Abstract. Alveolar soft-part sarcoma (ASPS) is an uncommon soft-tissue neoplasm, which is commonly found in the deep soft tissues of the extremities, with a propensity for recurrence and metastasis. However, the metastasis of ASPS to the heart is exceedingly rare. The present study reports such a rare case with cardiac metastasis. Using computed tomography, a 37-year-old man was diagnosed with brain, lung and spleen metastases from a previously treated ASPS in the left forearm. Cardiac metastasis was then diagnosed 1 month later. Despite chemotherapy and palliative whole-brain radiotherapy, the patient succumbed to the disease shortly after. This case suggests that the widespread metastases and cardiac involvement of ASPS may result in a poor outcome.

Introduction
First described in 1952 by Christopherson et al (1), alveolar soft-part sarcoma (ASPS) is a malignant, highly vascular tumor with distinct morphology compared with other soft-tissue sarcomas, and its cell of origin and biology have remained unclear. Clinically, ASPS constitutes <1% of all soft-tissue sarcomas (2) and occurs principally in individuals between 15 and 35 years of age, with a notable female predominance, particularly among patients <20 years of age (3). The tumor often originates in the muscle and deep soft tissues of the extremities, but it has also been found in tissues lacking skeletal muscle, such as those of the lungs, breasts, stomach, female genital organs and bones (4,5). This tumor type has an indolent clinical course, with a high tendency to metastasize via hematogenous dissemination (6). Metastases to the lungs, bones and brain are common and often asymptomatic, and may be detected after long disease-free intervals, while cardiac metastases from ASPS are rarely reported (7,8). Surgical excision of the primary and pulmonary metastases is the mainstay of treatment, although it remains rarely curative, and radiotherapy has a role in treating visual or microscopic residual disease following resection (9). Anthracycline-based chemotherapy regimens rarely elicit responses, and in general, ASPS is essentially impervious to well-established chemotherapy agents (10). Overall, the prognosis of ASPS is poor. The present study reports a rare case of ASPS with cardiac metastasis.

Case report
On August 10th 2012, a 37-year-old man was admitted to Shandong Provincial Hospital Affiliated to Shandong University (Jinan, Shandong, China) with complaints of headaches, dizziness and nausea over a period of 20 days. It was noted that 3 years previously, the patient had undergone surgery in other institution due to a progressively enlarging painless mass on the left forearm. Histopathological examination of the specimen confirmed ASPS (Fig. 1). During the current admission, computed tomography (CT) scans showed metastatic brain, lung and spleen lesions (Fig. 2), with no cardiac lesion. The patient was administered palliative whole-brain radiotherapy, which was delivered as 30 Gy in 10 fractions for 2 weeks. Chemotherapy consisting of 75 mg/m² cisplatin on days 1-3, 40 mg/m² pirarubicin on day 1, and 2 g/m² ifosfamide on days 1-3, every 3 weeks, was initiated during the palliative whole-brain radiotherapy. Despite the fact that the symptoms associated with cerebral metastasis were alleviated, a repeat CT scan that was performed after two cycles of chemotherapy demonstrated a new mass located in the upper segment and extending to the middle of the interventricular septum (Fig. 3), with contrast enhancement of the rim of the lesion consistent with cardiac metastasis. Transthoracic echocardiography was performed and this lesion was identified as a mass measuring...
40x21 mm within the interventricular septum (Fig. 4), without remarkable left ventricular outflow tract obstruction. The cardiac mass appeared hypoechoic, with blood flow signals detected. Ten days after the completion of two cycles of chemotherapy, the patient had no symptoms associated with the cardiac lesion and refused to continue further treatment.

Figure 1. Histology of the the left forearm specimen of alveolar soft-part sarcoma. (A) Hematoxylin and eosin staining; original magnification, x200. (B) Hematoxylin and eosin staining; original magnification, x400.

Figure 2. Computed tomograph images revealing the multiple metastases of (A) the lung, (B) the spleen and (C and D) the brain.

Figure 3. Computed tomography images revealing a 4.0-cm cardiac mass (arrows) in the (A) upper segment extending to the (B) middle of the interventricular septum.
The patient subsequently succumbed to cachexia and fast progression of the disease. Although a histopathological specimen could not be obtained from the heart, according to the CT scan and transthoracic echocardiography demonstrative tools, we believed the cardiac lesion to be cardiac metastasis of the ASPS from the forearm.

**Discussion**

ASPS is a rare soft-tissue malignancy that was originally alluded to by Smetana and Scott (11) in 1951 as a malignant tumor of the non-chromaffin paraganglia. In 1952, Christopherson *et al* (1) described it under its present name of ‘alveolar soft-part sarcoma’. Accounting for <1% of all soft-tissue sarcomas, ASPS most typically occurs in adolescents and young adults, with a female predominance (12,13). It is known that the most common primary sites of the tumor are the lower extremities, frequently the thigh or trunk (14). In the present case, a 37-year-old man was diagnosed with ASPS with the primary location of the left forearm. It is reported that ASPS is an indolent disease with characteristic slow growth, but that is associated with an poor overall outcome and a 5-year survival rate of only 20% in unresectable metastatic patients (15,16). Metastases to the lungs, brain and bones are common, and usually lead to a low survival rate. However, ASPS metastases to the heart are extremely unusual. In a review of the literature, only 2 papers on cardiac metastasis were found: Akiyama *et al* (17) reported the case of a 29-year-old man with ASPS of the brain and cardiac metastasis, while the case of a 13-year-old girl with metastases to the lungs and heart was reported by Campbell *et al* (18). In the present case, the metastatic mass was located in the septum, which was similar to the case by Akiyama *et al* (17), while Campbell *et al* (18) reported a mass in the left ventricle. In spite of the lack of histology, we hypothesized that the septal lesion in the present patient was a metastatic tumor, as the incidence of primary cardiac tumors ranges from 0.001 to 0.030% (19) and the mass developed in a short time period.

The traditional treatments for ASPS consist of surgery and radiotherapy (15,17). Treatment should begin with mass resection; in particular, a complete resection should be performed when possible. If only partial excision or a questionable surgical margin or symptomatic metastatic mass exist, radiotherapy should be added as a supplementary procedure. In the present case, the patient presented with headaches and nausea, which were produced by metastatic brain tumors, so the patient underwent palliative radiotherapy. Following the completion of radiotherapy, the symptoms were relieved and the patient’s quality of life improved. The role of adjuvant chemotherapy in ASPS remains uncertain (6). However, in advanced cases, such as the present patient with multiple metastases of the lungs, brain, spleen and heart, chemotherapy may be considered as a treatment option. Recently, a novel anti-tumor drug, cediranib, was used in a phase II trial by Kummar *et al* (15). Cediranib (AZD2171) is a potent, oral, small-molecule inhibitor of all three vascular endothelial growth factor receptor (VEGFR-1, -2 and -3) tyrosine kinases, which mediate angiogenesis and lymphangiogenesis (20,21). The study evaluated 43 metastatic, unresectable ASPS patients who were administered cediranib (30 mg) once daily in 28-day cycles, and it was observed that cediranib has substantial single-agent activity, producing an overall remission rate of 35% and a disease control rate of 84% at 24 weeks (15). The trial is still ongoing, and cediranib has not yet been approved for use in China.

Patients who are diagnosed with widespread metastases usually have a poor prognosis and ultimately succumb to their condition. It is reported that the median survival time in patients with these multiple metastases is 40 months (8,22). In the present case, the coexistence of ASPS with heart, lung, brain and spleen metastases in the 37-year-old male indicated a poor outcome. The patient succumbed 44 months after the initial ASPS diagnosed, which was similar to the aforementioned median survival time.

Magnetic resonance imaging (MRI) is an effective and highly sensitive tool for the diagnosis of ASPS. Typically, the MRI of ASPS features internal and external multilobulated signal changes, also presenting with high signals on T1-weighted imaging (T1WI) and T2WI. A low blood flow rate is observed in T1-weighted high signal images, while a high blood flow rate is observed on T1WI and T2WI, with multilobulated signal change (23,24). However, no MRI was performed for the present patient prior to the surgery three years previously.

In conclusion, the unusual primary location of ASPS in the forearm of a male adult, and the recurrence of ASPS with lung, brain, spleen and particularly cardiac metastasis, ensure that the present case is an exceptional example compared with those reported in the previous literature. It is difficult to make a timely diagnosis of ASPS, as patients mostly presented with a painless swelling, so that a histopathological examination is needed to confirm the diagnosis. In patients with these multiple metastases of the lungs, brain, spleen and heart, chemotherapy may be considered as a treatment option. Patients who are diagnosed with widespread metastases usually have a poor prognosis and ultimately succumb to their condition.
is required. MRI may be useful for the diagnosis when the symptoms of ASPS are non-specific. The main treatment principle is a radical resection, and radiation treatment is an adjunctive measure in selected patients. Radiation therapy should be considered for metastatic brain tumors, in order to relieve the symptoms and improve the patient's quality of life. In patients with ASPS, widespread metastases usually predict a poor prognosis. ASPS with cardiac metastasis are extremely rare. In general, treatments of cardiac metastasis are not found to be effective. In order to detect cardiac metastasis early, a heart examination should be performed in patients with ASPS.

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