A rare case of ovarian lymphangioma with elevated CA-125 causing pseudo-Meigs' syndrome

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Lymphangiomas are benign congenital malformations of the lymphatic system, thought to occur as a result of obstruction of the local lymph flow system. They can occur anywhere in the skin and mucous membranes. The most common sites are the head and neck, but sometimes they are found in the intestines, pancreas and mesentery. Lymphangioma of the ovary is an extremely rare lesion. It was first described in 1908. Clinical manifestations can vary, from an asymptomatic ovarian mass to an acute abdomen. These may be confused with malignant ovarian masses, resulting in extensive surgery. A 34-year-old woman presented with dull aching pain localised to the lower abdomen, present for the previous 6 months. Ultrasound and a contrast-enhanced computed tomography scan of the abdomen were suggestive of an enlarged right ovary with massive ascites. The cancer antigen 125 (CA-125) level was 685 units/mL. Diagnostic laparoscopy revealed chylous ascites, and further, a frozen section revealed a few atypical cells, additionally strengthening the diagnosis of a malignant tumour. Staging laparotomy with total abdominal hysterectomy and salpingo-oophorectomy was finally performed. Histopathology revealed lymphangioma, with no evidence of malignancy. CA-125 also showed a rapid decline following surgery. Lymphangiomas should be included in the differential diagnosis of ovarian cystic masses, especially in patients with chylous ascites. It is very important to discriminate such cases from other malignant tumours in order to avoid overtreatment. However, a careful follow-up for at least 2 years is needed for patients with lymphangioma of the ovary, to exclude recurrence.

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Lymphangiomas occur very rarely as tumours of the ovary. They are characterised by the presence of thin-walled vascular spaces lined by flat endothelial cells, with pale oeosinophilic homogeneous material within the vascular channels, which differentiates them from haemangiomas.[1] The most common sites of lymphangioma are the head, neck and upper body. Intra-abdominal lymphangiomas, although rare, are mostly found in relation to the intestine and mesentery.[2] Lymphangioma of the ovary is extremely rare. The majority of cases are benign, but some cases of malignant transformation into lymphangiosarcoma have also been reported.[3]

The presence of a pelvic mass in association with a markedly elevated cancer antigen 125 (CA-125) level is highly suspicious of a malignant ovarian tumour. The present case is reported in order to highlight the fact that there are also various benign conditions that can mimic such presentations, especially in premenopausal women, and lymphangioma of the ovary is one.

Case report

A 34-year-old woman came to the gynaecology out-patient department complaining of a dull aching pain localised to the lower abdomen for the previous 6 months. It was not associated with any bladder or bowel symptoms. There was no history of any menstrual abnormality. On examination, there was deep tenderness in the right iliac fossa, but no palpable lump or swelling. There was no organomegaly. On bimanual examination, the uterus was normal in size, and an adnexal mass of about 5 × 5 cm, soft in consistency, freely mobile and non-tender, was felt. The left adnexa was normal, and there was fullness in the pouch of Douglas. Per rectal examination was normal. Ultrasound revealed an enlarged right ovary with moderate ascites and mild pleural effusion. Contrast-enhanced computed tomography was suggestive of a multiloculated cyst in the right ovary, with thin internal septations and massive ascites.

Routine blood and urine investigations were normal. CA-125 was 685 units/mL. Carcinoembryonic antigen, lactic dehydrogenase and beta human chorionic gonadotropin were normal. In view of the markedly raised CA-125 and no definite cause of ovarian enlargement, a diagnostic laparoscopy was performed. About 1 500 mL of chylous fluid was present in the peritoneal cavity (Fig. 1). The right ovary was irregularly enlarged, about 6 × 6 cm,

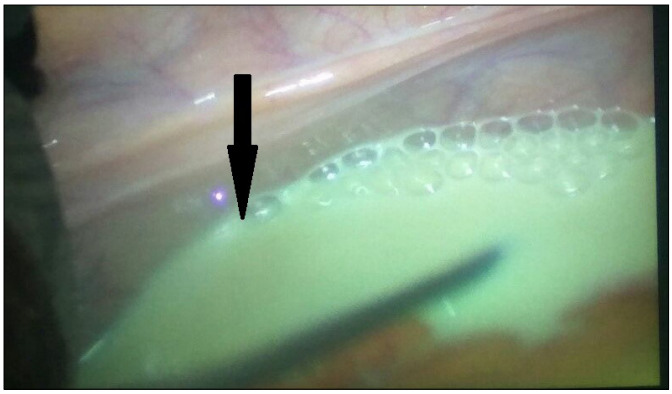

Fig. 1. Laparoscopic view showing the presence of chylous ascites.

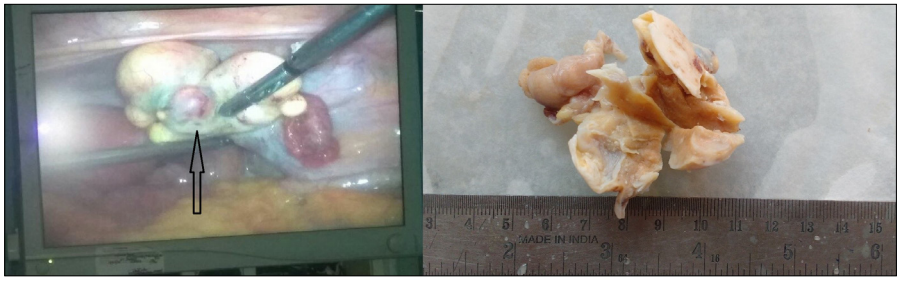

Fig. 2. Laparoscopic image of multi-cystic enlarged ovary mimicking malignancy, and cut section showing thin-walled cysts.

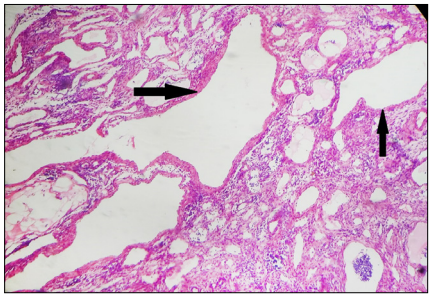

Fig. 3. 10 × 10 haematoxylin and eosin photomicrograph showing cystically dilated spaces lined by flattened epithelium, consistent with lymphangioma.

yellowish-white in colour and lobulated (Fig. 2). A right salpingo-oophorectomy was performed, and sent for frozen section, and was suggestive of many small cystic spaces filled with serous material, some showing atypical cells suggestive of malignancy. Staging laparotomy followed by total abdominal hysterectomy and bilateral salpingo-oophorectomy were performed. Histopathology suggested ovarian lymphangioma with no evidence of malignancy (Fig. 3). The patient recovered well in the postoperative period. Ca-125 fell to 162 units/mL on postoperative day 14, and <30 on postoperative day 30.

Discussion

Meigs' syndrome is defined as the presence of ascites and pleural effusion, together with fibroma or fibroma-like tumours, such as thecoma or granulosa cell tumour, in cases in which the removal of the tumour completely cures the patient.[4] Other benign tumours such as struma ovarii, mature teratomas or mucinous cystadenoma may also be associated. Fibroid uterus and secondary metastatic tumours of the ovary, if associated with hydrothorax, are referred to as pseudo-Meigs' syndrome.[5] This is probably the first case of lymphangioma as a cause of pseudo-Meigs' syndrome.

Lymphangiomas are generally benign tumours of the lymphatic system, composed of multiple cystic spaces lined by endothelium. They can be classified as cystic, capillary or cavernous, and may contain serous or chylous fluid.[6] Although they occur at any age, they are most common in the reproductive age group. Some cases have been described in neonates and children.[7,8] Owing to the paucity of cases, their exact incidence and malignant potential is not well known. They are usually benign, and only two cases with malignant transformation were found to have been reported to date.[3]

Their aetiopathogenesis is not clear. Some authors believe it to be due to sequestration of lymphatic vessels, which fail to communicate with the normal lymphatic channels, which may be either congenital or acquired, due to mechanical pressure, trauma, degeneration of lymph nodes, radiation, infection or surgery.[9] Proliferation of lymphatic nests due to chronic inflammation, especially in genetically predisposed individuals, has also been implicated as a causative factor.[10] In our patient, there were no signs of chronic inflammation in the peritoneal cavity, and so the exact cause of lymphangioma could not be ascertained. Chylous ascites is a very uncommon clinical finding, usually occurring are a result of the collection of thoracic or intestinal lymph in the abdominal cavity.[11,12] It is usually associated with chronic disruption of lymphatic flow, resulting from any cause. The cause of lymphatic obstruction may not be discernible in all cases. In the present case, malignant ovarian tumour was suspected at the initial medical examination owing to the presence of ascites associated with markedly raised CA-125.

CA-125 is a 200 kDa glycoprotein, and although initially identified on the surface of ovarian cancer cell line OVCA 433, it is widely distributed on the surface of both healthy and malignant cells of mesothelial origin, including pleural, pericardial, peritoneal and endometrial cells, as well as in the normal genital tract and amniotic membrane.[13] It is not present on the surface of normal ovarian cells, but is present in 80% of non-mucinous malignant ovarian tumours.[14] A very few cases of benign tumours, such as haemangioma of the ovary associated with a markedly raised CA-125, have also been reported in the literature.[15,16] However, to the best of our knowledge, no case of lymphangioma presenting with such high levels of CA-125, and presenting as pseudo-Meigs' syndrome, has been reported in the literature to date.

A careful follow-up for a period of at least 2 years is recommended in cases of lymphangioma, since they have a high recurrence tendency.

The present case has been reported owing to its rarity, and to highlight the fact that clinical suspicion of lymphangioma could have prevented the extensive surgery that this patient had to undergo at a young age.

Conclusion

Ovarian lymphangioma should also be part of the differential diagnosis in patients presenting with ovarian enlargement, especially in association with chylous ascites. Clinical suspicion of lymphangioma, along with a thorough knowledge regarding its management, could prevent radical surgery, especially in premenopausal women.

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