Alveolar soft-part sarcoma of the tongue : report of a case

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Abstract

Alveolar soft-part sarcoma(ASPS) is a rare, aggressive malignancy of uncertain histologic origin with a propensity for vascular invasion and distant metastasis. ASPS may mimic benign vascular neoplams of malformation but careful evaluation of the unique imaging features on CT scans, MR images, and angiograms lead to the correct diagnosis. ASPS of the tongue is slow-growing, painless mass, especially ASPS of the base the tongue is difficult to be noticed by patient, dentists or oral and maxillofacial surgeons on oral examintion because of its location and clinical resemblance to a benign lesion. And it leads to delayed or inadequate diagnosis.

We report radiologic and clinical features of an ASPS of the basal portion of the tongue in a 17-year-old boy, showing normal appearance, but palpation of the tongue and floor of the mouth reveals the tumor. Among the 23 cases of a primary ASPS of tongue reported, 7 cases occured on the basal region of the tongue, inculding the present one. There has been no recurrence or metastasis as of 3 years postoperatively.

Key words
Alveolar soft-part sarcoma, Tongue

Alveolar soft-part sarcoma(ASPS) is clinically and morphologically distinct soft tissue sarcoma that was first described as an entity in 1952 by Christopherson and his co-workers[1]. It is a rare, aggressive malignancy of uncertain histologic origin with a propensity for vascular invasion and distant metastasis. This extremely vascular tumor accounts for less than 0.5-1% of soft-tissue sarcomas, and seen most commonly in adolescents and young adults. It is somethat more frequent in women than in men, the ratio approximately being 2:1. The tumor is primarily found within the skeletal muscles or musculofascial planes of the extremities and rarely arises in the head and neck,especially the tongue[2-6]. Among the 23 cases of ASPS of the tongue reported, 7 cases occurred on the basal region of the tongue, including the present one[7,8].

ASPS of the tongue is slow-growing, painless mass, may mimic benign vascular neoplasms of malformations but careful evaluation of the unique imaging features on CT scans, MR images, and angiograms lead to the correct diagnosis[9]. This article focuses on clinical features of ASPS of the basal portion of the tongue to be noticed incidentally on palpation by patient's dentist and emphasize the unique imaging features that led to correct diagnosis.

REPORT OF A CASE

A 17-year-old boy in good general health was referred to us with a painless swelling of his tongue. The swelling had been noticed incidentally by his dentist, he being unaware of it. On oral examination there was a large pulsatile mass which was elastic, soft, round, circumscribed but not tender, measured approximately 3cm in diameter, in the left side on the tongue. The tongue was normal in color, no ulceration or hemorrhage was present(Fig. 1). There was no cervical lymph node enlargement. General examination was enssentially within normal limits. Since this was thought to be a hemangioma, needle

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aspiration was perfomed and blood was aspirated.

CT scans showed 3.5×3.7×4.3cm round hypervascular tumor in anterior portion of mouth floor with early washout, and multiple tortuous vascular channels was noted in adjacent areas, mistakenly believed to be a simple hemangioma(Fig. 2). MR imaging showed high signal intensity on unenhanced T2-weighted images and isosignal on T1-weighted images. Numerous signal void structures within the mass and dilated vascular structures around the mass were seen, which indicated hypervascular mass. It showed homogeneous enhancement on postcontrast T1-weighted images, however, intensity of enhancement was not so strong. It was suggested to be attributed to arteriovenous shunting in the tumor. It has well-defined margin and there showed no evidence of infiltration to adjacent structures(Fig. 3-5). Preoperative angiography and embolization were performed. Angiography showed a large hypervasuclar mass predominately supplied from the left lingual artery with small contribution from the right lingual artery, where the greatest bulk of tumor was(Fig. 6,7). Tumor embolization was accomplished by polyvinyl alcohol particles and coil.

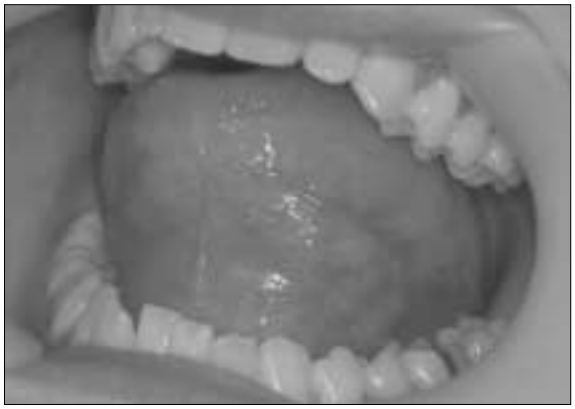

Fig. 1. Clinical appearance of the tongue. On palpation there was a pulsatile mass which was elastic, round and circumscribed, measured approximately 3cm in diameter, in the left side on the tongue.

Since the lesion was thought to be benign, we decided to excise the tumor by an intraoral approach. An incision approximately 3cm in length was made along the floor of mouth from the left side of the tongue extending to midlilne. By alternative sharp and blunt dissection, a portion of the lesion was exposed, vessels on the surface of the tumor was clamped and ligated and a specimen was removed by sharp dissection in one piece(Fig. 8). The tumor looked encapsulated and was readily separated from the surrounding tongue musculature. The postoperative course was uneventful, the patient did not require transfusion during the perioperative period. Histological examination showed large round or polygo-

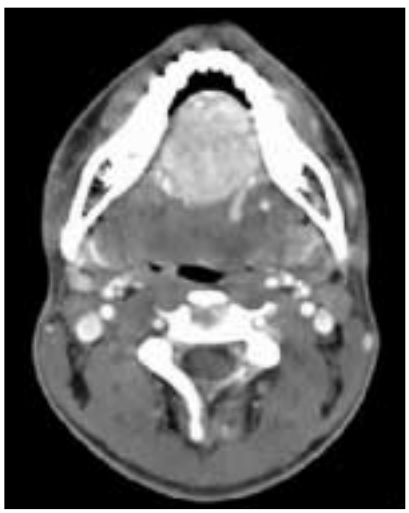

Fig. 2. Axial contrast-enhanced CT shows a well-circumscribed hypervascular tongue mass.

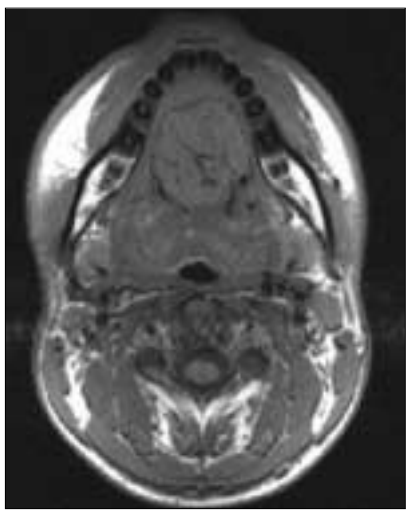

Fig. 3. Axial unenhanced T1-weighted image shows a isointensity signal lesion in tongue. Signal void structures are seen within the mass.

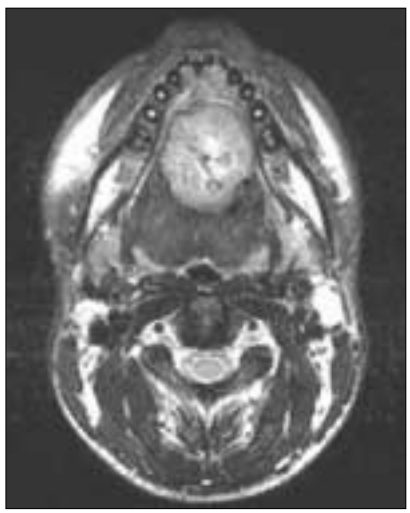

Fig. 4. Axial unenhanced T2-weighted image shows a lesion with high signal intensity. Signal void structures are also seen within the lesion. The lesion has well-defined margin, suggesting that it is not infiltrated to adjacent structures.

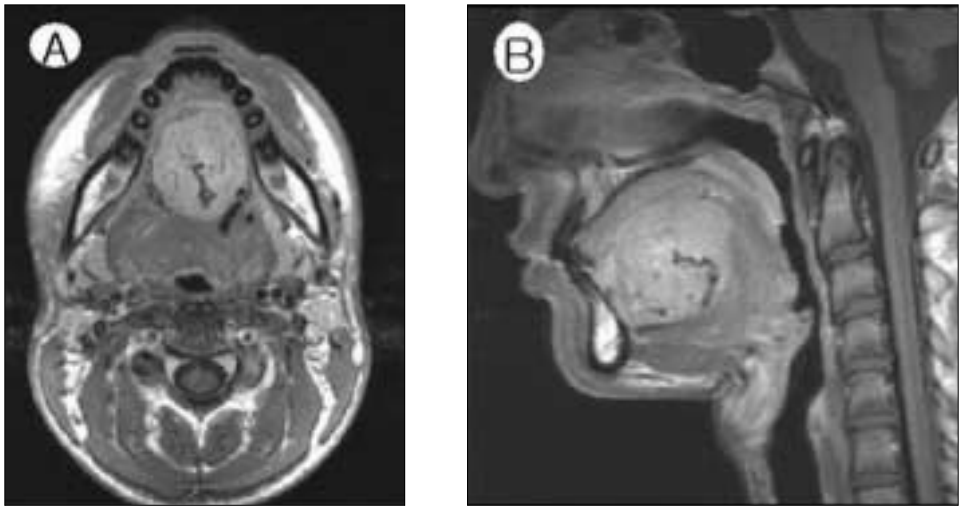

Fig. 5. Postcontrast T1-weighted axial and sagittal images show intense enhancement with numerous feeding vessels. Signal void structures are once again seen.

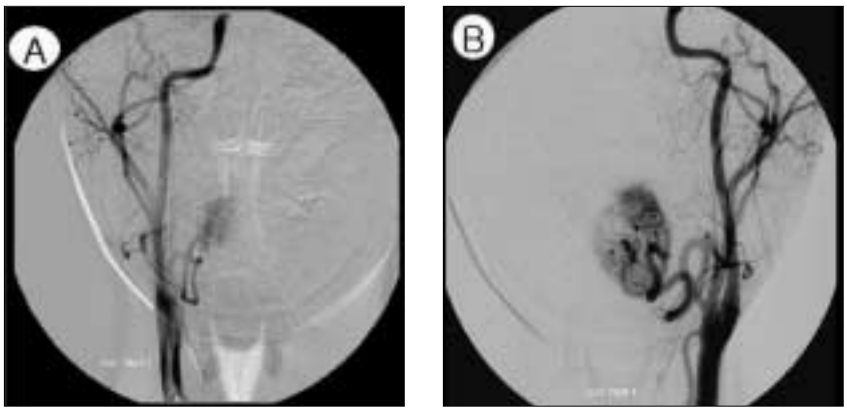

Fig. 6. A-P projection angiograms show large hypervasuclar mass predominately supplied from the left lingual artery with small contribution from the right lingual artery.

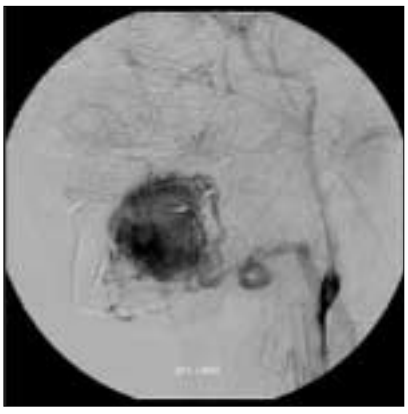

Fig. 7. Lateral-projection angiogram shows a hypervascular mass with irregular, tortuous vessels.

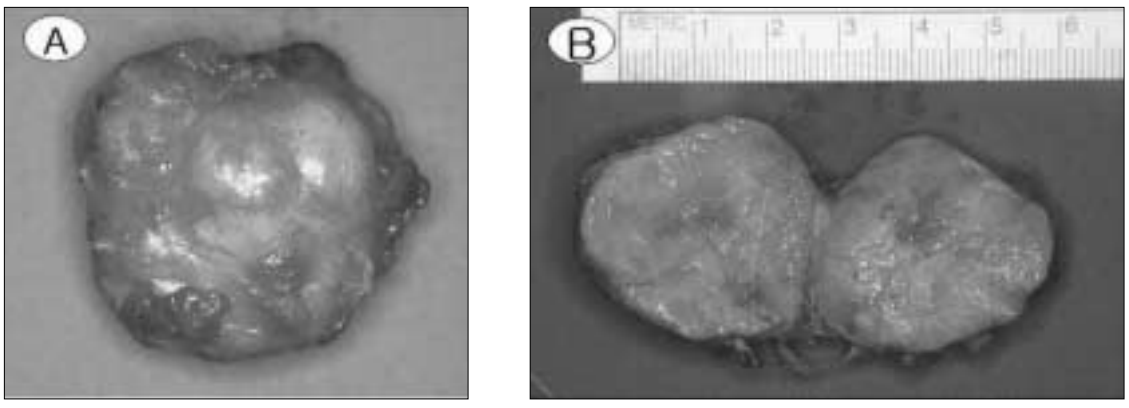

Fig. 8. The size of the resected tumor was 4×3.5×3 cm. It was round and well circumscribed.

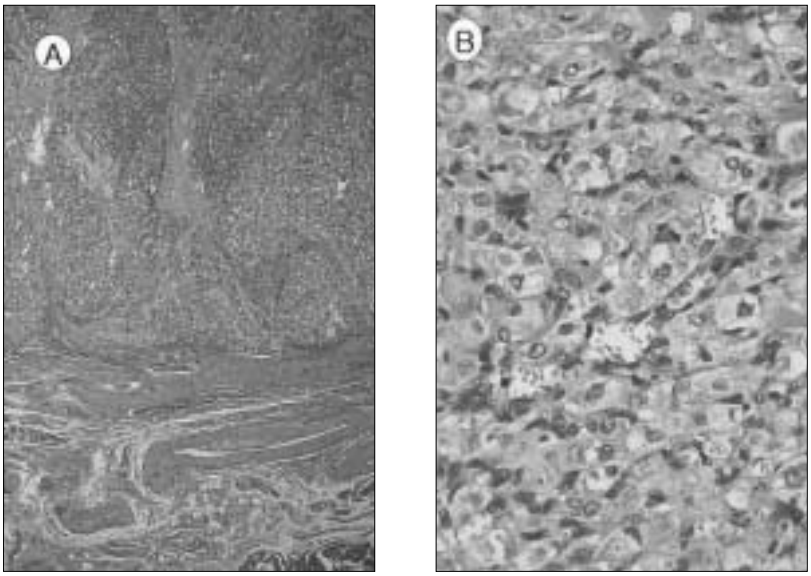

Fig. 9. A, Photomicrograph shows soild sheets of tumor cells, typical of alveolar soft-part sarcoma(Hematoxylin and eosin stain,original magnification ×40). B, Photomicrograph shows large round or polygonal tumor cells with vesicular nuclei and prominent nucleoli (Hematoxylin and eosin stain, original magnification ×400).

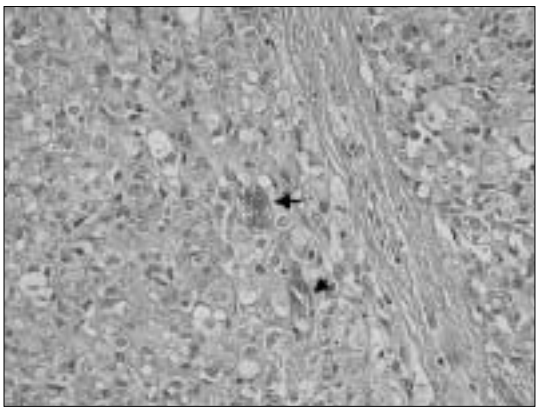

Fig. 10. PAS stain after diastase digestion shows rod-shaped crystalline inclusions in the cytoplasm, diagnostic of ASPS(arrows,original magnification ×400).

nal tumor cells with vesicular nuclei and prominent nucleoli, and solid sheets of tumor cells. PAS(periodic acid-Schiff) stain after diastase digestion showed rod-shaped crystalline inclusion in the cytoplasm. A final diagnosis of ASPS was made(Fig. 9, 10). There were no evidence of metastatic spread on chest and abdominal CT examination. Because of close margins, it were necessary to do a wide excision. However his parents refused further surgery. The patient had postoperative radiation therapy in a specialized center. There has been no recurrence or metastasis as of 3 years postoperatively.

DISCUSSION

ASPS derives its name from its histologic appearance and suspected origin[1]. Histolgically, the tumor shows variably sized alveolar structures circumscribed by delicate vascular channels and bands of fine connective tissue[5]. ASPS has a close clinical and imaging resemblance to common benign vascular tumors such as hemangioma, which may lead to misdiagnosis and inadequate or delayed treatment. Benign vascular soft-tissue vascular lesions are best classified into two categories, hemangioma and vascular malformation. The term "hemangioma" is restricted to the classic involuting vascular lesion of infancy. Vasuclar malformations are divided into low-flow and high-flow subtypes, characterized by the predominant vessel involved; namely, capillary, venous, or lymphatic involvement. Vascular malformations are present at birth, grow in proportion to patient growth, and do not involute. Rarely, they may spontaneously enlarge through the development of arteriove-

nous fistulas, thrombosis, or ectasia. In contrast, ASPS in not present at birth, grow slowly, and occurs in an older age group[9].

The following is a brief summary of radiographic images of ASPS, based on recent reports[7-10]. Unenhanced CT imaging of ASPS show a low-attenuation lesion similar to muscle with peripheral enhancement. In contrast-enhanced CT, it may show significant enhancement at the periphery of the lesion, together with central low-attenuation areas, it demonstrate necrosis of central areas. But our patient did not have central necrosis. Several imaging features help distinguish ASPS from benign hemangiomas and venous malformations(cavernous hemangioma). CT imaging of hemangioma in contrast typically shows a slightly hyperattenuated lesion on unenhanced images, with intense homogeneous enhancement after contrast material administration. Hemangiomas and venous malformation may also contain phleboliths seen as punctate calcifications on unenhanced CT images. MR images of ASPS show an signal intensity greater than muscle but less than fat. Unenhanced T1-weighted images typically show hyperintense signal as compared muscle that is not as intense than that seen with hemangiomas but more intense than that of other soft tissue sarcomas that are typically isointense relative to muscle, together with signal void structures, are characteristic of this lesion on MRI. Similar to hemangioma, ASPS has very high signal intensity on unenhanced T2-weighted images, but signal void structures are also seen. Postcontrast MR images show intense, heterogenous or homogenous enhancement. Our case showed homogenous enhancement. In case of tumor necrosis, a central region of low signal intensity may be seen on pre- and postcontrast T1-weighted images, with corresponding high signal intensity on T2-weighted images. No tumor necrosis was seen in our patient.

Angiography shows a hypervascular mass with ateriovenous shunting in the arterial phase, followed by intense tumor staining by the contrast medium. Multiple, enlarged and tortuous vessels also seen. In spite of the rapid shunting of blood from the lesion, "wash out" of the contrast material from the lesions was slow. This is contrast to a high-flow vascular malformation such as an arteriovenous malformation that has and arteriovenous shunt but no persistent tumor blush and a low-flow vascular malformation that has prolonged contrast blush but no arteriovenous shunt. Massive bleeding during surgical resection of ASPS has been reported and when located posteriorly in the tongue may be difficult to control at surgery[11,12]. Preoperative embolization for ASPS is useful to control blood loss and the need for transfusion therapy.

No definitive conclusions about treatment of head and neck ASPS can be reached because of the rarity of the disease and the lack of reported follow-up. However, surgery appears to be cornerstone of therapy in most cases, varying from simple to radical excision. If excision is adequate, local recurrence in not a major problem and there is no advantage in radical surgery[13-15]. The role of chemotherapy and radiotherapy is unclear, although they are recommended by several authors[15-17]. The prognosis depends to a large extent on appropriate management of metastasis, which are unpredictable and can be delayed up to three decades after primary diagnosis of the tumor, the lungs are the most common site[6,14].

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