CASE REPORT

Atypical radiological features of a leiomyosarcoma that arose from the ovarian vein and mimicked a vascular tumour

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ABSTRACT. Leiomyosarcomas of the ovarian vein are extremely rare tumours. Most tumours are hypervascular and typically have a large avascular centre of necrosis. We experienced a case of a retroperitoneal leiomyosarcoma that arose from the ovarian vein with marked vascular proliferation and dilatation within the tumour. To the best of our knowledge, this is the first case report of a retroperitoneal leiomyosarcoma with an atypical vascular structure in the clinical literature.

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A leiomyosarcoma is a rare malignant tumour of the smooth muscle that arises in the alimentary tract, retroperitoneum, genitourinary tract or soft tissue. A primary leiomyosarcoma of vascular origin is a rare tumour, which arises most often from the inferior vena cava [1]. A few cases of leiomyosarcoma of the ovarian vein have been reported in previous studies [2–4]. Commonly, a retroperitoneal leiomyosarcoma is seen as a large mass containing a prominent cystic necrotic area. Recently, we experienced a case of a retroperitoneal leiomyosarcoma that arose from the right ovarian vein and had multiple intratumoural dilated vessels. In this case report, we describe the imaging findings of this tumour and review the relevant literature.

Case report

A 58-year-old woman visited our hospital because of the presence of a palpable mass in the right lower abdomen. Laboratory findings, including tumour markers, were all normal.

Ultrasound of the right lower quadrant showed the presence of a large echogenic, partially cystic mass. A colour Doppler sonogram demonstrated increased blood flow within the mass. A duplex Doppler sonogram also demonstrated increased blood flow and pulsatile venous waveforms within the tumour (Figure 1). Contrast-enhanced CT revealed the presence of a 9 cm well-circumscribed, heterogeneous retroperitoneal mass containing central low attenuation. Characteristically enhancing tortuous tubular structures, thought to be dilated vessels, were seen within the mass (Figure 2a). The vascular structures seemed to be connected to the right

ovarian vein, and the upper portion of the right ovarian vein was distended (Figure 2b). The mass seemed separated from the kidney, small bowel and inferior vena cava. The differential diagnoses should include a retroperitoneal mass with a large area of central low attenuation and intratumoural vessels, such as a haemangiopericytoma, angiosarcoma or leiomyosarcoma.

The patient underwent resection of the retroperitoneal mass. The mass was attached and was connected to the right ovarian vessels (Figure 3a). There was no definite invasion of the adjacent organs and the mass was found to originate in the right ovarian vein. Upon examination of a gross specimen, abundant large-calibre, thin-walled vascular spaces were found throughout the mass (Figure 3b). A microscopic examination revealed a tumour composed of interlacing fascicles of spindle cells with blunt-ended nuclei. The tumour cells showed moderate to severe nuclear atypia and frequent mitoses. The central portion of the tumour showed necrosis, hyalinisation and numerous large-sized, thin-walled vascular spaces (Figure 4). The use of immunohistochemical staining demonstrated that the tumour cells expressed smooth muscle cell antigens, such as smooth muscle actin and desmin, but showed no staining for S-100 or CD34 antigens. A diagnosis of leiomyosarcoma was made.

The patient was discharged from the hospital to home on the fifth post-operative day and remained symptom free at follow-up 2 months after discharge.

Discussion

Primary leiomyosarcomas of veins are rare tumours that arise mainly from the inferior vena cava. Moreover, leiomyosarcomas that arise from the ovarian vein are extremely rare – five cases have been reported in the clinical literature [2].

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Figure 1. A duplex Doppler sonogram shows hypervascularity within the solid mass and pulsatile venous waveforms in the intratumoural vessels.

A typical retroperitoneal leiomyosarcoma is solid, with large, conspicuous cystic zones corresponding to areas of necrosis [5, 6]. The presence of a solid or necrotic extraluminal retroperitoneal mass that does not originate from a retroperitoneal organ, and that has a contiguous intravascular enhancing appearance on CT scans, is virtually pathognomonic for a vascular leiomyosarcoma [7]. Tumours with large cystic areas of necrosis typically demonstrate an avascular centre surrounded by a thick hypervascular rind [7]. In our case, the tumour was attached to the right ovarian vein but showed extensive vascular proliferation and dilatation within the mass as seen on sonography and CT imaging. The dilated vascular structures were connected from the right ovarian vein. Thus, it was not difficult to presume that the mass originated in the right ovarian vein, but we

could not assume that the tumour was a leiomyosarcoma at first impression owing to the presence of prominently dilated intratumoural vessels.

A histological examination showed that the areas of central low attenuation in the tumour were consistent with necrosis and hyalinisation, and that the large-sized tumour vessels corresponded to the enhanced vascular structures demonstrated on CT scans. To the best of our knowledge, this is the first case report of a retroperitoneal leiomyosarcoma with marked intratumoural vessels in the English-language literature. We think that there are two possible causes of the dilated intratumoural vessels. First, the presence of intratumoural arteriovenous shunting may be suspected, as indicated on colour Doppler sonography by large internal vessels with low vascular impedance and arterialisation of the venous flow with increased pulsatility [8]. Second, the dilated vessels maybe associated with a neoplasm. Van Kints et al [9] described the progressive enlargement and dilatation of veins over several years of follow-up, during which the size of the tumour increased, suggesting that the appearance of veins was related to the presence of the neoplasm. In our case, increased arteriased venous pulsatile flow suggests the presence of intratumoural arteriovenous shunting.

The differential diagnosis for a mass containing intratumoural vessels could include angioliipoma, haemangioma and haemangiopericytoma. Vessels visible within an angioliipoma or a haemangioma do not resemble the large intratumoural vessels seen in the present case. Haemangiopericytomas often have large intratumoural vessels [10], but large vessels especially are typically located at the periphery. Moreover, the histology and immunohistochemical staining of the tumour in our case were compatible with leiomyosarcoma rather than with haemangiopericytoma. The tumour was characterised by fascicles of spindle cells with blunt-ended nuclei, diffuse smooth muscle actin and desmin positivity, and CD34 negativity.

(a)

(b)

Figure 2. (a) An axial contrast-enhanced CT image shows a well-defined retroperitoneal mass with an area of central low attenuation and centrally discrete enlarged vessels. (b) A coronal contrast-enhanced CT image shows intratumoural vessels connected to the right ovarian vein (arrow).

(a)

(b)

Figure 3. (a) Upon examination of a gross specimen, the mass is contiguous with the right ovarian vein. (b) On sectioning, the mass shows a solid appearance with a central yellowish area of degeneration and numerous irregular large vascular spaces.

Figure 4. A microscopic examination reveals a tumour composed of numerous, irregular, thin-walled and large-calibre vascular spaces.

In conclusion, our case showed an extremely rare leiomyosarcoma that arose from the ovarian vein but that was different from previously reported cases: the lesion showed highly vascular proliferation and dilatation within the tumour. Imaging findings indirectly suggested the presence of intratumoural arteriovenous shunting. When a retroperitoneal mass shows a vascular structure within the mass, a retroperitoneal leiomyosarcoma should be considered in the differential diagnosis.

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