Lymphangioma of Ovary

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Abstract

Lymphangioma of the ovary is a very rare tumour. The debate whether it is a hamartoma or a neoplastic entity is still ongoing. We report a case of lymphangioma of the ovary in a young woman. In this article, we describe the clinical presentation, histopathology and immunohistochemistry findings of the lymphangioma of ovary that are helpful to both gynaecologists and pathologists in the diagnosis of this rare entity.

Introduction

Lymphangiomas are uncommon, hamartomatous, often congenital malformations of the lymphatic system that usually involve the skin and subcutaneous tissues. Lymphangiomas can occur anywhere in the skin and the mucous membranes. Most common sites are the head and the neck, followed by the proximal extremities, the buttocks, and the trunk. However, they sometimes can be found in the intestines, the pancreas, and the mesentery. Lymphangioma of the ovary is a very rare tumour. It was first described in 1908[1] and as of 2006 only 17 cases have been described in the literature.[2]

Case Report

A 29 year old married female presented with complaints of menorrhagia since 6 months. There was history of dysmenorrhoea and leucorrhoea since 4 months. Ultrasound abdomen was suggestive of uterine fibroid in the posterior myometrium measuring 5.4 x 4.9 x 4.3 cm with a bulky left ovary. A contrast MRI showed a posterior wall fibroid and a bulky left ovary with prominent stromal tissue. FSH, LH, prolactin and CA 125 levels were in normal range.

Left sided salpingo- oophorectomy with removal of the fibroid with endometrial curetting and a cervical biopsy was performed. The right ovary was found to be unremarkable. On gross examination, the left ovary measured 5 x 4.5 x 2.5 cm. On cut surface multiple small thin walled cysts were seen at the cortex. The rest of the ovary was oedematous, whitish and spongy (Fig.1). The fallopian tube was unremarkable. The fibroid measured 4 x 3.5 x 2.5 cm.

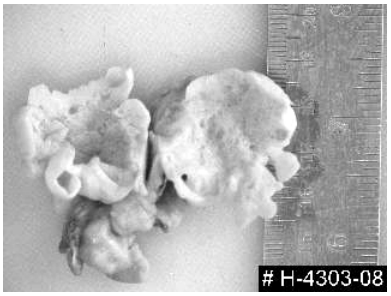

Fig. 1 Gross Photograph showing cut surface of left ovary showing multiple small thin walled cysts at the cortex.

On microscopic examination, the ovarian cortex contained multiple dilated follicular cysts, some of which were partially luteinized. The stroma deeper to that contained numerous vascular spaces of different sizes of which the inner surface were lined with flattened endothelial cells, showing neither cellular atypia nor proliferation. Few small lymphocytes were seen in the lumena (Fig. 2). The intervening stroma was fibrocollagenous and contained few scattered lymphocytes. IHC studies with CD 31 were positive in the cells lining the spaces confirming their endothelial nature (Fig. 3). The fallopian tube was unremarkable. The fibroid was seen to be an adenomyoma. The endometrial curettings revealed a proliferative pattern with a small hyperplastic endometrial polyp. The cervical biopsy showed mild

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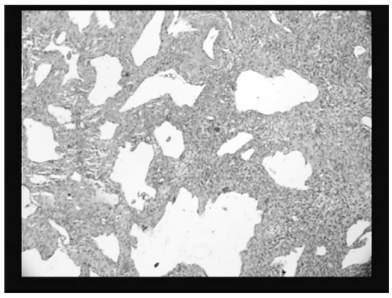

Fig. 2 Microscopy showing vascular spaces lined with flattened epithelial cells H→E Stain, 40x

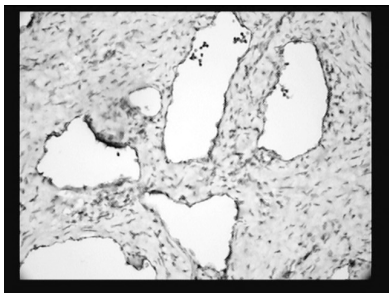

Fig. 3 IHC study shows CD31 positive cells lining the vascular spaces confirming their endothelial nature. Stain for CD31, 40x

endocervicitis.

Thus, a diagnosis of lymphangioma of the left ovary with multiple cortical follicular cysts was made. The patient's post operative period and subsequent follow up was uneventful.

Discussion

Lymphangioma of ovary is a very rare tumour with only 17 cases reported. The tumour is usually unilateral but bilateral cases have been reported.[3] Most of the reported cases are in the age group of 40 to 60 years, though one case was reported in a child.[4] The condition may be asymptomatic, and found incidentally, or may cause menstrual disturbances. The natural history of lymphangiomas is unpredictable although slow growth and indolent behaviour is usual. In fact, the malignant potential of this lesion is still not known because of the paucity of cases, with no reports of long term follow up in these cases. The pathogenesis of ovarian lymphangiomas is unclear and controversial. Various authors consider them to be hamartomatous malformations[5] or neoplasms.[6]

Macroscopically the tumour is small, with a smooth grey surface. On cut section, it is yellow, honeycombed and composed of numerous small cystic spaces exuding clear yellow fluid. Microscopically, lymphangioma of ovary is composed of closely packed thin walled vascular spaces lined by flattened endothelial cells and contain pale, homogeneous eosinophilic fluid. Lymphocytes may be seen within the vascular spaces.

Lymphangioma is differentiated from teratoma with a prominent vascular component by absence of other germ cell elements. Lymphangioma must also be distinguished from haemangioma and an adenomatoid tumour that contain thin walled vessel like spaces.

In contrast to haemangioma, lymphangioma does not contain blood cells in the vascular spaces. The adenomatoid tumour has solid areas and cells lining vessel like spaces, which are not endothelial in nature and stain positive with PAS and alcian blue as well as show positive staining for low molecular weight keratin and not for CD31. To conclude, ovarian lymphangiomas are rare and unusual tumours.

Discussion

1. Ovarian lymphangiomas are extremely rare tumours.
2. Lymphangioma must also be distinguished from haemangioma and an adenomatoid tumour that contain thin walled vessel like spaces by

special stains and immunohistochemistry.

References

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LEPTOSPIROSIS : TROPICAL TO SUBTROPICAL INDIA

Leptospirosis in humans is characterized by an acute febrile illness followed by mild self-limiting sequelae but at times causes more severe, and often fatal, multi-organ involvement. The clinical manifestations are highly variable. In general, the disease presents in four broad clinical categories : (i) a mild, influenza-like illiness : (ii) Weil's syndrome characterized by jaundice, renal failure, haemorrhage and myocarditis with arrhythmias; (iii) meningitis/meningoencephalitis; (iv) pulmonary haemorrhage with respiratory failure.

Presence of leptospira antibody by rapid tests or positive IgM ELISA in patients with clinical illness resembling leptospirosis make the case as probable leptospirosis. PCR detects antigen and is promising on both sensitivity and specificity, but is expensive and is most useful when patients present within four days of fever. Microscopic agglutination test (MAT) is the gold standard serologic test because of its unsurpassed diagnostic specificity. However, MAT is only performed in reference laboratories and requires acute and convalescent samples for diagnostic confirmation. Icterus and thrombocytopenia are markers of severe leptospirosis. Hypotension, myocarditis, cardiopulmonary arrest and adrenal haemorrhages are some of the features of more severe leptospirosis.

Third-generation cephalosporins are effective as well in the treatment of acute disease.

Doxycycline is reported to give some degree of protection to exposed individuals from non-endemic areas. Even if it does not always prevent infection in endemic area, it can reduce the severity of the disease and thus mortality and morbidity.

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