excision
Glomangioperi-cytoma	Spindle cells with perivascular hyalinization and staghorn vessels, low grade morphology	SMA (+), nuclear beta catenin (+), S100 (-)	Complete surgical excision
Solitary fibrous tumor	Ovoid to spindle cells arranged haphazardly and in short fascicles, pericytomatous pattern, ropey collagen bundles	CD34 (+), SMA (-), S100 (-)	Compete surgical excision

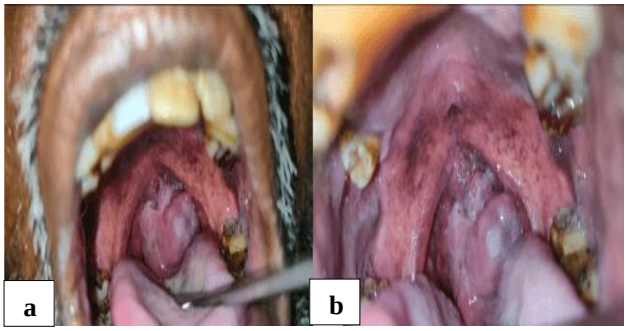

Figure 1: (a) and (b) Showing a smooth oropharynx mass attached to uvula and hanging in.

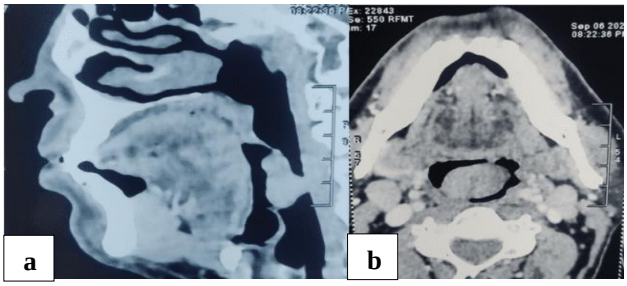

Figure 2: Contrast enhanced computed tomography shows homogenously enhancing mass arising from uvula hanging in oropharynx, (a) sagittal view and (b) coronal view.

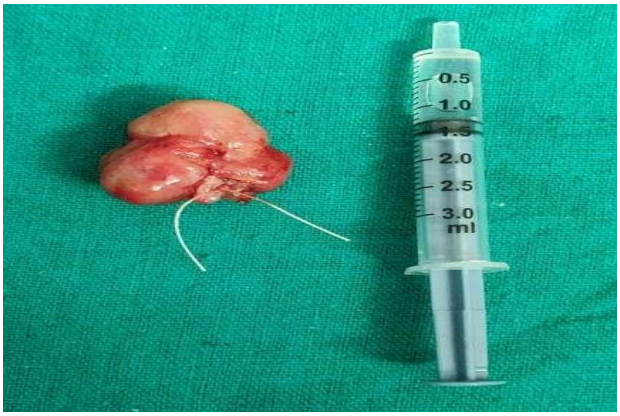

Figure 3: Post-operative specimen showing complete excision of tumour with 1 cm uvular margin shown by suture tie.

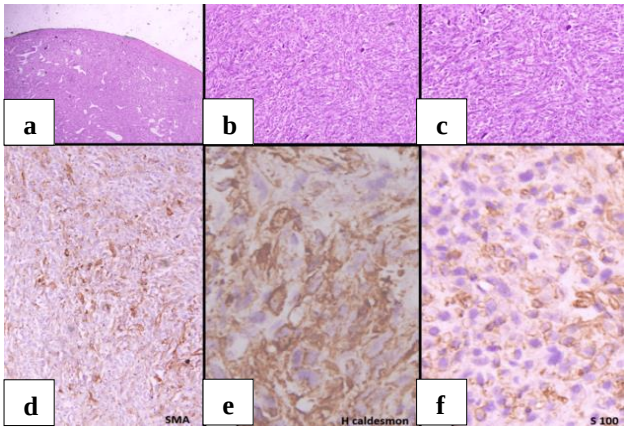

Figure 4: (a) Periphery of tumour shows well circumscription and pushing appearance, monomorphic spindle and epithelioid cells with compressed staghorn like appearance of blood vessels-hemangiopericytomatous pattern (haematoxylin & eosin x 100); (b) and (c) spindle and epithelioid appearance of the tumour cells with compressed interspersed blood vessels; and (d)-(f) tumour shows immunohistochemical positivity to smooth muscle actin (SMA), H Caldesmon, S 100.

DISCUSSION

Biphenotypic sarcoma is a new entity which was previously categorised with other soft tissue tumours. The disease occurs most commonly in fifth decade with female predominance and presents with symptoms related to nasal passage obstruction. Local invasion and intracranial extension can also be seen. Due to high propensity of such tumours to arise from sinonasal region, it was termed as biphenotypic sinonasal sarcoma (BSNS).[8] Although around 100 cases have been reported so far (common sites being nasal cavity and sinuses), origin of BSNS from uvula has not been reported before.

Microscopically, these sarcomas tend to show a low grade spindle cell morphology with fascicular arrangement. Stromal collagen, focal herring bone/ hemangiopericytomatous pattern are other features that can be seen.[9] The present case showed sheets and fascicles of long spindle cells with focal epithelioid morphology.

Immunohistochemistry is of paramount importance in diagnosing and differentiating biphenotypic sarcomas from other more common entities in this region. Rooper et al proposed an extensive immunohistochemistry panel due to the rarity and novelty of this entity and considerable morphologic overlap with other sarcomas that includes S100, SOX10, smooth muscle actin, calponin, desmin, myogenin, β-catenin, factor XIIIa, and cytokeratin. Biphenotypic sarcomas show dual expression of S100 and SMA universally, pointing towards dual neural and myogenic differentiation. Besides, these sarcomas can also show variable immunoexpression for desmin, myogenin, factor 8A, PAX8, nuclear beta catenin and CD34.[10] Few cases have also been reported with focal rhabdomyoblastic differentiation on morphology with nuclear myo-D and myogenin expression.[11,12] Loarer et al described 27 cases of biphenotypic sarcoma with focal immunoexpresson of Myo-D1 in the absence of morphologic evidence of rhabdomyoblastic differentiation.[13] Our case had no rhabdomyoblastic differentiation on morphology or on IHC. Rooper et al and Kakkar et al showed 10/11 and 5/6 cases to have focal nuclear expression of beta catenin however, Sethi et al described 3 case of BSNS with absence of nuclear beta catenin expression.[10,14,15] The current case also lacked beta catenin expression. A plethora of differential diagnosis were considered and ruled out one by one the basis of morphology and IHC. These included spindle cell carcinoma (co-expression of CK and vimentin), spindle cell melanoma (vimentin, S-100 and HMB-45 expression), synovial sarcoma (co-expression of CK and vimentin), leiomyosarcoma (SMA positive and negative for S100), glomangiopericytomas (perivascular hyalinisation and positive for SMA and nuclear beta catenin but negative for S100) and solitary fibrous tumor (CD34+) (Table 1).

Biphenotypic sarcomas characteristically demonstrate rearrangements of PAX3 with multiple fusion partners, the most common of which is MAML3.[12] Due to constraint of resources, cytogenetic analysis could not be done. Biphenotypic sinonasal sarcomas are treated by local excision, with or without adjuvant radiation treatment although the efficacy of adjuvant therapy versus re-excision is unknown.[9] Our patient underwent surgical excision with safe margin intraoperatively and histopathologically the margin is free of tumour by 0.7 cm. Considering the acceptable tumour free margin of resection and relatively safe location of tumour, the patient was not given adjuvant radiation therapy and advised for long follow up.

CONCLUSION

It is imperative to recognise the lesion as its natural course is completely different from its other counterparts. Its

proper identification is essential for correct diagnosis and treatment.

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