Lingual Alveolar Soft Part Sarcoma (ASPS): A Case Report and Literature Review

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Introduction

Alveolar soft part sarcoma (ASPS) was first described as soft tissue malignancy in 1952 by Christopherson et al as a tumour of uncertain histogenesis. Its histogenesis is still not entirely clear. However, the advent of modern immunohistochemistry and development of new tissue markers elucidated its origin to certain extent. Most cases of alveolar soft part sarcoma have been reported in the deep soft tissues of adolescents commonly in the upper half of the lower extremities. The lingual alveolar soft part sarcoma is extremely rare, and most of the head and neck tumours of this type had been reported in children. The rare sites include retroperitoneum, stomach, uterus, pulmonary vein, lung, sacrum, pituitary gland and breast. Herein, a case report describes alveolar soft part sarcoma, arising in the posterior part of a tongue.

Clinical Feature

Eighteen years old Iraqi was transferred to Salmaniya Medical Complex, Kingdom of Bahrain, for elective surgical treatment, who presented with two years history of a slowly growing lingual tumour, which eventually extended into the right side of the neck.

Gross Pathological Examination

Specimen received included tongue, right mandible, submandibular gland, external jugular vein, skeletal muscle and cervical lymph nodes. There was a fleshy greyish tumour measuring 8 X 7 cm, arising in the dorsum of the tongue and has infiltrated the neck muscle, submandibular gland, external jugular vein and surrounded the mandible on its medial aspect. All the resection margins were clear by at least 2 cm. Multiple blocks were submitted for histological examination including five blocks from the tumour. Sixteen cervical lymph nodes were identified; however, it was difficult to orientate the lymph nodes according to neck lymphatic drainage compartments. This was largely due to extensive direct infiltration by the tumour.

Microscopic Features

The tumour revealed partly alveolar and partly solid and organoid growth patterns, comprising nests of large malignant cells. The nests of tumour cells were surrounded by delicate fibrovascular septa, containing occasional thin walled vascular channels. The tumour cells were polygonal and rounded having vesicular nuclei with prominent nucleoli, and abundant eosinophilic granular cytoplasm (Figure 1). Some of the tumour cells had vacuolated cytoplasm. Mild nuclear pleomorphism was noted. Mitotic activity was noticeable (four mitoses per 10 HPF). Intracytoplasmic crystalline material was identified using PAS stain with prior diastase treatment (Figure 2). There was extensive vascular invasion including external jugular vein (Figure 3).

Immunohistochemistry showed positive cytoplasmic granular staining pattern of the tumour cells for Myo D1 (figure 4) and NSE (Figure 5) and focal cytoplasmic positivity for Desmine (Figure 6). The tumour cells were negative for alpha Smooth Muscle Actin, pan-cytokeratin marker MNF116, S-100...
protein, HMB-45 and chromogranin. Electron microscopy was not performed.

Fig. 1: Alveolar Soft Part Sarcoma showing Typical Alveolar Pattern, comprising Large Cells of Uniform Sizes with Mild Nuclear Pleomorphism. The Cells are dissociated in the Centres of the Tumour Nests. H&E. Magnification X 400.

Fig. 2: Note the Vascular Invasion by a Cluster of Tumour Cells. H&E. Magnification X 200.

Fig. 3: Cytoplasmic Crystalline Material Diagnostic for Alveolar Soft Part Sarcoma. PASD. Magnification X 400.

Discussion

Lingual alveolar soft part sarcoma is an extremely rare entity. Literature search indicates that only 5% of the cases of alveolar soft part sarcomas are lingual\textsuperscript{16}. The common location is the deep soft tissues of the extremities\textsuperscript{17}. Although the commonest among head and neck ASPS is lingual\textsuperscript{18}.

Lingual ASPS commonly presented in a younger age group as compared to the ASPS at other anatomical sites. The median age for the lingual ASPS reported is five years ranges from 3 to 21 years\textsuperscript{16}. In contrast, the current case presented at the age of eighteen. However, 70% of ASPS located at anatomical sites other than tongue, presented above the age of 20 years\textsuperscript{19}.

The lingual ASPS reports have shown a preponderance towards solid and organoid growth pattern associated with little alveolar pattern\textsuperscript{20, 21, 22}. Similar findings were observed in this case and cytological features were consistent with the cases reported in the literature. However, the mitotic activity was more, which is considered as a rare event in such cases.

Vascular invasion is a constant feature in all lingual alveolar soft part sarcomas as compared to the non-lingual ASPS. There is no correlation between the vascular invasion and long term survival in cases of lingual ASPS\textsuperscript{16}. This may perhaps be due to smaller tumour size (median 25mm) and early diagnosis, resulting in prompt surgical excisions. The tumour is much larger (80mm) in the current case, which is thought to be due to a delay in excess to the diagnostic and surgical treatment facilities.

The main differential diagnoses included salveolar Rhabdomyosarcoma, extra-adrenal malignant pheochromocytoma and malignant melanoma. Positive staining pattern of the tumour cells for MyoD1, NSE and Desmin supported the diagnosis of alveolar soft part sarcoma. Negative staining pattern for chromogranin, MNF116, HMB45 and S-100 protein excluded the latter two differential diagnoses. However, the immunohistochemical staining pattern of alveolar soft part sarcoma is similar to alveolar rhabdomyosarcoma. The diagnosis of alveolar soft part sarcoma was preferred over alveolar rhabdomyosarcoma due to consistent presence of large cells in all sections from the tumour. In contrast alveolar rhabdomyosarcomas always show large majority of small to medium sized cells with occasional presence of large multinucleated giant cells. This may explain the limitations of immunohistochemistry in establishing the diagnosis of alveolar soft part sarcoma in particular and sarcomas in general. Contrary to the statement, immunohistochemistry is a useful tool in excluding other differential diagnoses such as extra-adrenal pheochromocytoma and malignant melanoma, which have more specific immunophenotypes. This may emphasis a point that one has to resort to conventional
morphological criteria in establishing sarcomas diagnosis in general and alveolar soft part sarcoma in particular.

Molecular analysis may contribute to the diagnosis of alveolar soft part sarcoma, which has a characteristic translocation der(17)t(X;17)(p11.2;q25)\(^{23,24}\).

**Fig. 4:** MyoD1 shows Diffuse Granular Cytoplasmic and Focal Nuclear Staining of the Tumour Cells. Magnification X 400.

**Fig. 5:** NSE is Positive in all Tumour Cells. Magnification X 400.

**Fig. 6:** Desmin shows Focal Positivity of the Tumour Cells. Magnification X 400.
Adequate surgical excision with wide margins is a treatment of choice in most of the reported cases. A series of fourteen cases has shown that complete surgical excision alone was a treatment of choice and adjuvant chemotherapy was given to two cases. All those patients had relatively good outcome. It is difficult to predict good prognosis in this case, simply due to larger tumour size and extensive infiltration into the surrounding neck structures.

References

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