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Lymphangioma of Ovary – A Rare Pathological Entity

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Abstract **Case Report**

Lymphangioma of the ovary is an extremely rare lesion. It was first described in 1908. Lymphangiomas are benign congenital malformations of lymphatic system. They are thought to happen due to obstruction of local lymph flow system and they can occur anywhere in the skin and the mucous membranes. Most common sites are the head and the neck and sometimes they can be found in the intestines, the pancreas and the mesentery. Lymphangioma of the ovary is a very rare tumor. Typically, lymphangiomas are slow-growing tumors that remain asymptomatic for a long time, with the tumor being identified incidentally during histopathological examination after excision.

Keywords: Lymphangioma, ovary, asymptomatic.

INTRODUCTION

Lymphangiomas of the ovary are very rare and are usually encountered as incidental findings often located on the surface of the ovary or in the3,-5 parenchyma. It is uncertain whether lymphangiomas represent true neoplasms, hamartomas or lymphangiectasis. They probably arise from sequestered lymphatic sacs that fail to communicate with the draining lymphatic channels or are secondary to mechanical pressure, trauma, previous surgery or radiation. The common sites include head, neck, axilla and skin [1]. Less than 20 cases of ovarian lymphangiomas have been reported so far in the indexed literature. They are incidentally detected and are usually unilateral but can occasionally be bilateral. Usually seen in adult women, lymphangiomas of ovary can rarely occur in children. The histology differential diagnosis of lymphangioma of the ovary includes the more frequently observed hemangioma, secondary dilatation of lymphatic channels and an adenomatoid tumor. Hemangiomas show vascular spaces lined by continuous endothelial cells and lack lymphocytes in the wall. Secondary causes can be ruled out based on history and absence of stromal reaction. Adenomatoid tumor shows cystic spaces lined by continuous cuboidal or flattened mesothelial cells and do not show lymphocytes in the wall or stroma. The behavior of ovarian lymphangiomas is benign but few cases have recurred and occasional cases have shown transformation to a malignancy [1, 2].

CASE REPORT

A 40 –year – old woman presented with menorrhagia of 1 – year duration. Ultrasonography was done and confirmed as fibroid uterus of small size along with a right sided cystic ovarian mass, measuring 4x4 cms. No significant mass was detected on the left adnexal region. Total abdominal hysterectomy and bilateral salpingooophorectomy was done, Grossly, Uterus and cervix were unremarkable and both tubes were within normal limits. Both ovaries was cystic and measured 5cm in diameter, which on cutting show cystic spaces filled with clear fluid in some and pultaceous material in other small cyst [Figure 1].

Multiple sections were taken from the ovaries.One ovary showed simple subcortical cysts, whereas right ovary showed multiple vascular spaces of varying sizes separated by fibrous tissue septa with occasional lymphoid follicle formation in the stroma (Figure.2). These spaces were lined by flattened endothelial cells and filled with homogenous pale eosinophilic material. A few mature lymphocytes and occasional erythrocytes were also present. The stroma also showed sprinkling of lymphocytes [Figure 3].

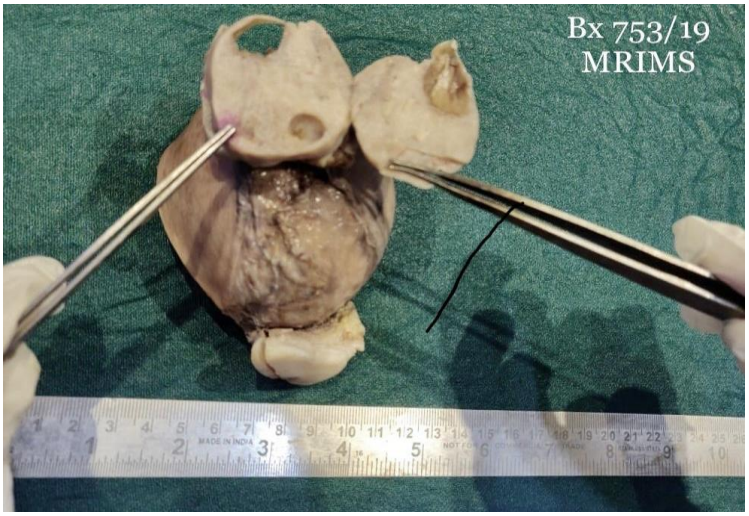

Fig-1: Gross showing cut section of ovary having multiple cysts and grey white areas

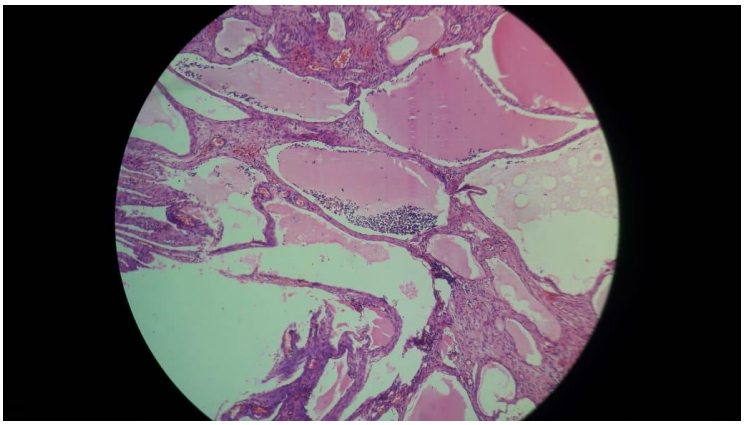

Fig-2: Section showing ovary with multiple vascular spaces of varying sizes separated by fibrous tissue septa filled with lymphocytes and collection of lymphocytes in between the stroma. [H&E,x40]

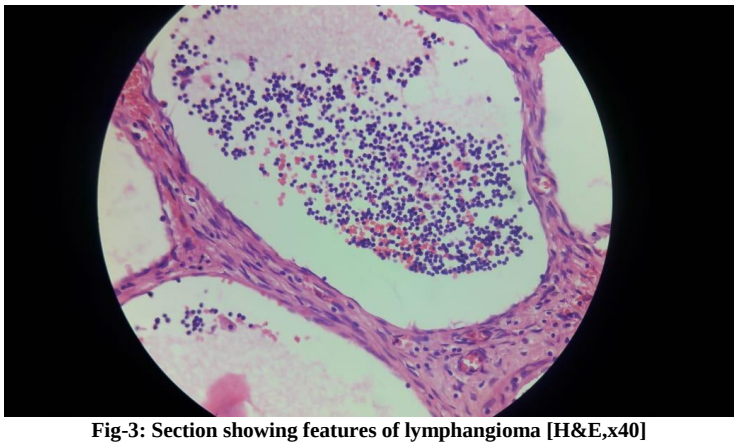

Fig-3: Section showing features of lymphangioma [H&E,x40]

DISCUSSION

Lymphangioma is a benign lesion of the lymphatic vessels that is characterised by lymphatic proliferation. 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. On ultrasound imaging, lymphangioma may appear as septated cystic lesions. 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.

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. Lymphangioma of the ovary is extremely rare.

The lesion is considered benign but a case of lymphangiosarcoma of the ovary has been reported. The lesion is usually unilateral but bilateral lesions have been reported. Earlier, lymphangiomas had been considered to be neoplastic in nature and capable of aggressive behaviour. However, some investigators now believe that lymphangiomas are hamartomas. Others believe that both neoplastic and hamartomatous theories are possible. Fibrosis of lymphatic channels following surgeries or infections leading to dilatation of the proximal channels has been postulated to be the underlying pathogenesis. Lymphangioma of the ovary following radiation therapy has also been reported.

Lymphangiomas are rare malformations, which arise from sequestration of lymphatic tissue which fails to communicate with lymphatic system [1]. Morphologically they are comprised of multiple cystic spaces lined by single layer of flattened endothelium, containing serous or chylous fluid and are classified into capillary, cystic or cavernous type [2, 3]. Lymphangiomas are most commonly seen in head, neck and axillary region but can occur anywhere in body. They can also be seen in intestine, pancreas and mesentery.

Lymphatic system is formed by a network of unidirectional vessels that collects excess fluid from interstitial tissue, which is transported to the regional lymph nodes and ultimately drains to venous system through the thoracic duct. The lymphatic system is present in all organs except in the brain, anterior chamber of the eye, bone marrow and spleen [3].

Pathogenesis of lymphangiomas is uncertain and matter of discussion. Some authors thought they are true neoplasms. According to well established theory, sequestration of lymphatic tissue during embryonic development can cause lymphangiomas. Various authors thought that impaired regional lymphatic drainage due to chronic salphingitis or radiation therapy may be the cause for cystic lymphangioma of ovary.

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. 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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