CASE REPORT

Lymphangioma: surrounding the ovarian vein and ovary

Korcan Aysun Gonen,[1] Remzi Abali,[2] Meltem Oznur,[3] Cuneyt Erdogan[4]

[1]Department of Radiology, Namik Kemal University, School of Medicine, Tekirdag, Turkey
[2]Department of Gynecology and Obstetrics, Namik Kemal University, School of Medicine, Tekirdag, Turkey
[3]Department of Pathology, Namik Kemal University, School of Medicine, Tekirdag, Turkey
[4]Department of Radiology, Uludag University, School of Medicine, Bursa, Turkey

Correspondence to
Dr Korcan Aysun Gonen, aysunbalc@yahoo.com

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SUMMARY

Lymphangiomas are usually benign lesions seen in the head and neck region in children. Intra-abdominal localisation is rare and the majority of these cases are in early childhood. Retroperitoneal lymphangiomas constitute approximately 1% of all lymphangiomas. They are generally diagnosed incidentally, may be asymptomatic or may present with a palpable abdominal mass. A limited number of cases of ovarian lymphangiomas have been reported in women, whereas there are no reported cases of paraovarian localisation. We present a rare case of lymphangioma located in bilateral paraovarian region and along the left ovarian vein with radiological findings.

BACKGROUND

Lymphangiomas are benign lesions characterised by proliferation of lymphatic vessels. Approximately 50% of all are presented at birth and 90% are diagnosed before the age of 2.[1] During this period, the most common localisation is head- neck region. Intra-abdominal lymphangiomas have been reported in the gastrointestinal tract, mesentery, liver, pancreas, gallbladder, spleen and adrenal gland.[2] There are reported cases in the pelvic area, rarely in ovaries or even in fallopian tube and inguinal region.[3–8] Retroperitoneum is the second most common localisation for the abdominal lymphangiomas after small bowel mesentery. It is seen in elderly and is often asymptomatic and diagnosed incidentally.

We present a rare case of a lymphangioma localised in bilateral paraovarian region, extending up to the renal region by surrounding left ovarian vein in a woman in menopause, together with multidetector CT (MDCT) findings.

CASE PRESENTATION

A 54-year-old female patient admitted with long-standing mild abdominal pain and abdominal distension. There was nothing important in medical history and physical examination.

With a prediagnosis of ovarian cystic lesion and uterine myoma, a whole abdominal imaging was performed by 16-slice MDCT (Bright Speed 16; General Electric Medical Systems Co., Ltd, Milwaukee, Wisconsin, USA). The CT scan demonstrated multiloculated hypodense lesions, larger on left (approximately 3×5 cm in size), in bilateral adnexal region. Lesion on the left side was extending by surrounding ovarian vein up to the point about 2–3 cm distal to point of joining with renal vein (figure 1). Its density was consistent with liquid, and it was hypointense on T1-weighted MRI and hyperintense on T2-weighted MRI and the lesion was not enhanced on postcontrast images. In addition, examinations revealed multiple subserosal and intramural degenerated myomas, largest 10 cm in diameter and with intense homogeneous contrast enhancement, at the level of the uterine corpus and fundus. Ascites was not detected.

TREATMENT

The patient underwent abdominal hysterectomy with bilateral salpingo-oophorectomy. Fallopian tubes were normal and both ovaries were atrophic intraoperatively. Yellow multilocular cystic masses with chylous content were present in bilateral paraovarian region, extending to parametrium and renal region by surrounding left ovarian vein. In addition, nine intramural and subserosal myomas, the largest having a diameter of 10 cm and the smallest 0.7 cm, were found in the uterus.

OUTCOME AND FOLLOW-UP

Histopathological examination of the excised-multilocular cystic masses was compatible with lymphangioma (figure 2). In a follow-up period of 1 year, clinical and radiological examinations revealed no recurrent disease.

DISCUSSION

Most of retroperitoneal lymphangiomas are asymptomatic and are usually discovered incidentally during radiological examinations made for any reason in the elderly, or during surgery and autopsy. Clinically the most common findings are slow-growing abdominal mass, abdominal pain, loss of appetite and nausea and vomiting. Cystic growth occurs due to increase in lymphatic flow and closure of drainage channels. Thus, abdominal distention and compression to adjacent organs occur. Ascites, bleeding, rupture, torsion and volvulus are rare complications of retroperitoneal lymphangiomas, and severe abdominal pain and fever may occur.[9 10]

Lymphangiomas are divided into three types according to size of lymphatic gaps, as capillary or simple, cavernous and cystic.[11] Cystic masses contain chylous or serous content and are lined with a layer of endothelium; are also known as cystic hygroma. They have soft consistency and typically develop in the fetus. Seventy-five per cent of them are seen in neck, axillar and inguinal region.[8] Retroperitoneal lymphangiomas are mostly cystic in nature.[12]

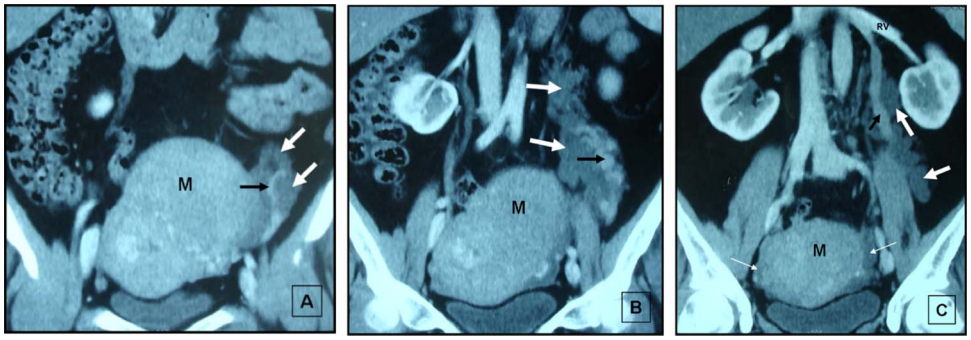

Figure 1 Venous phase coronal multiplanar reconstruction CT images show multiloculated cystic lesion (*white arrows*) along dilated left ovarian vein (*black arrow*) (A–C), and in bilateral adnexal region (*thin white arrow*) (C), degenerated myoma (M), 10 cm in diameter and with intense homogeneous contrast enhancement. M, myoma, RV, renal vein.

A variety of disorders such as inflammatory, fibrotic and genetic factors, mechanical pressure, traumatic causes and degeneration of lymph nodes, endothelial lymphatic vascular permeability disorders are responsible for development of lymphangiomas.[1]

Ovarian lymphangiomas are rare and have been published in a few reports. Most of the them are unilateral.[3–5 13] The clinical significance is less and considered to be benign. They are usually asymptomatic and diagnosed incidentally during ultrasonographic examination or surgery.[3] The aetiology is not clear; some authors accept it as a true neoplasm, while some authors claim that they develop as a result of lymphatic proliferation which result from lymphatic circulation disorders, hamartoma and malformations.[3 4] Disruption of lymphatic drainage was held responsible for ovarian lymphangiomas in two female patients who respectively had bilateral chronic follicular salpingitis and received radiotherapy and chemotherapy due to Wilm's tumour.[4 14] In another case, ovarian lymphangioma was accompanied by chylous acid.[13] Akyildiz *et al*[3] have reported a case of ovarian lymphangioma in menopausal period and deterioration of lymphatic circulation due to leiomyoma, 35 cm in diameter, was held responsible for this. In our case, there were also multiple myomas with the biggest having a diameter of 10 cm and lymphatic drainage could possibly be impaired as a result of compression of these myomas. However, the ovaries of our patient were atrophic; the lesions were in the paraovarian region and were extending cranially by surrounding the ovarian vein on the left side. As far as we know, there is no case of such a lymphangioma previously defined in the literature.

Preoperative recognition of retroperitoneal lymphangiomas is rare. Radiological methods provide important information for determining the diagnosis and effective surgical approach, in particular MDCT and MRI demonstrate the nature of the lesion and the relationship between neighbouring organs. Definitive diagnosis is made by histopathological examination.

As spontaneous regression of the cyst is rare and there is risk of recurrence, surgical treatment is necessary. Simple total excision is the preferred method. Peritonitis, bleeding, abscesses and torsion may occur rarely after the surgery. Although spread to retroperitoneum is very rare, it is a fatal complication.[9 15]

Lymphangiomas in adult women can rarely be seen in ovaries. This is, to our knowledge, is the first case of lymphangioma which is localised in paraovarian region and around the ovarian vein, and they can be demonstrated by MDCT.

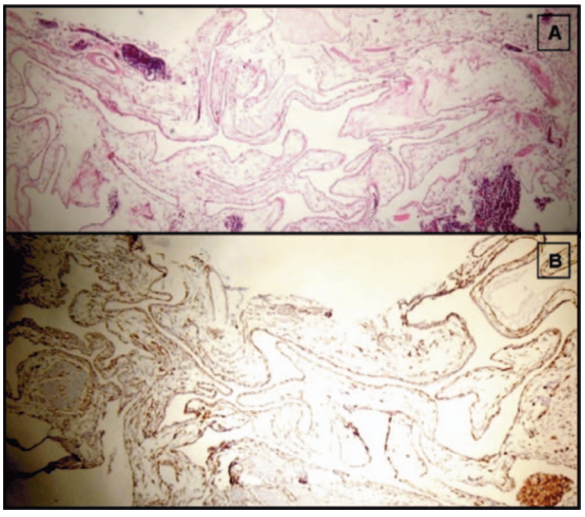

Figure 2 Histopathological examination: thin-walled lymphatic vessels, lined with squamous endothelium, which contain lymphatic aggregates in their stroma are seen (A). CD34 positivity of lymphatic vessels is shown by immunohistochemistry (B).

Learning points

- Lymphangiomas are usually seen in the head and neck region and pelvic localisation are rare.
- A limited number of cases of ovarian lymphangiomas have been reported in women, whereas there are no reported cases of paraovarian localisation.
- Radiological methods, in particular multidetector CT and MRI provide important information for determining the diagnosis and effective surgical approach, but definitive diagnosis method is histopathological examination.
- It should be considered that lymphangiomas can be localised in paraovarian region and around the ovarian vein.

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Competing interests None.

Patient consent Obtained.

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