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CASE REPORT

Alveolar soft-part sarcoma of the retroperitoneum

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Abstract A 33-year-old woman was referred to Iida Municipal Hospital because of left back pain. Computed tomography showed a tumor (17 × 11 × 10 cm) in the left retroperitoneal space. T1- and T2-weighted magnetic resonance imaging showed an inhomogeneous mass with marginal blood vessels. The tumor was resected via lumbar oblique incision with the thoraco-abdominal approach. The tumor weighed 1800 g and consisted of nests of 5–100 large, loosely arranged, polygonal cells, surrounded by capillaries, resembling alveoli. The tumor cells were rich in cytoplasm, containing periodic acid–Schiff- and diastase-positive granules and typical crystals. The pathological diagnosis was alveolar soft-part sarcoma (ASPS). Alveolar soft-part sarcoma is a rare soft-tissue tumor that accounts for approximately 0.5–1% of soft-tissue sarcomas. Such tumors originating in the retroperitoneal space are extremely rare. Herein is reported a case of ASPS of the retroperitoneum with radiological and pathological findings.

Key words alveolar soft part sarcoma, retroperitoneum, sarcoma.

Introduction

Alveolar soft-part sarcoma (ASPS) is a rare soft-tissue tumor that accounts for approximately 0.5–1% of soft-tissue sarcomas.¹ Alveolar soft-part sarcoma occurs most often in the soft tissues of the pelvis and lower limbs, and is very rare in the retroperitoneal space.^{1,2} Portera *et al.* reported that only six (8%) of 70 ASPS patients treated from 1959 to 1998 at MD Andersson Cancer Center had ASPS of retroperitoneal space origin.¹ We found only four other cases of ASPS of retroperitoneal space origin in the literature.^{3–6}

Here we report a case of ASPS that originated in the retroperitoneal space, along with radiological and pathological findings.

Case report

A 33-year-old woman was admitted to Iida Municipal Hospital with left back pain, cough, and fever. She had occasionally had slight left back pain for 4 years. Computed tomography showed a large tumor (17 × 11 × 10 cm) in the left retroperitoneal space (Fig. 1). The tumor was restricted to the left kidney, stomach, spleen, and pancreas without continuity.

T1- and T2-weighted magnetic resonance imaging (MRI) showed an inhomogeneous mass with marginal blood vessels (Fig. 2). Laboratory investigation revealed no endocrine abnormalities.

Resection of the tumor was performed via lumbar oblique incision with the thoraco-abdominal approach (tumor weight, 1800 g; operation time, 297 min; bleeding volume, 1250 mL). Postoperative course was uneventful, and cough and fever both disappeared. Pathological diagnosis was ASPS with vascular invasion, and there was no continuity with the adrenal gland.

The patient refused adjuvant chemotherapy. Currently, 6 months after surgery, there is no local recurrence or metastasis.

Discussion

Alveolar soft-part sarcoma has a characteristic appearance on light microscopy,⁷ consisting of nests of 5–100 large, loosely arranged, polygonal cells, surrounded by capillaries, resembling alveoli (Fig. 3). The tumor cells are rich in cytoplasm, containing periodic acid–Schiff- (Fig. 4) and diastase-positive granules and typical crystals. The nuclei are round or oval, have an irregular chromatin pattern, and their nucleoli are clear. Mitoses are infrequent. Although there is no general consensus, the origin of ASPS

is considered to be myogenic.⁸

Alveolar soft-part sarcoma tends to be of equal or slightly higher signal intensity than skeletal muscle on T1-weighted MRI,⁹ and to have a high and heterogeneous signal intensity on T2-weighted MRI (Fig. 2).^{9,10} The presence of a large soft-tissue mass associated with large peritumoral vessels (Figs 1,2) is strongly suggestive of ASPS.¹⁰ There is strong and almost uniform enhancement with i.v. contrast medium.¹⁰ Small areas that fail to have enhancement may represent tumor necrosis. These findings were also seen in the present case.

Alveolar soft-part sarcoma affects primarily younger patients: the peak age of incidence is between 15 and 35 years.¹ Alveolar soft-part sarcoma is also characterized by unusual patterns of metastatic spread. For example, brain metastases have been reported as a common feature of metastatic ASPS, whereas they are relatively unusual with other high-grade sarcomas. Portera *et al.* reported that the 5-year actuarial survival rate of localized ASPS was 88%, and that the 5-year disease-free survival rate was 71%.¹ Alveolar soft-part sarcoma is very rare in the retroperitoneal space, so the difference in ASPS clinically and prognostically between the pelvis, lower limbs and in the retroperitoneum is not known. In the present case there was vascular invasion. This patient will be followed up, taking special care regarding brain metastasis.

In summary, we report a case of ASPS in a young woman with radiological and pathological findings. Such lesions occurring in the retroperitoneal space are very rare.

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- Fig. 1** Enhanced computed tomography showing a vascular mass measuring 17 × 11 × 10 cm in the retroperitoneal space.
- Fig. 2** Alveolar soft-part sarcoma tends to have a high and heterogeneous signal intensity on T2-weighted magnetic resonance imaging.
- Fig. 3** Hematoxylin–eosin staining. The tumor consisted of nests of large, loosely arranged, polygonal cells, surrounded by capillaries, resembling alveoli.
- Fig. 4** Periodic acid–Schiff (PAS) staining. The tumor cells were rich in PAS-positive cytoplasm.

Fig.1



Fig.2

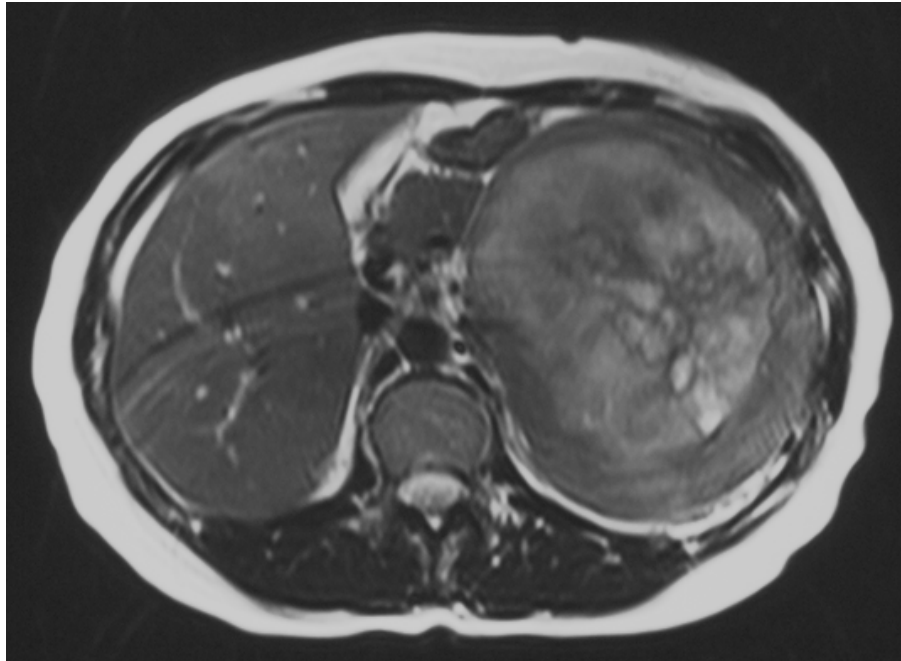
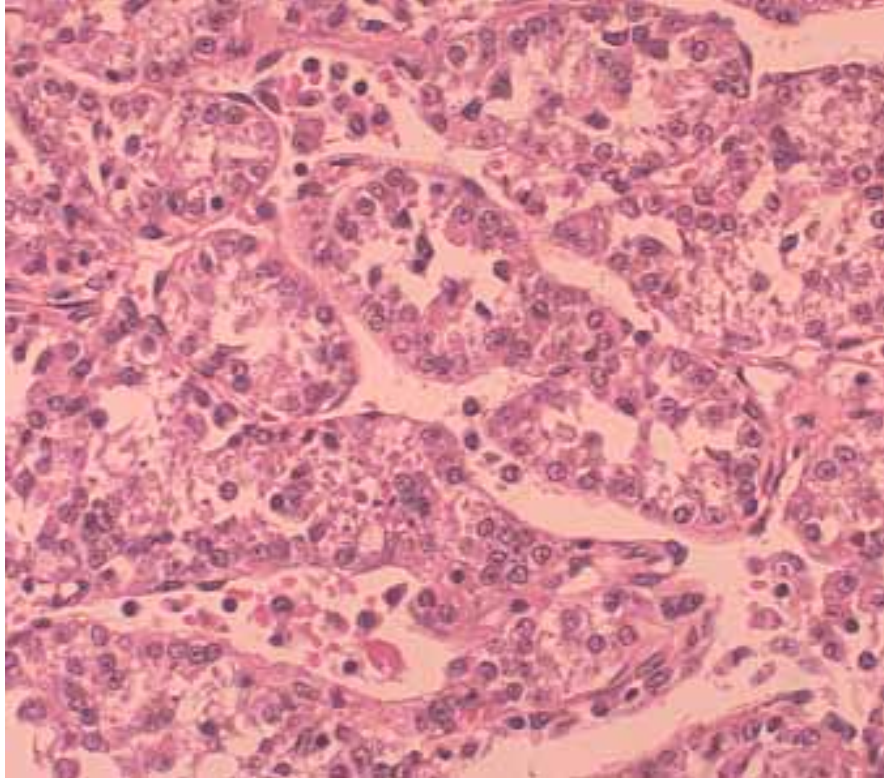
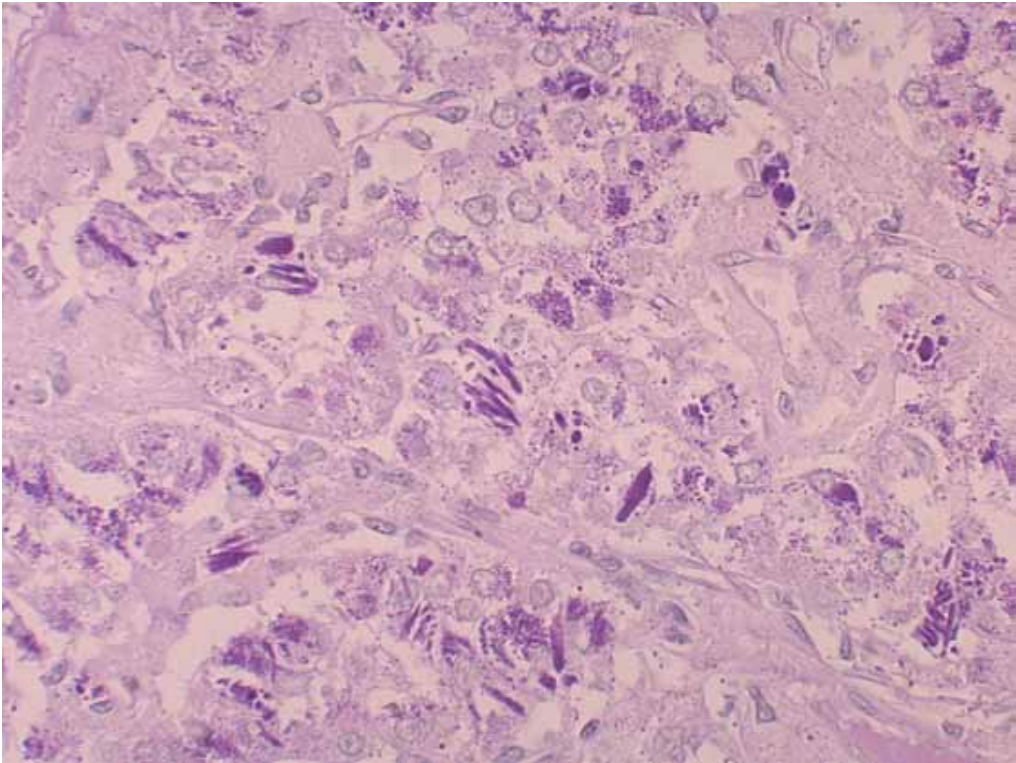


Fig.3



400x

Fig.4



400x