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Original Article

Alveolar soft part sarcoma: Clinical presentation, treatment, and outcome in a series of 13 patients

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Abstract

Background: Alveolar soft part sarcoma (ASPS) is a rare soft tissue tumor that typically affects young patients. Similar to other soft tissue sarcomas, it has high pulmonary metastasis ability, whereas compared with other soft tissue sarcomas, it has high brain metastasis ability. Because of the rarity of the disease, most studies on ASPS have been case reports and small series studies.
Method: We performed a retrospective study to evaluate the clinical and pathological features and oncological results in a consecutive series of patients with localized or metastatic ASPS treated at our institute between 1994 and 2014. Demographics, location, severity of disease, treatment provided, progression-free survival, and overall survival were evaluated.
Results: A total of 13 patients were investigated. The most common locations of primary tumor were the thigh ($n = 6$, 47%), followed by the flank ($n = 3$, 23%), forearm ($n = 2$, 15%), and calf ($n = 2$, 15%). Three patients were initially diagnosed as having hemangiomas elsewhere. These patients received unplanned intralesional excision. All the patients received wide tumor resection at our institute. Over the average follow-up period of 80.5 months (range: 36−133 months), the 5-year overall survival rate was 67.5%. Four patients were continuously disease free (31%), six were living with disease (46%), and three died of disease (23%). Of nine patients who presented with distant pulmonary metastasis, two had bony and brain metastases. The 5-year survival rate was 66.7% in patients who received chemotherapy and those who did not ($p = 0.941$).
Conclusion: The treatment strategy for ASPS is wide resection, and postoperative chemotherapy may be crucial for long-term survival. In addition, this type of tumor has a high distant metastasis rate at the time of diagnosis, particularly in the lungs and brain.
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Keywords: Alveolar soft part sarcoma; Chemotherapy; Metastasis; Surgery

1. Introduction

Alveolar soft part sarcoma (ASPS) is a rare, clinical, and distinctive soft tissue sarcoma that typically affects young patients and was initially described by Christopherson in 1952.[1] It is generally believed to account for 0.5%−1% of all soft tissue tumors.[2,3] ASPS typically occurs in adolescents and young adults aged 15−35 years, particularly in females, who

Conflicts of interest: The authors declare that they have no conflicts of interest related to the subject matter or materials discussed in this article.

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exhibit an incidence rate of 60%.[4,5] The location of the tumor is typically in the deep soft tissue, primarily in the thighs or buttocks; however, it is also found in the arms, breasts, genital area, chest, and retroperitoneal tissue.[6] Radiographically, ASPS is characterized by calcification through plain radiography. In addition, magnetic resonance imaging (MRI) has revealed high signals in both T1- and T2-weighted images and internal and external multilobulated signal changes (Fig. 1).[7,8] Histopathologically, periodic acid-Schiff-positive findings may reveal rhomboid-shaped crystalline material, and ASPS cells also have round, regularly placed nuclei with vesicular chromatin and a prominent central nucleolus (Fig. 2).[9] Genetically, ASPS is caused by an unbalanced translocation, namely der(17)t(X:17)(p11;p25), which results in the formation of an ASPL–TFE3 fusion gene.[10] Because of the rarity of this disease, most studies on ASPS have been case reports and small series studies. In the present study, we first analyzed the patients' demographic and tumor characteristics in Taiwan. Second, we evaluated the factors related to clinical outcomes of ASPS.

2. Methods

The medical records of 23 patients pathologically diagnosed as having ASPS who received surgical treatment between June 1994 and July 2014 were analyzed in our hospital. Those treated at other departments ($n = 10$) were excluded. Two patients were diagnosed as having ASPS in other areas such as the nasopharyngeal area ($n = 1$) and retroperitoneal tissue ($n = 1$). Eight were receiving chemotherapy without surgical intervention. No patients received the treatment protocol in our department. A total of 13 patients were enrolled in this study. The mean age of the patients was 21 years (range: 9–52 years). The mean follow-up period was 80.5 months (range: 36–133 months). We performed a retrospective study to evaluate the patients' demographic and tumor characteristics. In addition, the clinical outcomes were analyzed and compared based on several factors.

The anatomic location and depth of each tumor were evaluated and recorded by the first medical doctor. A tumor was classified as deep if it invaded the deep fascia in the

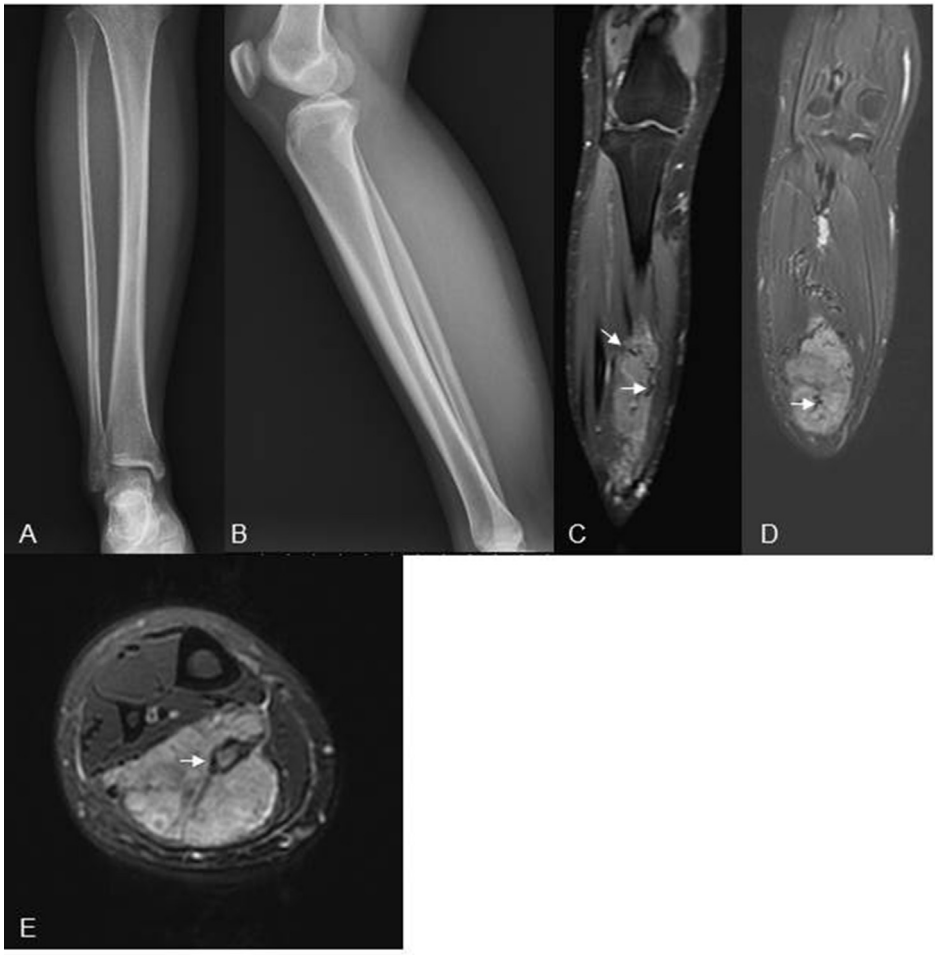

Fig. 1. Image showing a right calf mass present for 16 months in a 19-year-old women. (A) Radiography showing the anterior-posterior view of the right lower leg. No obvious calcification or bone erosion can be observed. (B) Radiography showing the lateral view of the right lower leg. Soft tissue mass over the calf area can be observed. (C) Coronal post-contrast T1-weighted image showing inhomogeneous enhancement of the tumor in the right lower calf with ill-defined margin and tortuous vessel (arrow). (D) Coronal fat-suppressed T2-weighted image revealing the irregular signal tumor with intra-tumoral signal voids (arrow). (E) Axial fat-suppressed T2-weighted image revealing the tortuous vessel with flow voids (arrow).

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