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A case of retroperitoneal vascular malformation

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Introduction

The occurrence of retroperitoneal vascular malformations is rare,[1] and the accurate diagnosis of these tumors is frequently difficult using available imaging modalities.[2] Here, we report a rare case of retroperitoneal vascular malformation that was histopathologically diagnosed using a specimen obtained during ultrasound-guided fine needle biopsy.

Case presentation

A left retroperitoneal tumor was incidentally detected in a 56-year-old Japanese man during computed tomography (CT) examination. The patient had no symptoms and medical history. CT revealed a 7 × 5 × 4-cm-sized solid mass with an irregular margin that was located in the retroperitoneum just below the left kidney. The tumor was slightly enhanced and had several small calcifications in its peripheral area. The tumor extended beyond the lateroconal fascia (Fig. 1). A magnetic resonance imaging (MRI) scan showed a tumor with iso-signal intensity on T1-weighted images, high-signal intensity on T2-weighted images, and no abnormal signal on a diffusion-weighted image (Fig. 2). Positron emission tomography revealed a tumor with no abnormal uptake. Test results for tumors markers and other screening laboratory data were within normal range. The patient underwent ultrasound-guided fine needle biopsy. Histopathological examination revealed proliferation of smooth muscle with surrounding fibrous tissues, soft tissues and stratified squamous epithelium (Fig. 3). Immunohistochemical findings were positive for cluster of differentiation 31, desmin, and elastic van Gieson, but negative for D2-10. No abnormal proliferation was observed in the vascular endothelia (Fig. 4). An immunohistochemical feature and the absence of abnormal proliferation of the tumors led to a final diagnosis of vascular malformation. Because the patient was asymptomatic, watchful waiting was selected as the treatment strategy. There have not been any notable changes in his case during the year that has passed since the diagnosis.

Discussion

The classification for vascular anomalies was established by the International Society for the Study of Vascular Anomalies in 1996. The main organizational principle of this classification is categorizing vascular lesions into vascular tumors (neoplastic), vascular malformations (non-plastic), and provisionally unclassified vascular anomalies.[3] Vascular malformations are usually benign but have vascular tumor-like growth patterns. They usually manifest at birth or soon thereafter. Abnormal dense collections of blood vessels may occur in the muscles, internal organs and mucous membranes or more commonly on the skin surface. Vascular malformations are histopathologically classified as capillary, lymphatic, venous, arteriovenous and combined types. Furthermore, they are classified as low- and high-flow types based on the hemodynamic status.[4] In our case, vascular malformation was diagnosed because no abnormal proliferation was observed in the vascular endothelia. However, we could not classify the type of vascular malformation using the small needle biopsy tissue.

Retroperitoneal neoplasms are relatively rare.[5] Among retroperitoneal tumors, vascular malformations are extremely rare. The diagnosis of retroperitoneal tumors is difficult because they are located deep within the trunk. CT and MRI are usually used for the diagnosis of such tumors. Plain radiography usually demonstrates a soft-tissue mass. Calcifications and phleboliths may also be present and are characteristics of the low-flow type. MRI offers the additional advantage of demonstrating flow dynamics of tumors and thus has now become the first choice for evaluating these tumors. However, accurate preoperative diagnosis of retroperitoneal tumors is difficult because the degree of

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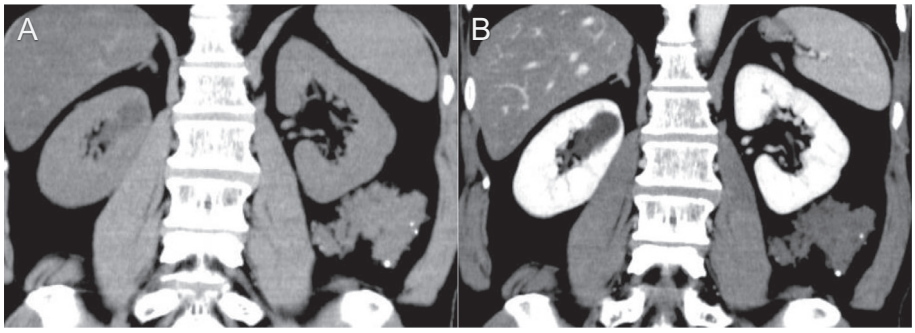

Fig. 1. Plain computed tomography showing a 7 × 5 × 4-cm-sized tumor with an irregular margin and several small calcifications in its peripheral area (A) and with no enhancement (B).

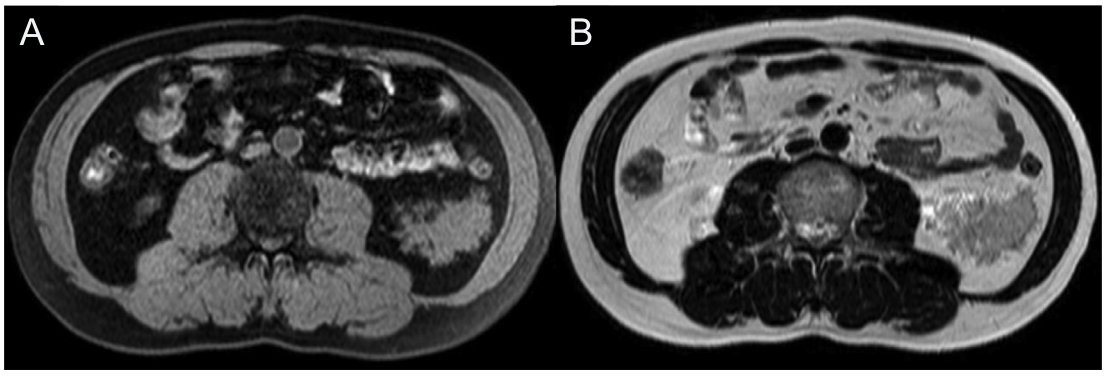

Fig. 2. Magnetic resonance imaging scan showing a tumor with iso-signal intensity on T1-weighted images (A) and high-signal intensity on T2-weighted images (B).

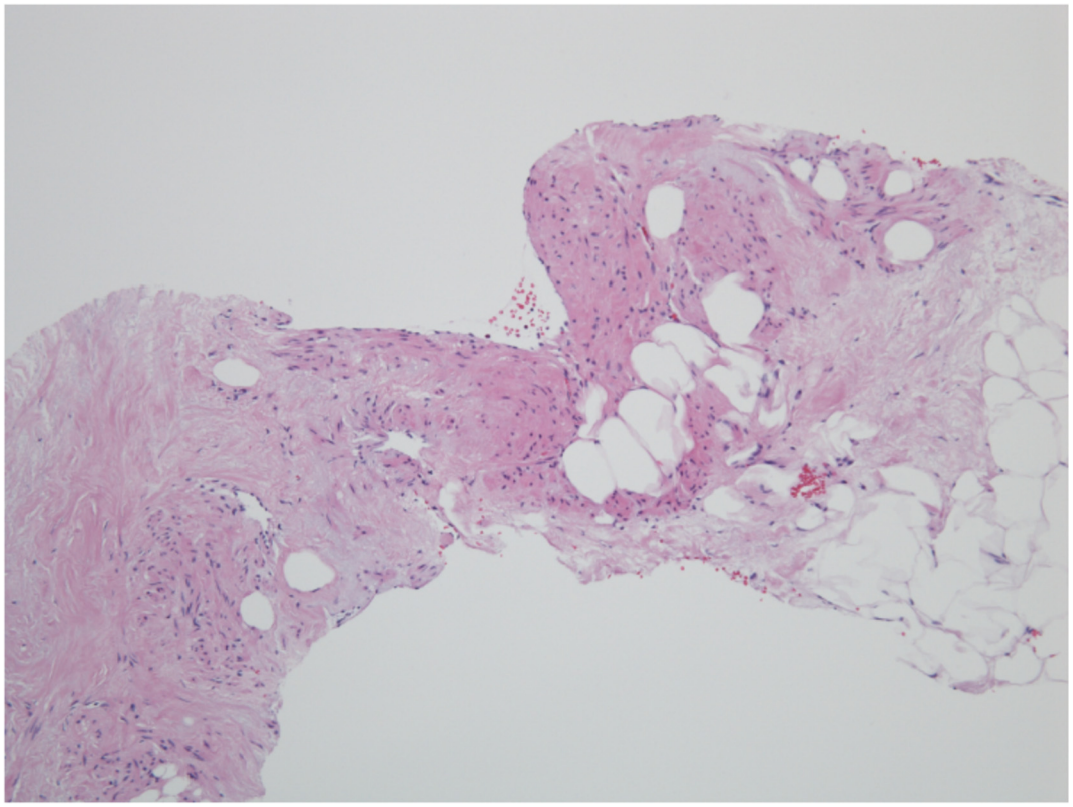

Fig. 3. H and E, 10 × , Photomicrograph obtained at histological examination showing proliferation of smooth muscle with surrounding fibrous tissues, soft tissues and stratified squamous epithelium.

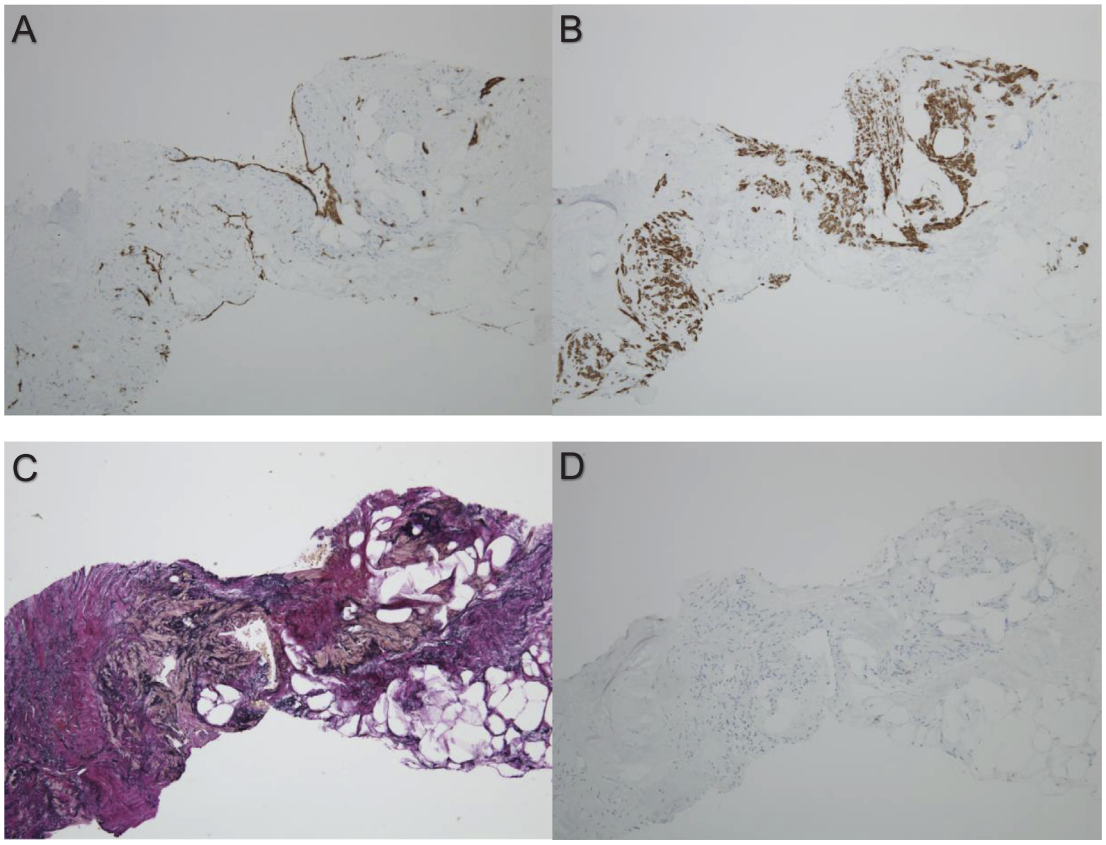

Fig. 4. Immunohistochemical findings were positive for cluster of differentiation 31 (A), desmin (B), and elastic van Gieson (C), but negative for D2-10 (D).

signal intensity may vary depending on specific tumor components. In our case, a preoperative diagnosis could not be made using various imaging modalities. In addition, we could not exclude other retroperitoneal tumors (e.g., neurogenic tumors, sarcoidosis, amyloidosis and lymphangiogenic tumors). These differential diagnoses of the tumor are essentially benign neoplasms that manifest clinical symptoms only when the tumor compresses an adjacent organ or tissue. Watchful waiting is usually a common treatment strategy for asymptomatic vascular malformation.[4] However, in our case, needle biopsy helped us to obtain a precise diagnosis. Because such tumors are benign and asymptomatic, we could avoid invasive procedures and select watchful waiting as the treatment strategy.

Conclusion

Needle biopsy is useful for obtaining precise diagnoses of unidentified tumors in the retroperitoneum. When encountering a tumor that cannot be identified using imaging modalities, needle biopsy should be performed.

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