Alveolar soft part sarcoma of tongue base — a rare presentation of a rare tumor

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Abstract

Alveolar soft part sarcoma is a rare, aggressive malignancy of uncertain histologic origin and enigmatic clinical behaviour. It has a characteristic histopathological picture, with a propensity for vascular invasion and distant metastasis. We report a case of alveolar soft part sarcoma involving the tongue base in an adolescent female. She underwent laser assisted excision of the tongue base tumour followed by post-operative radiotherapy. The clinical presentation, histopathological picture, immunohistochemical & cytogenetic studies, radio-imaging, management protocols and prognosis of this tumor have been discussed.

Keywords Lingual Alveolar Soft Part Sarcoma (ASPS) · Immunohistochemistry

Introduction

Alveolar soft part sarcoma (ASPS) is a rare malignant tumor of connective tissues with an unclear aetiopathogenesis. It was coined by Christopherson et al in 1952 [1]. It is among the least common of soft tissue sarcomas. Its incidence has been reported to be less than 1% of all primary soft tissue malignant neoplasms. This extremely vascular tumor is most commonly found in the lower extremities (44%). About 27% of ASPS occur in the head and neck region, primarily in children, the common sites being tongue, larynx, oropharynx and orbit [2, 3]. Lingual ASPS represents 10% of all ASPS. ASPS of the tongue is a slow-growing, painless tumor that typically occurs in females aged between 15 and 35 years. This lesion may mimic benign vascular neoplasms, but careful evaluation of the unique imaging features seen on CT scans, MR images and angiograms lead to the correct diagnosis.⁽⁴⁾ International literature so far reports nearly 60 cases of Lingual ASPS [11, 12, 13].

Case Report

A 13-year-old girl presented to us with complaints of slowly progressive dysphagia for solids and odynophagia along with a *hot-potato* voice of 4 months duration. She had episodes of blood stained sputum, often following vigorous brushing and hawking habits. Her oropharyngeal examination revealed a soft, pinkish smooth surfaced mass of size 2 x 2.5 cms arising from the base of tongue in midline, extending onto both valleculae. All other otolaryngological and head & neck examination were unremarkable, except for intense oropharyngeal erythema. The clinical picture was suggestive of lingual thyroid due to its characteristic location. Her hematological & thyroid function tests were normal. A Technetium 99 Scan revealed normal functioning thyroid tissue in the neck. There was no uptake by the tongue base mass.

A contrast CT of the oropharynx showed a much larger (4 x 3.5 cms) well-circumscribed, heterogeneously

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enhancing lesion within the tongue, mistaken to be a haemangioma by the radiologist. Further assessment with MR imaging & Angiography showed heterogeneous enhancement of the lesion due to high vascularity, along with flow voids suggestive of areas of central necrosis within the tumor. No definitive feeding vessels could be identified by the MR Angiography. Extensive pre-operative investigations such as, CT neck, CT chest, abdominal ultrasound and nuclear medicine scans were performed for tumor staging and no distant metastases were noted. The tumor was stage grouped as T3 N0 M0 (Stage III) as per the UICC nomenclature. As the tumour was highly vascular no tissue biopsy was attempted.

In view of the superficial extension of the lesion onto the surface of the base of tongue & both valleculae, laser resection of this vascular lesion was planned after preparation for blood transfusion on table. Using Boyle Davis mouth gag with a small tongue blade, KTP/532 laser with 9 watts power settings a wide excision of the entire mass with a cuff of normal uninvolved tissue all around, was performed with a hand held probe. On gross examination after surgical resection, the tumor was noted to be highly vascular, well circumscribed and encapsulated. The excised tissue was sent for histopathological examination & immunohistochemical studies. The patient was kept on Ryle's tube feeds for two weeks & recovered well post-operatively with improved speech and swallowing. She remained asymptomatic on clinical examination at 6 months follow-up.

The histopathologic findings were suggestive of a Granular cell tumour, with nests of large granular cells separated by fibrovascular stroma. The neoplasm showed vascular invasion within the margins of the specimen. The cells were PAS positive with diastase-resistant intracytoplasmic inclusions. Specimen analysis with Immunohistochemistry proved positive for Desmin, Muscle specific Actin and Sarcomeric Actin suggestive of myogenic origin of the tumor. Immunohistochemical stains for HMW, LMW keratins, Neuron specific enolase and S-100 were negative. Evidence of sarcomatous constituents within the tumor, provided the pathological diagnosis of Alveolar soft part sarcoma. The case was discussed with radiation oncologist & a post-operative course of radiotherapy (4.5 grays delivered in 6 cycles) was given. The patient had an uneventful recovery period & is presently symptom free at six months follow up.

Discussion

Alveolar soft part sarcoma is a slow growing but highly malignant tumour. It typically presents as a slowly enlarging, well-circumscribed and often encapsulated mass. It has uncertain histogenesis & predominantly affects female children and adolescents. Earlier ASPS was considered to be of neural origin, but more recent immunohistochemical studies have proved it to be a tumour of uncertain differentiation [5].

The median age group is around 15–35 years and there is female preponderance before the age 30 and a slight male predominance over age 30.

The tumor is grossly well circumscribed & highly vascular. Microscopically, it is characterized by a strikingly uniform pseudo-alveolar pattern of polygonal cells, that contain characteristic cytoplasmic crystalline inclusion bodies [5]. In the head & neck regions, it may present in the orbit, oropharynx, larynx & laryngopharynx [2, 3]. It has a particular affinity to metastasize to the lungs & brain. In many cases, metastases in the lungs or brain are the first manifestation. The incidence of metastases seems to be highest in the lungs (38%) followed by the brain (33%). It may present with multiple lymph nodes, presumably even while the sarcoma itself may be still small & undiagnosed. Cervical lymph node metastasis involvement occurs in about 7% of cases of head & neck ASPS [4, 7]. Local recurrence after surgery has been reported in upto 20% of cases. Metastases of this tumor can occur more than ten years after primary tumor removal & radiation. This makes evaluation of the curative treatment & prognosis of ASPS extremely difficult [9].

ASPS derives its name from its histologic appearance and suspected origin. As a result of formalin fixation, granular cells that are normally arranged in well-defined nests separated by a rich vascular network lose cohesion and create a pseudoalveolar pattern. These large cells may resemble gemistocytic astrocytes, atypical neurons, or metastatic renal cell carcinoma [7]. The nuclei of these tumor cells appear uniform and vesicular with prominent nucleoli showing the characteristic *apple core* concave indentation. The histogenesis remains controversial and this tumour is classified under *Tumours of uncertain differentiation* in the 2002 WHO classification of soft tissue tumors. There are no consistently positive findings on IHC. Desmin is sometimes positive, particularly on frozen sections. There is often cytoplasmic reactivity only for Myo D1. Myogenin staining is consistently negative. In 25% of cases there is S100 protein or NSE positivity. There is no expression of chromogranin, NF protein. CK or EMA. ASPS are deceptively benign histologically with no atypical mitotic figures or nuclear pleomorphism. Demonstration of PAS-positive diastase-resistant crystals with the appropriate clinical history confirms the diagnosis [5, 7]. Cytogenetic analysis of soft part sarcomas consistently show the chromosomal translocation abnormality of der(17) t(X;17)(p11;q25) wherein 2 gene loci are involved, namely TFE3 (Location Xp11) and ASPSCR1 (Location 17q25).

Lingual ASPS usually presents with a mass in base of tongue associated with dysphonia, odynophagia & a varying degree of dysphagia. Clinical examination often reveals pulsation with a thrill and may mimic vascular malformations, which may lead to misdiagnosis and inadequate or delayed treatment. Although there is no overlying ulceration or bleeding, significant hemorrhage can occur as the tumor grossly extends through the overlying mucosa [10].

On MRI, most of the alveolar soft part sarcomas demonstrate increased signal intensity on both T1W and T2W images with flow voids in the center and at the margins of the tumor [2]. The recognition of these characteristic MR findings may lead to the early diagnosis of ASPS, especially when the clinical presentation is that of a slow growing soft tissue mass in young adult patients. In addition to hemangioma and vascular malformation, other less likely differential diagnoses include hyperplastic lingual thyroid, dermoid, and squamous cell carcinoma. These neoplasms are easily differentiated from ASPS on the basis of CT and MR findings. Lingual thyroid typically occurs in the posterior oral tongue and does not have areas of necrosis or flow voids. Although rare, dermoids of the tongue appear as hypoattenuated and well-circumscribed masses on CT images that may contain fat and typically show peripheral enhancement of the cystic wall. Squamous cell carcinoma of the tongue accounts for nearly one quarter of oral cavity cancer but typically does not have well-circumscribed borders, flow voids, intense enhancement, and hyperintense signal on T2-weighted images as seen in ASPS [2, 4, 7].

Aggressive laserization of the lingual mass with wide margins, using CO₂, Nd-YAG or KTP/532 laser have proven the logical method for complete tumor clearance with low morbidity rates as compared to conventional surgical procedures wherein haemorrhage can be a major intra-operative complication [8, 11, 12]. KTP/532 laser has become an ideal solution to deal with such well circumscribed vascular tumors of the oropharynx [9, 10]. Massive bleeding during surgical resection of ASPS has been reported and when located posteriorly in the tongue may be difficult to control at surgery. Preoperative embolization for ASPS is useful to control blood loss and the need for transfusion therapy. Aggressive surgical excision is recommended, because local recurrence is common [9, 10]. The role of post-operative radiotherapy has remained limited. Chemotherapy is reserved for pulmonary metastasis. Prognosis is guarded, as 5-year and 20-year survival rates are reported to be 59% and 15%, respectively, with local recurrence and distant metastasis primarily responsible for the poor prognosis of this neoplasm [5, 7, 9].

Conclusion

Lingual ASPS should be considered in the differential diagnosis of a slow growing hypervascular soft tissue mass in a young adult. CT and MR imaging features of ASPS suggest the correct diagnosis and will help the surgeon in

performing a wide surgical resection to reduce the risk of local recurrence. Preoperative staging proves vital due to the frequency of metastasis at presentation and the relatively poor prognosis of ASPS. Long term post-operative follow-up is mandatory for all patients with this deceptively indolent and highly malignant neoplasm.

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