

Ultrasonographic and MR Findings of Alveolar Soft Part Sarcoma

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Alveolar soft part sarcoma (ASPS) is a rare soft-tissue sarcoma, commonly occurring in children and adolescents. The tumor mostly involves the lower extremities. The prognosis of the patient depends on whether there is metastasis. We present a 19-year-old female with ASPS in her right lower leg. Grayscale and color Doppler ultrasound showed a well-defined hypoechoic lesion with hypervascularity and very low resistive index (RI). Magnetic resonance imaging revealed iso signal intensity to muscle on T1-weighted images, high signal intensity to muscle on T2-weighted images with signal voids, and good enhancement after gadolinium administration. In a mass with hypervascularity and very low RI on sonography and hypervascularity with flow voids on magnetic resonance imaging, ASPS should be considered. [*J Chin Med Assoc* 2009;72(6):336–339]

Key Words: alveolar soft part sarcoma, color Doppler ultrasound, MRI

Introduction

Alveolar soft part sarcoma (ASPS) constitutes less than 1% of soft tissue sarcomas, with young peak age and slight female predominance.^{1,2} The lesion is most commonly found in the proximal lower extremities.³ The imaging modalities for this tumor include plain film, sonography, computed tomography (CT), and magnetic resonance imaging (MRI). ASPS commonly shows equal or higher signal intensity to muscle on T1-weighted images and high signal intensity on T2-weighted images, with intra- and extratumoral flow voids. Marked enhancement is seen after gadolinium administration.^{4–9} Sonographic findings of ASPS reveal well-circumscribed hypoechoic lesion with hypervascularity.^{6,10} No resistive index (RI) of color Doppler ultrasound (CDUS) for ASPS has previously been reported. Here, we present the sonographic, CDUS and MRI findings of ASPS in the right lower limb of a 19-year-old female.

Case Report

A 19-year-old female complained of swelling in her right thigh for about 3 years. She went to visit her doctor because of increased size of the lesion and painful sensation. Physical examination revealed a tender mass in the right lower extremity. The patient was then referred to our department for further evaluation. Grayscale and CDUS revealed a hypoechoic mass measuring 6.3 × 4 × 3 cm in size at the lateral aspect of the right lower leg with hypervascularity and very low RI, with suspected arteriovenous shunt formation (Figure 1). MRI revealed a well-enhanced mass between the right peroneous longus and brevis muscles. The lesion showed iso signal intensity to muscle on T1-weighted images and high signal intensity on T2-weighted images. Some flow void serpiginous lines in the peripheral region of the lesion were also noted (Figure 2). Tentative diagnoses included hypervascular sarcomas, such as ASPS and angiosarcoma.



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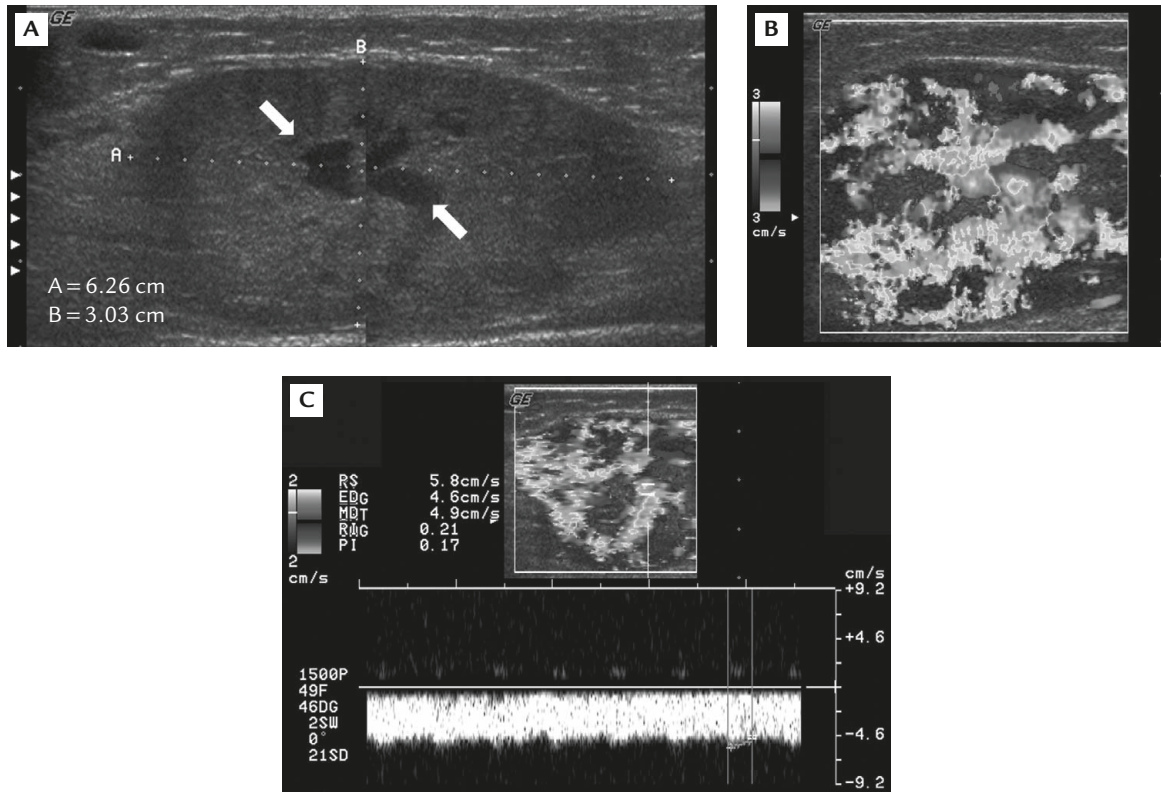


Figure 1. Sonography of the right thigh. (A) Grayscale ultrasound reveals a well-defined mass lesion with tubular structures (arrows). (B) Color Doppler ultrasound shows that these tubular structures are extremely hypervascularized. (C) The lowest resistive index of the lesion was 0.24.

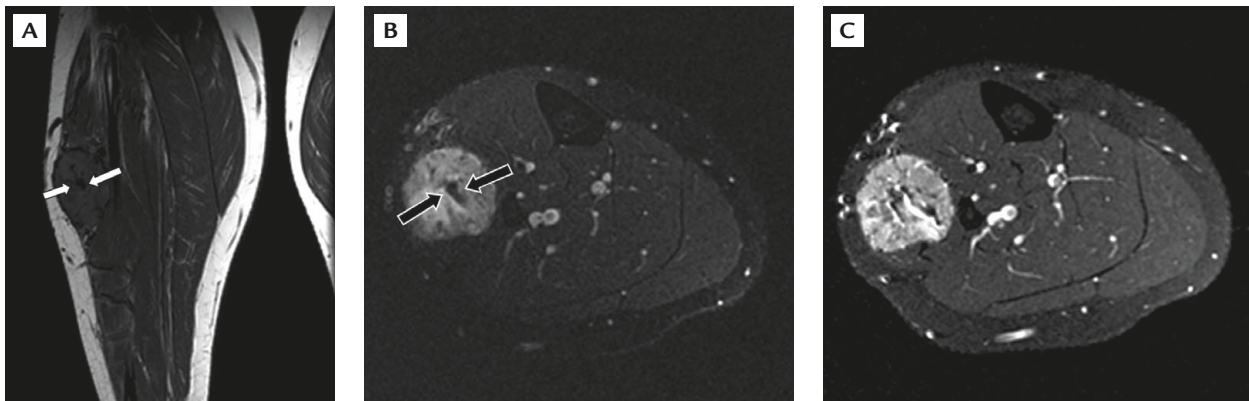


Figure 2. Magnetic resonance imaging of the right thigh. (A) T1-weighted (TR/TE: 550/14) coronal view shows an iso signal intensity mass lesion with signal voids (arrows). (B) T2-weighted (TR/TE: 3000/55) axial view with fat saturation shows a mass lesion with high signal intensity and signal voids (arrows). (C) The mass is strongly enhanced after gadolinium injection.

Wide excision was performed. Histology demonstrated alveolar soft-tissue sarcoma. Microscopy revealed large tumor cells with vesicular nuclei, prominent nucleoli, and granular cytoplasm, clustered in well-defined nests separated by delicate fibrous tissue. Arteries and dilated venous lumens were also seen (Figure 3), with possibly direct shunt of artery and vein.

Discussion

The name *alveolar soft part sarcoma* refers to its typical microscopical morphology of large granular cells organized in an alveolar-like array and separated by a rich vascular network.³ It is a rare soft-tissue sarcoma occurring in the second to fourth decades of life.^{1,2}

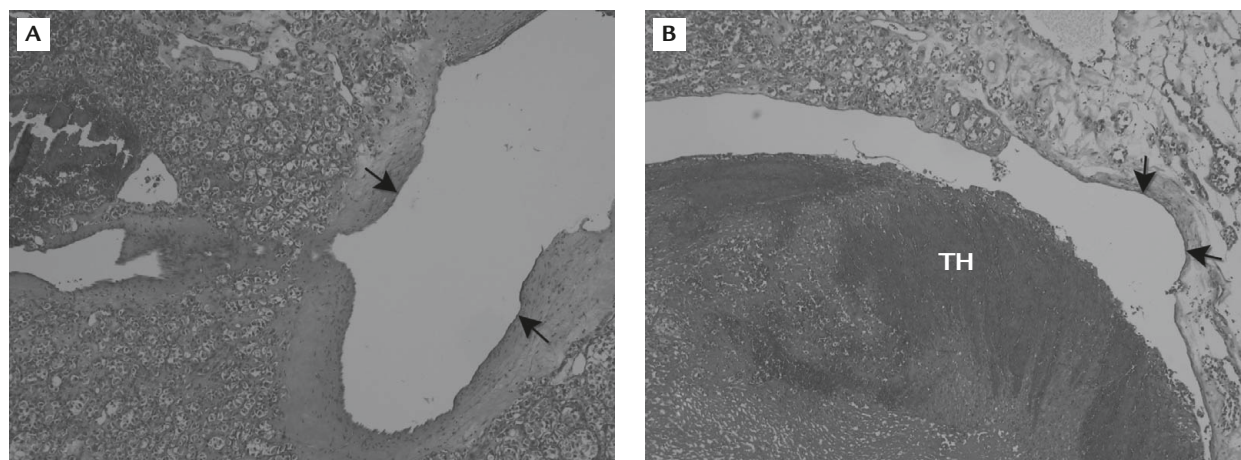


Figure 3. Tumor histology with hematoxylin and eosin stain (40 \times). (A) Photomicrograph shows large tumor cells with vesicular nuclei and granular cytoplasm. The artery (arrows) can be seen. (B) The dilated vein (arrows) with tumor thrombus (TH).

It can present in any region of the body, but is most commonly found in the lower extremities, followed by the trunk and upper extremities. Metastasis has been reported in 20–25% of patients at diagnosis.¹¹ The lungs are the most frequently affected organ.¹² The long-term outcome of patients varies with disease extension. The 5-year survival rate of patients with localized ASPS is 60–88%.^{1,11,13} However, if there is metastasis, only 10% of patients survive >5 years from the time of diagnosis.^{11,13}

Unenhanced T1-weighted MRI of ASPS usually show equal or higher signal intensity to muscle, and T2-weighted MRI shows very high signal intensity. Intra- and extratumoral flow voids are seen on both T1- and T2-weighted images, representing enlarged feeding arteries and draining veins. Marked enhancement is seen after gadolinium administration.^{4–9}

There are a few reports describing the sonographic findings of ASPS, which are as follows: well-circumscribed hypoechoic lesions with hypervascularity and without sound through transmission or acoustic enhancement.^{6,10} In our case, CDUS revealed extreme hypervascularity and very low RI, about 0.24–0.58. Low RI may indicate the presence of arteriovenous shunt.¹⁴ The histology also demonstrated artery and dilated vein with direct shunt between them.

The imaging findings of our patient suggested the differential diagnoses of hemangioma, arteriovenous malformation (AVM), hemangiopericytoma, and angiosarcoma.^{4,15} We can distinguish ASPS from hemangioma by the presence of flow voids and central necrotic area of ASPS, and from AVM by significant soft tissue component and slow washout of contrast medium of ASPS. Hemangioma and AVM show indistinct margins and sometimes phlebolith on sonography. The peak ages for hemangiopericytoma and

angiosarcoma are older, and their RI are higher than that of ASPS.¹⁵ Another differential diagnosis is hypervascular metastasis, such as from thyroid cancer or renal cell carcinoma,¹⁶ in which case the previous malignancy history of the patient should be sought. It is difficult to distinguish hypervascular metastasis with low RI from ASPS without knowing the previous malignancy history of the patient. With regard to other soft-tissue sarcomas, they are not as hypervascular or with such low RI as ASPS.¹⁵

In conclusion, we presented the specific sonographic and MRI findings in a young woman with ASPS in her right thigh, which were iso- or high signal intensity on T1-weighted images, high signal intensity on T2-weighted images, intra- or extratumoral flow voids and hypervascularity on MRI, and well-circumscribed with extreme hypervascularity and very low RI on sonography. In a mass with hypervascularity and very low RI, ASPS should be considered.

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