

CASE REPORT

Primary alveolar soft part sarcoma of the chest wall: A case report

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Abstract

A 26-year-old man presented to the emergency department due to headache, nausea, and vomiting. He had a right subclavicular slow-growing mass. Histopathological evaluation showed alveolar soft part sarcoma. The patient was found to have multiple cerebral and pulmonary metastases. So far, he has got three cycles of brain radiotherapy.

KEYWORDS

alveolar soft part sarcoma, case report, neoplasm metastasis

1 | INTRODUCTION

Soft-tissue sarcomas are a diverse category of malignancies that have at least 70 distinct histologic subtypes.^{1,2} Alveolar soft part sarcoma (ASPS) is an extremely rare subtype of sarcoma that accounts for <1% of all soft-tissue sarcomas,³ which was initially characterized by Christopherson et al. in 1952.⁴

Alveolar soft part sarcoma is more frequent in young adults and adolescents, particularly those between the ages of 15 and 35. Before the age of 30, there is a female-to-male predominance, with the ratio reversing as individuals get older.³ ASPS is most commonly manifested as a painless, slow-growing lesion that induces relatively minor functional impairment.⁵ Primary tumors are

frequently huge, vascularized masses that appear as pulsatile masses. In children, the head and neck, particularly the orbit and tongue, are typical sites of involvement, but in older individuals, it affects the muscles of the upper and lower extremities. Despite of its slow growth pattern, overall prognosis is poor due to fast metastatic dissemination to the bone, lungs, lymph nodes, and brain.³

The majority of prior studies have concluded that the mainstay of treatment is total surgical excision of the tumor mass in its early stages. Other treatments, including as radiation therapy, chemotherapy, and biologic therapy, are less well recognized, and the evidence is still unclear.⁶ Given the disease's rarity, comprehensive and detailed knowledge of its clinical course, pathophysiology, and effective therapy remains challenging. In this case

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report, we describe the accurate diagnosis and treatment of primary ASPS of the chest wall in a young man who presented with headache, nausea, and vomiting.

2 | CASE PRESENTATION

A 26-year-old man presented to the Emergency Department of Ghaem Hospital (Mashhad, Iran) with complaints of headache, nausea, and vomiting for 1 week. The patient had a 2-year history of painless swelling in the right subclavicular region that had gradually increased in size. Due to his low economic state, he did not seek medical attention throughout this time period. The mass progressively restricted his right arm's movements, although it was not painful. Moreover, he has been suffering from progressive anorexia and a loss of weight (around 5 Kg in the previous 5 months). His family history, social history, and his review of systems were unremarkable.

In the physical examination, a tender, warm and soft mass measuring 10 × 11 cm was detected over the anterior portion of the upper right hemithorax. Pulmonary and cardiac auscultation were normal. The extension movement of the right arm was restricted in both active and passive activities. There was no evidence of papilledema, optic atrophy, or focal neurologic deficit. Initial laboratory assessment of the patient illustrated in Table 1.

An ultrasonographic assessment of the right upper hemithorax revealed a heterogeneous, hypoechoic, highly vascular soft-tissue mass. Also, abdominal ultrasonography was unremarkable. A computerized tomography (CT)

scan of the chest revealed a heterogeneous enhancing mass with internal necrosis and prominent surrounding veins, behind the right pectoralis major muscle with no bone involvement (Figure 1). A brain CT scan and magnetic resonance imaging (MRI) were conducted due to the patient's headache to assess suspected cerebral involvement, and several brain metastases were detected (Figure 2). Imaging and clinical findings were used to accurately diagnose of a soft-tissue malignant neoplasm with cerebral and pulmonary metastases. As illustrated in Figure 3, histologic findings revealed neoplastic proliferation of epithelioid cells with eosinophilic cytoplasm and nuclear atypia with alveolar pattern.

According to the immunohistochemistry (IHC) analysis, the tumoral cells were reactive for vimentin and myoD1 but were negative for ck, pax8, ema, CD10, CD5, CD3, LCA, Arginase, RCC marker, Heppar A, Melan A, S100, Chromogranin and Synaptophysin. The IHC and histologic findings pointed to ASPS as the most likely diagnosis. The patient has undergone three cycles of brain radiotherapy and has refused all other treatments.

3 | DISCUSSION

Alveolar soft part sarcoma is a very rare vascular soft-tissue malignancy that has generally poor prognosis.⁷ Children and adolescents are most commonly affected by ASPS. Nevertheless, the typical range of age for presentation is 15–35 years, and our patient matches into this range. The majority of investigations have reported a female preponderance in adult patients, while no such preference has been observed in adolescents.⁶ Lieberman and colleagues studied the histopathological and clinical characteristics of 102 ASPS patients and found that the median age of patients at the time of diagnosis was 22 years, with a 1.5 to 1 female preponderance.⁸

Although the precise etiology of ASPS is unknown, investigations suggest that genetic alterations probably play a significant role.⁹ ASPS develops in the presence of skeletal muscles or musculofascial planes, which reveals the tumor's high affinity for the buttocks, thighs and chest or abdominal walls.¹⁰ ASPS of the chest wall is scarce, and we only have limited information about therapeutic options and outcomes. The majority of investigations have classified these malignancies as truncal sarcomas, with a poor prognosis. In a study by Gordon et al., 149 patients with chest wall sarcoma were evaluated and only 3% of them had ASPS.¹¹

Alveolar soft part sarcoma typically manifests as a painless, slow-growing tumor with a high risk of metastasis. As this tumor grows slowly and insidiously, patients typically present with a long clinical history and a

TABLE 1 Laboratory analysis of the patient

Laboratory finding (unit)	Result	Reference value
WBC (1000/mcL)	7.8	3.9–12
Polymorphonuclear (%)	84	-
Lymphocyte (%)	13.3	-
RBC (10 ⁶ /mcL)	5.48	4.5–6.2
Hb (g/dl)	13	11–16
Platelets (1000/mcL)	400	150–450
Blood urea (mg/dl)	28	15–45
Creatinine (mg/dl)	0.8	0.6–1.3
Sodium (mEq/L)	137	135–145
Potassium (mEq/L)	3.7	3.5–5.5
AST (IU/L)	44	Up to 40
ALT (IU/L)	58	Up to 40
Alkaline phosphate (IU/L)	219	80–306

Abbreviations: ALT, alanine aminotransferase; AST, aspartate aminotransferase; Hb, hemoglobin; RBC, red blood cell count; WBC, white blood cell count.

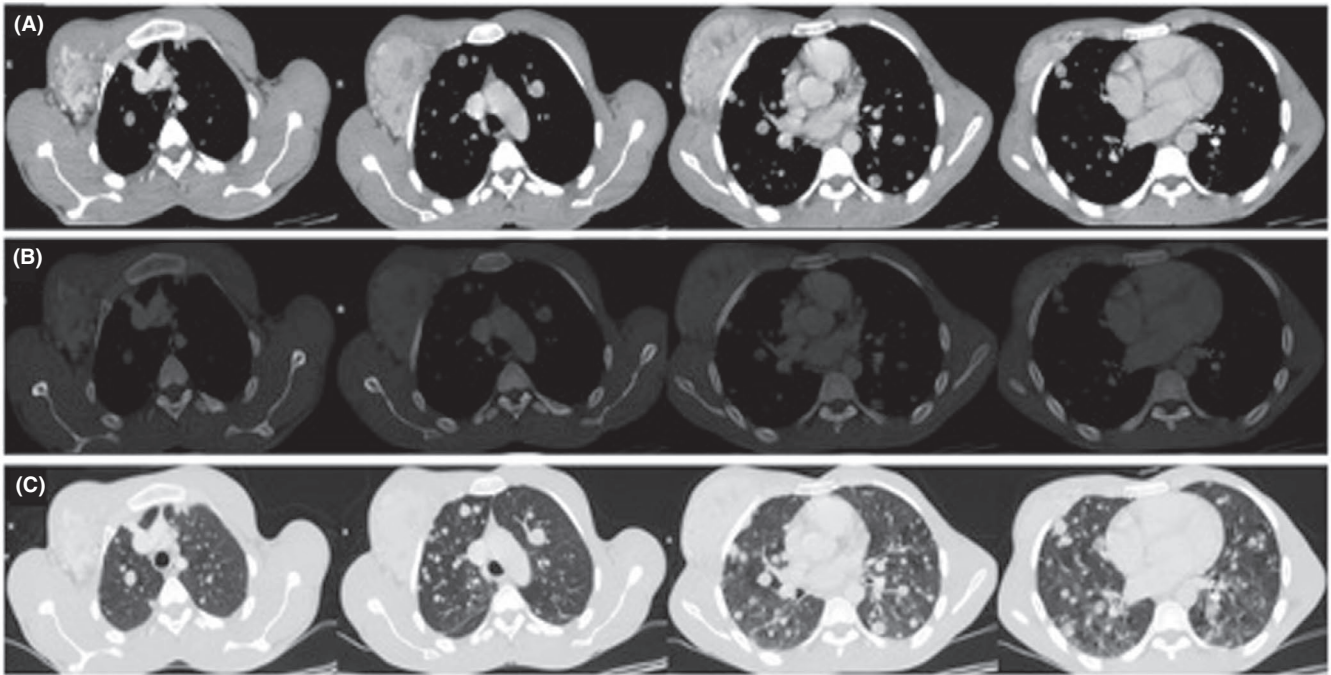
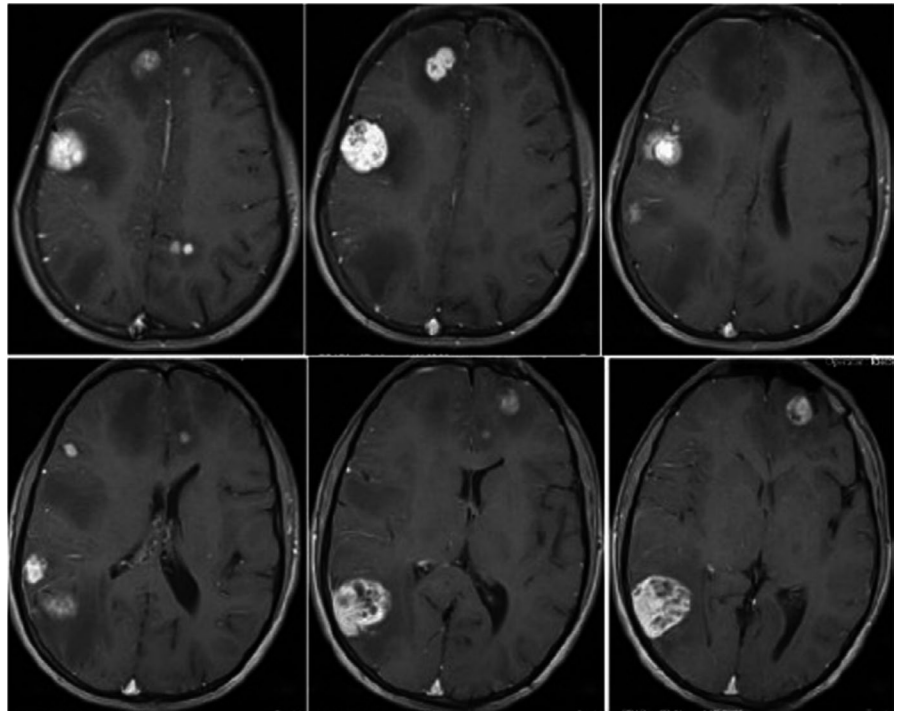


FIGURE 1 Computed tomography scan (A) mediastinal, (B) bone, and (C) lung window (venous phase after contrast): (A) Heterogeneous enhancing mass with internal necrosis and prominent surrounding veins, behind the right pectoralis major muscle was detected. (B) There was no evidence of bone involvement. (C) In the fields of both lungs, there are several metastases with a random distribution

FIGURE 2 T1-weighted post-contrast images show multiple metastases with strong enhancement and mass effect, as well as severe vasogenic edema around them



massive mass, like in our case, which had a significant swelling in the right hemithorax. Lung metastases are the most common, followed by bone and brain metastases.¹² As in our patient, these metastases are found in approximately 20%–25% of patients at the time of diagnosis.⁶

Except for the bone metastases, our patients had several lung and cerebral metastases. Our patient did not consult a doctor despite the evident swelling on his chest wall, and he presented with symptoms of cerebral and pulmonary metastases, a characteristic that is more common in individuals with tumors in the extremities rather than the chest wall.

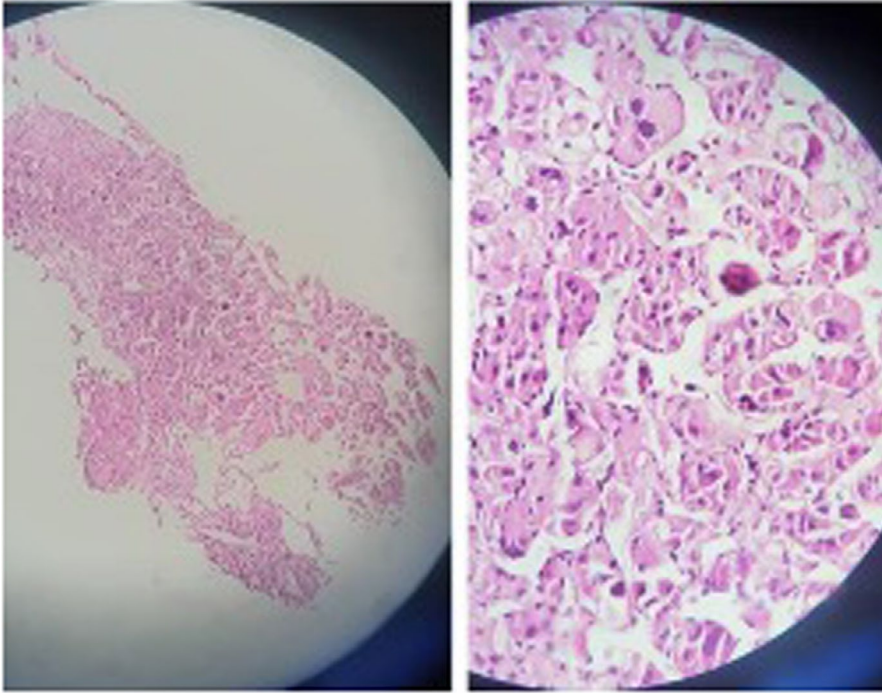


FIGURE 3 Light microscopic view shows neoplastic proliferation of epithelioid cells with eosinophilic cytoplasm and nuclear atypia with alveolar pattern (H&E stain, $\times 40$, $\times 100$)

Clinical suspicion and clinicopathological correlations with proper radiologic investigations are required for accurate diagnosis and treatment of this rare tumor. Radiologic findings include prominent venous vascularity on CT scan and high-signal intensity on T1- to T2-weighted images on MRI. Cui et al. investigated MRI characteristics in 12 patients with ASPS and found that low signals of radiating flow voids with high signals of slow blood flow or blood sinuses in the central region of the tumors have high diagnostic importance. Moreover, MRI and CT scan can be used to determine tumor-free resection margins.¹³ Nevertheless, final diagnosis and the best definitive diagnostic method is biopsy, which is obtained by a surgical procedure.¹⁴

Surgical excision and/or systemic therapy for metastatic disease are commonly used in the treatment of ASPS. If removing the whole tumor is not feasible or the tumor has disseminated, surgery and radiotherapy may be used simultaneously.¹⁵ Chemotherapy still hasn't been proven to be useful in brain metastases, while being beneficial for the primary lesion. This is due to the medications' inability to pass the blood–brain barrier.¹⁶ Also, the effective treatment of ASPS necessitates supportive medical care and long-term follow-up.¹⁷

Lieberman et al. found that ASPS is a very indolent malignancy with a long clinical duration and late metastases, with survival rates of 77%, 60%, 38%, and 15% at 2 years, 5 years, 10 years, and 20 years, respectively.⁸ Pennacchioli et al. followed 33 patients for a median of 72 months and found that they had a 5-year survival rate of 68.7% and a 10-year survival rate of 53.4%. They concluded that the

prognosis of ASPS is mostly influenced by the disease's characteristics as well as surgical quality, with the best outcomes seen in smaller, well-resected tumors.¹⁸

4 | CONCLUSION

Alveolar soft part sarcoma of the chest wall is an uncommon and aggressive neoplasm that has a high chance of spreading to other parts of the body. The major symptom of patients is typically a painless mass. This neoplasm must be extensively evaluated clinically, radiographically, and histopathologically in order to reach an accurate diagnosis.

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CONFLICTS OF INTEREST

The authors declare that there are no conflicts of interest.

AUTHOR CONTRIBUTION

All authors contributed to the study's conception and design. OS, FK, AG, AHJ, and BA performed material preparation, data collection, and acquisition. AG, FK, HSB, and OS involved in writing the first draft of the manuscript. All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

CONSENT

This study was approved by the Ethics Committee of Mashhad University of Medical Sciences, Mashhad, Iran. All tests were carried out in compliance with the institution's specified rules and regulations. Furthermore, the patient's written informed consent was acquired for the publishing of this case report.

DATA AVAILABILITY STATEMENT

The data are also available on request.

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