Review

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Alveolar soft-part sarcoma of the oral cavity: A review of literature

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Abstract

An alveolar soft-part sarcoma is a malignant neoplasm primarily affecting the soft tissues of head and neck. The aim of the present review is to systematically present the demographic and clinico-pathological data of articles published in the English medical literature. A comprehensive search of the databases (PubMed, Medline, SCOPUS, Web of Science, and Google Scholar) along with cross references to the published articles on alveolar soft-part sarcoma for eligible studies/case reports published since 1957 till date was done to retrieve the data. A total of 74 cases were identified and analyzed from 42 papers published in the English medical literature. All the clinical, radiographic, and prognostic features were analyzed and presented along with the treatment strategies. Alveolar soft-part sarcoma is a rare and aggressive malignancy of uncertain histologic origin with a propensity for vascular invasion and distant metastasis. This neoplasm requires careful clinical, radiographic, and histopathologic evaluation to reach to the correct diagnosis.

Keywords

Alveolar soft-part sarcoma, alveolar soft sarcomas, connective tissue tumors, soft-tissue tumors

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Introduction

Alveolar soft-part sarcoma (ASPS), with an ICD-O code of 9581/3,[1] accounts for less than 1% of soft-tissue sarcomas.[2] This extremely vascular tumor arises in 27% of head and neck cases, out of which 25% cases occur in tongue.[3] Christopherson et al.[4] were the first to coin the term and describe the lesion in 1952. Although ASPS may occur at any age, it demonstrates a strong predilection for adolescents and young adults.[5] ASPS accounts for 5% of all pediatric soft-tissue sarcomas other than rhabdomyosarcomas.[6]

Vascular invasion is frequent, and metastatic disease is frequently present at the diagnosis of ASPS. Lung metastases are seen in 42%–65% of patients, while brain and bone are the next most common sites of metastasis, with lymph node involvement seen in only 10% of patients.[7] 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.[3]

Overall survival rates have been reported to be 62% after 5 years, 42% after 10 years, and 18% after 20 years. ASPS accounts for 5% of all pediatric soft-tissue sarcomas other than rhabdomyosarcomas.[8] No specific treatment

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protocols have been developed, which makes its management difficult especially in children.

The aim of the present review was to comprehensively appraise the clinical, radiological, histopathological, histogenetic, and therapeutic aspects of ASPS and thus attempt to further speculate on the possible biologic profile of the tumor to enhance knowledge about this unusual malignancy.

Methods

Search strategy and selection criteria

This systematic review was conducted according to the guidelines of the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement. We performed a comprehensive search of the databases (PubMed, Medline, SCOPUS, Web of Science, and Google Scholar) along with cross references to the published articles on ASPS for eligible studies/case reports published since 1957 till date. Keywords included a combination of "alveolar soft-part sarcomas (ASPS)," "alveolar soft sarcomas," "connective tissue tumors," and/or "soft-tissue tumors." Additional citations identified through the reference lists of selected references and bibliographic linkages were included in the review. Journals related to subjects such as oral pathology, oral surgery, and oral medicine were also searched for aforementioned keywords. Inclusion criterion consisted of human case reports on ASPS, and extra-oral, metastatic, recurrent cases were excluded from the study. For the analysis, the following clinico-histopathologic data were pooled: age, gender, onset, duration, size, location, shape, color, tenderness, fixation, overlying mucosa, histopathologic characteristics, radiographic presentation, immunohistochemical observations, metastasis, treatment rendered, and recurrence status along with its genetics.

Results and discussion

In the present review, a total of 74 cases were identified and evaluated from 42 research papers published in the English medical literature (Table 1). The clinico-pathological data are summarized in Table 2.

Clinical features

Incidence. ASPS is a rare aggressive sarcoma accounting for less than 1% of soft-tissue sarcomas.[2] According to Aiken and Stone,[3] this extremely vascular tumor is most commonly found in the lower extremities (44%) and arises in the head and neck in 27% of cases, with 25% of head and neck cases occurring in the tongue. It accounts for 0.1% of sarcomas of the head and neck. In children and infants, the most common sites of occurrence are orbit (41%) and the tongue (25%).[5]

Age and gender distribution. Wang et al.[30] have studied 18 cases of ASPS and found a mean age of 20.2 years with an age range of 3–61 years, and there was a slight female predilection (M:F ratio of 1:1.25). However, Fanburg-Smith et al.[16] studied 14 cases and found a mean age of 5 years with an age range of 3–21 years. Male predilection was observed with male-to-female ratio of 1.3:1. While in our analysis, we have found out an age range of 1.5–64 years with a mean of 18.25 ± 15.243 years. There were 31 (41.9%) males[9,10,15,16,23,25,27–29,31–36] and 43 (58.1%) females[2,3,5,8,11–14,16–21,24,26,27,36–46] in the review and male-to-female ratio of 1:1.4 with female predilection. Gender predilection in our review is in accordance with Wang et al.[30] and male-to-female ratio is in contrast with that of Fanburg-Smith et al.[16] According to Kim et al.,[36] most of the patients were from the second decade with slender female predilection.

Location and clinical presentation. According to Wang et al.,[30] base of the tongue was the most common site followed by cheek. Fanburg-Smith et al.[16] also observed lingual ASPS in 14 patients. Our analysis also provides evidence that tongue is the most commonly affected site followed by cheek. Of the 60 cases, 42 (70%) cases were present in the tongue. Out of which, 16 (26.66%) cases did not mention the specific part of the tongue, 11 (18.33%) were present on the base of the tongue, 8 (13.33%) on the dorsum of the tongue, 4 (6.66%) on the ventral surface of the tongue, 2 (3.33%) were on the left lateral dorsum of the tongue, while in 1 (1.66%) case, only lateral surface has been mentioned. The growth rate is mentioned in 12 cases. Out of which, six (50%) exhibited slow growth, two (16.66%) rapid, two (16.66%) gradual, and two (16.66%) were progressive in nature.

Most common clinical presentation was a well-circumscribed mass (2/5; 40%). Most of the lesions resembled vascular lesions appearing bluish (3/7; 42.85%) to dark red in color. All the cases described were non-tender (8/8; 100%). Consistency of the lesion was described as firm (4/12; 33.33%) or soft (3/12; 25%) or between elastic and friable (1/12; 8.33%) or just palpable (2/12; 16.66%). Out of four, two (50%) cases were fixed, while one (25%) was unattached and the remaining one was fluctuant (25%). Maximum cases demonstrated smooth and lobulated surfaces (3/5; 60%), 2 were erythematous (40%), 2 (40%) were ulcerated, while 1 (20%) had surrounding induration. The size of the lesion varied from 1.2 cm × 1 cm to 6 cm × 6 cm. Mean size of ASPS was found to be 3.84 ± 1.945.

Radiographic examination. Among all the cases reported, 15 cases have described radiographic picture of the lesion. One (6.66%) case revealed a large tongue. Four cases (26.66%) revealed well-circumscribed lesion, while one (6.66%) showed a lytic lesion. Four (26.66%) cases presented a high signal intensity on T1-weighted and

Table 1. Summary of clinico-pathological data on alveolar soft part sarcoma.

Parameters	Values	%
Papers	42	
Total cases	74	
Age (years)		
Mean	18.25 ± 15.243	
Range	1.5 to 64	
Gender		
Male	31	41.9
Female	43	58.1
Growth		
Slow	6	50
Rapid	2	16.66
Gradual	2	16.66
Progressive	2	16.66
Duration		
Mean	12.725 ± 23.665	
Range	1 month to 9 years	
Size		
Mean	3.87 ± 1.978	
Range	1.2 × 1 cm^2 to 6 × 6 cm^2	
Site		
Tongue	42	70
Only tongue	16	26.66
Lateral tongue	1	1.66
Left lateral dorsum of tongue	2	3.33
Base of tongue	11	18.33
Dorsum of tongue	8	13.33
Ventral tongue	4	6.66
Cheek	9	15
Mandible	4	6.66
Gingiva	2	3.33
Mouth	1	1.66
Buccal mucosa	1	1.66
Buccal space	1	1.66
Tenderness		
Total	8 cases	
Tender	0	0
Non-tender	8	100
Consistency		
Total	12	
Firm	4	33.33
Hard	2	16.66
Soft	3	25
Palpable	2	16.66
Between elastic and friable	1	8.33
Radiographic features		
Total	15	100
Well circumscribed	4	26.66
Lytic lesion	1	6.66
High signal intensity (T1 and T2)	4	26.66
Hyperintense mass	2	13.33
Enhancing lesions	6	40
Follow up (years)		
Total	46 cases	100
0 –1	12	26.08
1–2	11	23.91
2–3	10	21.73
3–4	4	8.69
4–5	5	10.86
5–6	1	2.17
6–7	1	2.17
7–8	1	2.17
21–22	1	2.17
Metastasis		
Total	8 cases	100
Lung	7	87.5
Lung and liver	1	12.5
Lung and skeletal sites	1	12.5
Bi-lateral lymph nodes	1	12.5
Treatment		
Total	49 cases	100
Surgery	35	71.42
Surgery + chemotherapy	7	14.28
Surgery + radiation	1	2.04
Surgery + chemotherapy + radiation	2	4.08
Chemotherapy	3	6.12
Extirpation	1	2.04

T2-weighted magnetic resonance image. One case (6.66%) displayed enlargement of cervical and left submandibular lymph node. Two cases (13.33%) unveiled a hyper-intense single mass, while six (40%) demonstrated enhancing lesions.

Histopathology. The histogenesis of ASPS has been thought to have a neural, neuroendocrine, or myogenic origin. Histologically, the tumor presents a proliferation of large polygonal/polyhedral cells with ample eosinophilic and granular cytoplasm [periodic acid–Schiff (PAS)-positive diastase-resistant intra-cytoplasmatic material], divided by delicate vascular channels and bands of fine connective tissue that confer an organoid pseudo-alveolar pattern to the tumor. This aspect has been ascribed to the necrosis of the tumor cells. Tumor cells are large polyhedral cells with a large vesicular nucleus and 1 or 2 prominent nucleoli and demonstrate low mitotic figure (4 per 10 high power fields).[25,47] PAS-positive diastase-resistant crystals in the cytoplasm have been found in 80% of the cases. In the present review, out of all the cases, only three cases were PAS positive.[3,29,39] Throughout the tumor, the fine connective

Table 2. Immunohistochemical findings in alveolar soft part sarcoma.

Year	Author(s)	IHC
1990	Takita et al.[9]	Actin (+), desmin (+), vimentin (+)
1993	Carson et al.[10]	NSE (+)
1998	Hunter et al.[11]	Vimentin (+), desmin (+) myoglobin (−), CK (−), EMA (−), neural filament (−), glial fibrillary acidic protein (−), serotonins (−), synaptophysin (−), met-enkephalin (−), leu-enkephalin (−)
1999	Bentley et al.[12]	Vimentin (+), S100 (+), desmin (+), actin (−)
2000	Kimi et al.[13]	Myoglobin (+), sarcomeric actin (+), NSE (+), CK (−), vimentin (−), desmin (−), SMA (−), S100 protein (−), neurofilaments (−), chromogranin A (−), factor VII antibodies (−)
2000	Yoshida et al.[14]	MYOD1 (+), desmin (+), myoglobin (+), alpha SMA (+), vimentin (+), NSE (+), factor VII (−), keratin (−)
2001	Charrier et al.[2]	Desmin (+), vimentin (+), S100 (−), CK (−), HMB45 (−), KLI (−)
2003	Aiken and Stone[3]	HMW (−), LMW (−), S100 (−), PAS (+)
2003	Richards et al.[15]	Vimentin (+), NSE (+)
2004	Fanburg-Smith et al.[16]	Desmin (+), 14 cases SMA (+), vimentin (−), neural/melanocytic (−), MYOD1 (−), histiocytic (−), epithelial markers (−)
2005	do Nascimento Souza et al.[17]	NSE (+), vimentin (+), desmin (+), S100 protein (+), CK AE1/AE3 (+), EMA (+), neurofilament (+), synaptophysin (+)
2006	Ryu et al.[18]	CD64 (+), S100 (−), myoglobin (−), desmin (−), NSE (−), chromogranin (−), synaptophysin (−)
2007	Raghunandhan et al.[19]	Desmin (+)
2009	Rodríguez-Velasco et al.[5]	Vimentin (+), S100 (−), EMA (−), glial fibrillary acidic protein CD68 (−)
2009	Baglam et al.[20]	NSE (+), S100 (+), SMA (+), chromogranin (−), synaptophysin (−), HMB45 (−), vimentin (−), desmin (−), pan-CK (−)
2009	Wakely et al.[21]	Vimentin (+), pan-CK (−), S100 protein (−), HMB45 (−)
2010	Min et al.[22]	MYOD1 (+), desmin (+), TFE3 (+); HMB45 (−), vimentin (−), CD34 (−)
2010	Eley et al.[23]	MYOD1 (+), muscle actin (+)
2011	Conde et al.[24]	VEGF (+)
2012	Argyris et al.[25]	1. NSE (+), CK AE1-AE3 (−), desmin (−), S100 protein (−) 2. TFE3 transcription facto (+), diffuse cytoplasmic NSE (+), MYOD1 (+), CD68 (+), desmin (+), CK AE1/AE3 (−), vimentin (−), SMA (−), smooth muscle myosin heavy chain (−), melanocytic markers S100 (−), HMB45 (−), melan A (−), neuroendocrine markers synaptophysin and chromogranin (−)
2013	Martínez et al.[26]	Myoglobin (+), actin (+), desmin (+), CK (−), S100(−), chromogranin (−)
2014	Wang et al.[27]	PAS (+), vimentin (+), S100 (+), CK (+), HMB45 (−), SMA (+)
2014	Meng et al.[28]	MYOD1 (+), desmin (+), vimentin (+), KPI (−), S100 (−), MSA (−), CD34 (−), chromogranin A (−), synaptophysin (−)
2014	Kinger et al.[29]	MYOD1 (+), S100 (−)

tissue septa around the alveolar structures containing tiny vascular channels with various sizes were evident. There was a loss of intercellular cohesion, with neoplastic cells clinging to the fibrovascular septae, marked in a "pseudo"-alveolar pattern. It has been observed that ASPS tumors in very young patients (<3 years) exhibit a solid morphology, while lesions in older patients (>5 years) develop an organoid pattern. Fanburg-Smith et al.[16] have proposed that the neoplasm's architecture may be an age-related feature.

The pathognomonic histologic feature of ASPS, first designated by Masson,[48] is the existence of granules and rod-like or rhomboid-shaped crystalline inclusions within the cytoplasm of the tumor cells. These crystalline inclusions are found in 25%–100% of cases,[16,48] both in primary and metastatic ASPS. PAS-positive diastase-resistant granules may represent precursors to the rod-shaped crystals. The histogenesis of ASPS is still ambiguous. Historic descriptions of this neoplasm as malignant granular cell myoblastoma, malignant granular cell tumor, and malignant non-chromaffin paraganglioma imitate some of the proposed histogenetic sources from neural or Schwann cell, paraganglia, and skeletal muscle. In an effort to demonstrate

a myogenic origin of ASPS, the characteristic crystalloids encountered in ASPS have been decoded as being constituted of Z-band muscle constituents.[49] Ultrastructurally, some tumor cells show abundant crystalloids with periodicity and membrane-bound intracytoplasmic granules.[14]

Immunohistochemistry. Due to the varied immunohistochemical findings, the exact identity of the tissue type from which ASPS derives still remains unsolved. Many investigators have tried various immunohistochemical markers to reach to the accurate diagnosis of ASPS. Of all the 16 cases, 7 (43.75%) cases showed vimentin positivity (+),[2,5,9,11,12,14,15,17,21,27,28] while 9 (56.25%)[13,16,20,22,25] cases showed negative expression for vimentin. Three cases (18.75%) showed S100 positivity (+),[12,17,20,27] while 13 cases (81.25%) showed negative S100 expression.[2,3,5,13,18,21,25,26,28,29] Out of the nine cases, one case (11.11%) showed positivity for cytokeratin (CK) (+),[27] while eight (88.88%) cases showed negative expression.[20,21,25,26] Argyris et al.[25] had carried out immunohistochemical study on ASPS using TFE3 transcription factor and found positive expression (+) in accordance with the results found by Min et al.[22] He also found positivity with diffuse cytoplasmic neuron-specific enolase (NSE), MYOD1, desmin, and CD68 similar to Min's findings, while Rodríguez-Velasco et al.[5] found negativity for CD68. Argyris et al.[25] found negative expression using CK AE1/AE3, vimentin, smooth muscle actin (SMA), smooth muscle myosin heavy chain, S100, HMB45, Melan A, synaptophysin, and chromogranin. Kimi et al.[13] found positivity with sarcomeric actin, NSE, and negative expression for CK, vimentin, desmin, SMA, S100, neurofilaments, chromagranin, and factor VIII. All of the six cases in the review were found to be HMB45 negative.[2,20–22,25,27] SMA was found to be positive in 18 cases (85.71%)[9,14,20,23,26,27] and negative in 3 cases (14.28%).[12,13,25] Desmin (d-33) expression was positive in 18 cases (85.71%),[2,9,11,12,14,16,17,19,22,26,28] while negative in 3 (14.28%).[13,18,20] From the seven cases, five (71.42%) cases showed positivity (+) for MYOD1,[14,22,23,25,28,29] one (14.28%) case demonstrated strong positivity (++), while the remaining one (14.28%) showed negative expression.[16]

Studies have shown negative expression for CD34 in two cases.[22,28] In the present review, most of the cases showed negative expression for neuroendocrine markers (synaptophysin and chromogranin),[11,13,18,25,26,28] while one (10%) case exhibited positive expression.[17] Myoglobin presented positivity in three (60%) ((2 (+); 1 (++)),[13,14,26] while negative expression in two (40%) cases.[11,18] NSE expression was positive in six (66.66%) cases,[10,13,14,15,17,20,25] while negative in three (33.33%) cases.[18]

Metastasis. Despite very indolent growth, ASPS has a high proclivity for metastasis. It most commonly spreads to the lungs in 42%–65% of cases, and less common sites are the bones and the brain.[7] In the present review, we have found seven (87.5%) cases metastasizing to the lungs, one (12.5%) case each to the lung and liver, lung and multiple skeletal sites, and to the bilateral lymph nodes. There is also a reported case of metastasis to the oral cavity in the literature.[47]

Genetics. ASPS displays t(X;17)(p11;q25) linking the TFE3 gene. This is used to validate the diagnosis of ASPS.[50] The antibody against TFE3 displays inconsistently strong nuclear positivity in most of the ASPS cases. The der(17) t(X;17)(p11;q25)[51] hints to rearrangement and fusion of ASPL and TFE3 genes. Two modifications of ASPSCR1/TFE3 gene fusion have been reported. Both encrypt a chimeric protein consisting of the ASPL N-terminal region fused to TFE3 basic helix-loop-helix and leucine zipper DNA-binding domains. The expression of ASPSCR1/TFE3 chimeric protein is deliberated to cause transcriptional deregulation and has been linked as a possible tumorigenic mechanism. Detection of t(X;17) or ASPL-TFE3 fusion records are highly specific and sensitive markers for interpreting the diagnosis of ASPS[29,27] outstanding the accuracy of immunohistochemical stain for TFE3.[52] Argyris et al.[25] found TFE3 + VE signifying translocation of TFE3 gene and presence of an ASPSCR1/TFE3 fusion transcript, which has been authenticated by other authors as well.[21,22,24,30]

Differential diagnoses. Differential diagnoses of ASPS comprise tumors with large cells organized in nests with eosinophilic/clear cytoplasm, such as malignant melanoma, renal clear cell carcinoma, adrenal cortical carcinomas, and hepatocellular carcinoma. Paraganglioma is also incorporated as it shows organoid and granular pattern. Cell schwannoma also bestows light granular cytoplasm and is incorporated as a part of the differential diagnosis.[17,53,54] All these differential diagnoses can be expelled with adequate PAS and immunohistochemical analysis. ASPS is negative for epithelial, melanocytic, and neuroendocrine markers. Vimentin and NSE can be positive, but are not specific. Muscle markers can be immunoreactive and some researchers consider ASPS as a subtype of rhabdomyosarcoma, but is not widely accepted.[16,53] Immunoreactivity for pan-CKs AE1–AE3, epithelial membrane antigen (EMA), neurofilaments, and synaptophysin antibodies have also been elaborated.[17]

The differential diagnoses for ASPS include paraganglioma, renal clear cell carcinoma, and granular cell schwannoma. The differential diagnosis from metastatic renal cell carcinoma is grounded on the fact that lack of CD10, EMA, RCC, PAX2, and CA9 expression in ASPS. Extrarenal rhabdoid tumor and rhabdoid tumor of the kidney are malignant neoplasms with expression of vimentin, keratins, EMA, CD99, synaptophysin, and NSE. Apart from this, identification of mutations and homozygous

deletions of the SMARCB1 (INI1) gene or absence of nuclear staining for INI1 are commonly encountered in rhabdoid tumors, but absent in ASPS.[55] Sometimes, ASPS may attain clear cytoplasmic features and simulate lipomatous neoplasms such as hibernoma, lipoblastoma, or liposarcoma. These are usually positive for S100 protein and display precise morphologic alterations such as lipoblasts, differentiating them from ASPS.[56] Paraganglioma displays a distinctive nested "zellballen" architecture[57] and reveals positivity for neuroendocrine markers, while S100 protein highpoints the auxiliary sustentacular cells.

TFE3 is used to identify ASPS but not so specifically, as it may stain the nuclei of granular cell tumor,[52] paraganglioma, translocation-related renal cell carcinomas, and adrenocortical carcinoma.[58] If there is uncertainty about the diagnosis, RT-PCR for the ASPSCR1/TFE3 fusion transcript is a more precise method.[52,57] A comparable but common t(X;17)(p11;q25) translocation and ASPSCR1/TFE3 fusion transcripts have also been acknowledged in a type of renal cell carcinoma developing in pediatric and young adult patients.[59]

Treatment. Although various treatments have been proposed, complete resection is recommended for ASPS to avoid its recurrence. The most common surgical intervention, which was followed in various cases in this review was resection (35/49; 71.42%) followed by combination of surgery and chemotherapy (7/49; 14.28%). A few cases were treated with a combination of surgery, chemotherapy, and radiation (2/49; 4.08%). A few cases were also treated with a combination of surgery and radiation (1/49; 2.04%). A case was also treated with only chemotherapy (3/49; 6.12%) and the remaining one was extirpated (1/49; 2.04%).

Limitations

As per the inclusion criteria, the present review encompasses full-text articles in English language and excludes abstracts whose full text could not be retrieved. Moreover, case reports in foreign languages except English were excluded which may have caused loss of relevant data. Undiagnosed or misdiagnosed cases of ASPS were not included in the present review.

Future implications

A comprehensive search and inclusion of published and unpublished data/case reports in other languages should be incorporated to improve the review on this rare entity.

Conclusion

In conclusion, ASPS showed distinct female predilection with predominance in the first decade of life. It showed strong predilection for soft tissues of head and neck, especially the tongue followed by cheek with painless well-circumscribed swelling as the most common presentation. Histologically, the tumor showed a proliferation of large polygonal/polyhedral cells with abundant eosinophilic and granular cytoplasm (PAS-positive diastase-resistant intra-cytoplasmatic material), separated by delicate vascular channels and bands of fine connective tissue that confer an organoid pseudo-alveolar pattern to the tumor. Due to unavailability of follow-up data in many cases, it would be inappropriate to comment on the best treatment modality of this neoplasm.

Conflict of interest

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