

Lymphangioma of the ovary

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ABSTRACT

Lymphangioma of the ovary is an extremely rare lesion. There are only 19 cases reported in the literature. Lymphangioma is usually unilateral and asymptomatic, presenting as an incidental finding during routine gynaecologic procedures. It is composed of aggregates of lymphatic spaces in ovarian stroma and the endothelial cells lining these spaces stain positively with CD-31 and CD-34 on immunostaining. The main differential diagnosis is an adenomatoid tumour which can be differentiated from the lymphangioma by immunohistochemical studies. We report this rare lesion in a 42-year-old Malay lady diagnosed after total abdominal hysterectomy and right salpingo-oophrectomy. A literature review is presented and the histological and immunohistological findings along with differential diagnosis are discussed.

Keywords: Adenomatoid tumour, lymphangioma, immunohistochemistry, ovary

INTRODUCTION

Lymphangioma is a benign lesion of the lymphatic vessels that is characterised by lymphatic proliferation. Lymphangioma of the ovary is extremely rare and only 19 cases have been reported.^{1,2} It is a benign vascular lesion, lined by a single layer of flattened endothelial cells. However, a single case report of its malignant counterpart has been reported.³ Earlier reports suggested lymphangiomas to be neoplastic but it is now believed to be hamartomatous in origin.^{4,5} Most are asymptomatic and are usually discovered incidentally during routine gynaecological procedures. Manifestation as chylous ascites has

also been reported.² On ultrasound imaging, lymphangioma may appear as septated cystic lesions.¹ Despite the benign nature, it is important for clinicians to be aware of its rarity and to consider it in the differential diagnosis of unilateral multi-cystic lesion of the ovary. We report this rare lesion in a 42-year-old Malay lady who was diagnosed after a total abdominal hysterectomy and right salpingo-oophrectomy. The histological features, immunohistological findings and differential diagnosis are discussed and the literature reviewed.

CASE REPORT

A 42-year-old Malay lady presented to the Gynaecology Department with a history of menorrhagia. She was known to have diabetes mellitus that was diagnosed during her

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first pregnancy and had been controlled with diet and oral hypoglycaemic agents. She had four previous normal deliveries. Apart from the occasional urinary tract infection during the ante-natal period, there was no other significant disease. Her present routine haematological and biochemical profiles were within normal range.

Vaginal examination revealed an enlarged 16-week sized uterus. The right ovary was noted to be enlarged. The left ovary was normal. A diagnosis of fibroid uterus was made by ultrasound scan. The

exact cause of the right ovarian enlargement could not be ascertained. Therefore, it was decided to proceed to total abdominal hysterectomy and right salpingo-oophorectomy. A biopsy of the left ovary was also done.

The specimen was of uterus with cervix measuring 16 x 12 x 10 cm with a 5 cm right fallopian tube and a 4 x 2 x 2 cm attached right ovary. The left ovarian biopsy was 1.2 x 0.5 cm grey white tissue. The cut surface of right ovary was greyish with tiny cysts. The cut surface of the uterus showed a seven cm diameter intramural fibroid. On

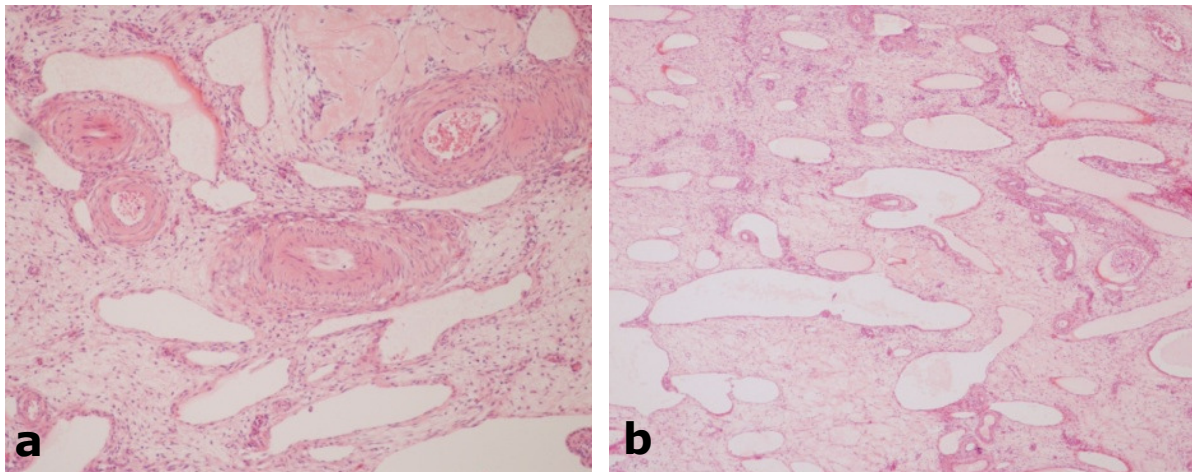


Fig 1: a) Histology of the ovary showing dilated endothelium lined vascular spaces in ovarian stroma. (H & E, x4), b) Histology showing dilated endothelium lined vascular spaces in ovarian stroma (H & E, x4).

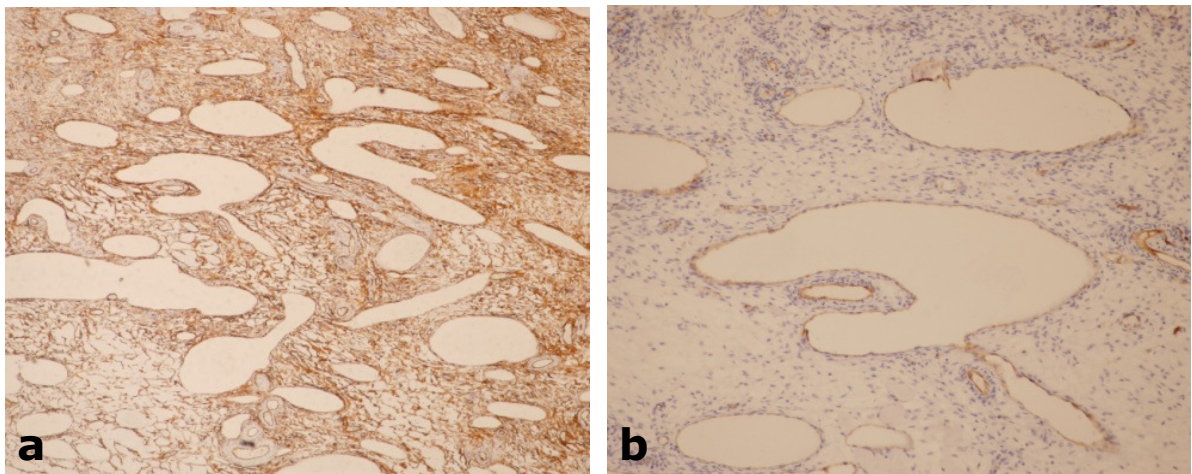


Fig 2: a) Histology of the ovary showing endothelium of lymphatic spaces and stroma staining positive (brown) with CD-34 (x4), b) Histology showing positive (brown) staining with CD-31 (x10).

histological examination, the right ovary showed a small follicular cyst. The ovarian stroma contained a large number of intercommunicating thin walled vascular channels lined by a single layer of flattened endothelial cells. Amorphous eosinophilic fluid was seen in the lumen. There was no endothelial hyperplasia and no necrosis or haemorrhage (Figures 1a and 1b). The lining cells stained positively with CD-31 and CD-34 (Figures 2a and 2b). These findings were consistent with a diagnosis of lymphangioma of the right ovary. The possibilities of haemangioma and adenomatoid tumour were excluded by the absence of blood in the lumen and negative AB/PAS, cytokeratin and vimentin staining respectively. The left ovarian biopsy showed a hyalinised corpus luteum. There were no dilated lymphatics. The cervix showed chronic cervicitis with squamous metaplasia. A mitotically active leiomyoma was in the uterus and the endometrium was in the proliferative phase. The right fallopian tube was normal with no vascular or lymphatic dilatation. The patient was well and asymptomatic when she was last seen in clinic.

DISCUSSION

The lymphatic system consists of a network of uni-directional vessels that collects excess fluid from the interstitium, transports it to the regional lymph nodes and ultimately drains to the venous system via the thoracic duct. The lymphatic vessels are seen in all organs except in the brain, anterior chamber of the eye and in systems with rich sinusoidal networks such as bone marrow and spleen.

Lymphangiomas are commonly seen in the head, neck and the upper body regions.

Intra-abdominal visceral lymphangiomas are very rare with most seen in relation to the intestine and the mesentery.⁶ Lymphangioma of the ovary is extremely rare. To our knowledge, this is the first case to be detected and reported in our local setting. Lymphangioma of the epididymis has been reported before in our local setting.⁷

The lesion is considered benign but a case of lymphangiosarcoma of the ovary has been reported.³ The lesion is usually unilateral but bilateral lesions have been reported.⁸ Earlier, lymphangiomas had been considered to be neoplastic in nature and capable of aggressive behaviour.⁴ However, some investigators now believe that lymphangiomas are hamartomas. Others believe that both neoplastic and hamartomatous theories are possible.⁵ Fibrosis of lymphatic channels following surgeries or infections leading to dilatation of the proximal channels has been postulated to be the underlying pathogenesis.⁸ Lymphangioma of the ovary following radiation therapy has also been reported. This was a case that had been previously treated for Wilm's tumour during childhood with radiation therapy.⁹ In our patient, the right ovarian stroma contained intercommunicating vascular channels lined by a single layer of flat endothelial cells that were immunoreactive to CD-31 and CD-34. Cytokeratin (LMW) and vimentin were negative. Our patient had an intramural mitotically active leiomyoma with normal parametrial vessels and lymphatics. There was no inflammation around the fallopian tubes. Since there was no evidence of infection and no obstructive pathology noted in the surrounding lymphatics, we believe the lesion to be hamartomatous.

The main differential diagnosis is an adenomatoid tumour of the ovary. Adenomatoid tumour is a benign solid tumour of mesothelial origin affecting both the male and female genital tracts. This tumour is positive for cytokeratin (LMW) and negative for the endothelial markers CD-31 and CD-34.⁵ The lesion was not due to pressure of the fibroid as the right fallopian tube and the uterus did not show any vascular or lymphatic dilatation.

Almost all the cases of lymphangioma of the ovary reported in the literatures were found incidentally during routine gynaecological examinations or procedures. Interestingly, most ovarian lymphangioma are asymptomatic. However most were found in the presence of other gynaecological pathologies.

In conclusion, our case highlights the importance of being aware of this rare entity and of considering it in the differential diagnosis in patients found to have multi-cystic lesions of the ovary even though it typically has a benign course.

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