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Case Report

Giant cystic lymphangioma of the right ovary: A case report and review of literature

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Abstract

Lymphangiomas are rare benign congenital malformations of the lymphatic system. They are thought to happen due to obstruction of the local lymph flow system. Lymphangiomas may arise in any part of the body. Approximately 95% of lymphangiomas are found in the head, neck and axilla, and 5% appear in other parts of the body. Lymphangioma of the ovary especially huge one is exceptional. Lymphangiomas are usually slow growing and patients remain asymptomatic for a long time. We report a case of a very huge lymphangioma of the right ovary associated with lymphangioma of the peritoneum in postmenopausal woman treated by total excision of the both cysts.

***Keywords*:** Lymphangioma, benign congenital tumors, lymphatic system, ovarian cyst.

BACKGROUND

Lymphangiomas are rare tumors of the lymphatic system comprised of multiple cystic spaces lined by endothelium. They are generally benign tumors. Lymphangiomas can be classified as capillary, cystic and cavernous. They contain serous or chylous fluid. (Alqahtani et al., 1999). Lymphangioma of the ovary is rare and there are only few reports in literature. (Hornick and Fletcher, 2005; Fernández et al., 1998; Steyaert et al., 1996; Luo et al., 2004; Scheye et al., 1994; Konen et al., 2002). Lymphangiomas may arise in any part of the body. Clinical manifestations vary from asymptomatic masses to acute abdominal pain. Ultrasonography (US), computed tomography (CT), and magnetic resonance imaging (MRI) are the useful imaging modalities for the diagnosis of lymphangioma. However, lymphangiomas diagnosis becomes difficult because it can be confused with a malignant ovarian mass, especially in postmenopausal woman. They are most often the incidental findings during routine abdominal or pelvic imaging studies.

Case report

A 55 years old postmenopausal woman admitted in surgical department for a huge lymphangioma cysts of right ovary and the peritoneum for 2 years of evolution. Since 20 weeks it becomes very huge causing difficulties for breathing. She was followed-up and treated for diabetes mellitus, asthma and arterial hypertension for last 5 years. There is no history of hemoptysis or melena, or fever, she is obese and weighting 124 kg and the BMI was 48.4 kg/m2. (Photo 1)

General and systemic examination of the patient was characterized by good general conditions with no pallor, no jaundice and no lower limbs oedema. Examination of respiratory and cardiovascular systems revealed no abnormality except wheezing in the both chest and the functional respiratory exploration was within normal limits. Blood was within normal values except blood sugar which was slowly higher. Chest x-ray examination and ECG were without any abnormalities. Ultrasound examination and CT scan showed a huge cystic lymphangioma of the right ovary; the cystic lymphangioma of the peritoneum was not detected (seen) because it was attached to the big ovarian one by thine adherence. After a short preparation, the patient was operated and removal of both cysts was carried out. The first largest Cyst (3.5 liters) was sucked, the second peritoneal Cyst was removed subsequently (Photos 4-5-6-7-8-9). Post-operative period was uneventful. She was discharged after 5 days with a body weight of 111.5 kg. The histological result was in favor of cystic lymphangioma of the right ovary and peritoneum.

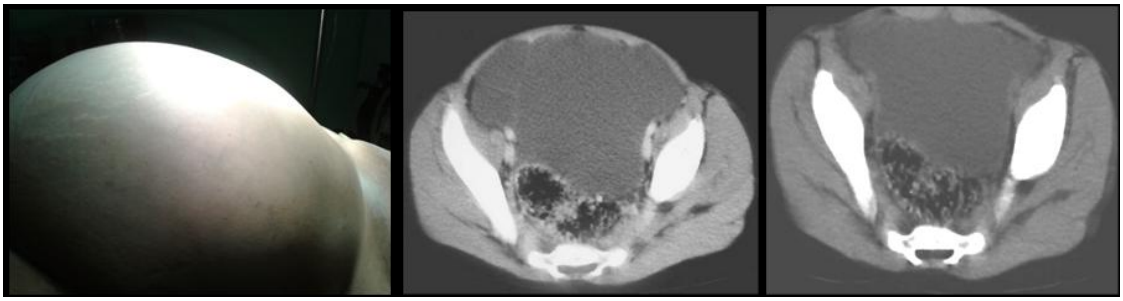
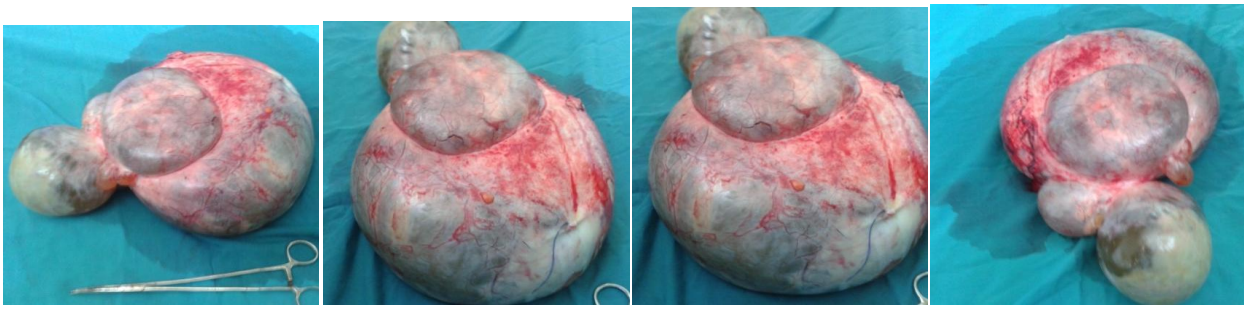
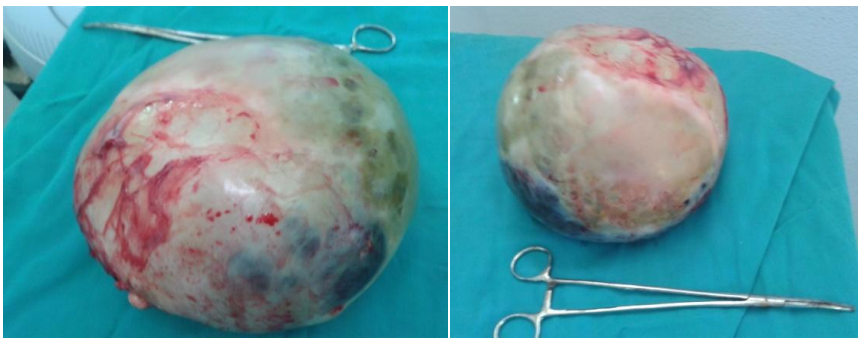


Photo 1. Showed distended abdomen photos 2 and 3: CT scan showing a huge right ovarian cyst



Photos 4-5-6 and 7. Showed the right ovarian lymphangima cyst after having been sucked about 3.5 liters



Photos 8 and 9. Showing the second lymphangioma cyst of the peritoneum.

DISCUSSION

The lymphatic system consists of a network of unidirectional 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 (Kafeel et al., 2010). Cystic cavernous lymphangiomas are uncommon benign and low growing tumor derived from the lymphatic vessels (Alqahtani et al., 1999). Its etiology is remains unclear. Congenital malformation of the lymphatic vessels leading to sequestrations of lymphatic tissue seems to be the cause of this condition (Roisman et al., 1989). Several forms of lymphangiomas, including simple capillary, cavernous and cystic, have been described. The classification of lymphangiomas is mainly based on the size of the lymphatic space and the nature of the lymphatic wall (Chen et al., 2005; Rieker et al., 2000). The cystic form is characterized by a thin irregular wall covered by endothelium, smooth muscle, foam cells, and lymphatic tissue; it appears as a multiseptated, cystic mass with or without intracystic debris and has no connection with the adjacent normal lymphatics system. (Alqahtani et al., 1999; Hornick and Fletcher, 2005; Fernández et al., 1998; Steyaert et al., 1996; Luo et al., 2004; Scheye et al., 1994; Konen et al., 2002; Kafeel et al., 2010; Roisman et al., 1989; Chen et al., 2005; Rieker et al., 2000; Goh et al., 2005). Thin walled lymphatic vessels situated one type. The cavernous type is composed of dilated lymphatic vessels and lymphoid stroma and has connections with normal adjacent

lymphatics (Chen et al., 2005). The size and location of the lymphangiomas are the determinants of the clinical presentation. It is more frequently found in childhood (60% presents at birth, and 90% are detected by the end of the second year of life, and mostly are diagnosed in the first five years of life (Alqahtani et al., 1999; Hornick and Fletcher, 2005; Fernández et al., 1998).Occasionally, the tumor is also discovered in adult in various other anatomic sites (Alqahtani et al., 1999). It most often occurs in the head and neck, axilla, or groin of young children. It is rarely found as intra-abdominal masses when occurring in the abdomen (2-10% in the internal organs) (Hornick and Fletcher, 2005; Fernández et al., 1998; Steyaert et al., 1996; Luo et al., 2004; Scheye et al., 1994; Konen et al., 2002; Kafeel et al., 2010; Roisman et al., 1989; Chen et al., 2005; Rieker et al., 2000; Goh et al., 2005; Losanoff et al., 2003). The clinical manifestations vary from an asymptomatic state to acute onset of severe abdominal pain Griffa et al., 2006; Gabata et al., 2005. Cystic lymphangiomas might arise with acute abdominal pain associated to bowel obstruction, signs of peritonitis, chronic abdominal swelling that is detected by palpation of a cystic mass or abdominal swollenness with lower extremities lymphoedema (Steyaert et al., 1996; Luo et al., 2004; Scheye et al., 1994). The abdominal ultrasonography is the procedure of choice for establishing the diagnosis, even during the antenatal stage (Alqahtani et al., 1999; Hornick and Fletcher, 2005; Fernández et al., 1998; Steyaert et al., 1996; Luo et al., 2004; Scheye et al., 1994; Konen et al., 2002). A cystic lesion with multiple thin septa is the typical Ultrasonography finding of a mesenteric lymphangioma (Chen et al., 2005). Cavernous lymphangiomas appear as solid masses on CT. MRI are more useful than CT for detecting fluid-filled cystic lesions like cavernous lymphangiomas (Rice et al., 1943). The acute lymphangiomas associating intracystic hemorrhage are more difficult to diagnose, CT-scan and celioscopy might be useful approaches in this context (Konen et al., 2002). Sequential ultrasonography and CT-scan examinations showed progressive enlargement of the cystic masses, increase of fluid echogenicity and wall thickening associating multiplication of septa **8**. Lymphangioma of the ovary is extremely rare. It is composed of aggregates of lymphatic spaces in ovarian stroma and the endothelial cells lining these spaces stain positively with CD-31, D2-40 and CD-34 on immunostaining and all were negative for keratin. The main differential diagnosis is an adenomatoid tumour which can be differentiated from the lymphangioma by immunohistochemical studies. The radiology is the revealing diagnostic tool. The lesion is considered benign but a case of lymphangiosarcoma of the ovary has been reported. (Rice et al.,1943). The lesion is usually unilateral but bilateral lesions have been reported. (Evans et al., 1999). 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. (Kurman, 2001). Fibrosis of lymphatic channels following surgeries or infections leading to dilatation of the proximal channels has been postulated to be the underlying pathogenesis. (Evans et al., 1999). The final diagnosis is always based on the histological findings, since this examination shows extensive myofibroblastic areas and objectifies the lymphatic character of the lesion (Fernández et al., 1998; Steyaert et al., 1996; Luo et al., 2004). The main differential diagnosis is an adenomatoid tumor of the ovary. Adenomatoid tumor 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.(Kurman, 2001) (Brunei). Although cystic lymphangiomas are benign tumors, they may compress adjacent structures via a mass effect. Other complications, including rupture, secondary infection, volvulus, or intestinal obstruction, have also been reported (Ricca, 1991; Troum and Solis, 1996). The gold standard treatment modality of cystic lymphangiomas is surgical excision. Treatment with OK-432, steroids, bleomycin, fibrin glue, or Ethibloc has not been established to be superior to surgery (Fernández et al., 1998; Steyaert et al., 1996). However, some authors recommend conservative management of asymptomatic cystic lymphangiomas due to the spontaneous regression rate of 10% (Alqahtani et al., 1999; Hornick and Fletcher, 2005; Fernández et al., 1998; Steyaert et al., 1996). Other treatment modalities including aspiration and injection of sclerosant agents may be performed for emergent decompression; however, these approaches are not recommended for elective therapy due to the high recurrence rates. The treatment is mainly surgical; it consists of enucleation when feasible; the segmental intestinal resection is achieved when the cyst adheres intimately to the bowel (Hornick and Fletcher, 2005; Fernández et al., 1998; Steyaert et al., 1996; Luo et al., 2004; Kafeel et al., 2010). Few reported cases of diffuse malformation required an extensive bowel resection, which might yield short bowel syndrome (Hornick and Fletcher, 2005; Fernández et al., 1998; Steyaert et al., 1996; Luo et al., 2004; Scheye et al., 1994; Konen et al., 2002; Kafeel et al., 2010). The resection could be performed with laparoscopic technique without large incisions. Tumors were cystic masses associated to areas of fat necrosis and hemorrhage. Often, cysts contain thick, gelatinous or milky fluid (Scheye et al., 1994). Laparotomy or laparoscopy both are acceptable routes of surgery for treatment. There are several reports in the literature of successful open resection of intra-abdominal lymphangiomas, but only few such report describing the laparoscopic treatment of an ovarian lymphangioma. (Fernández et al., 1998; Steyaert et al., 1996). It has inherent advantages in the form of less intra-operative

blood loss, early recovery, less morbidity, and low complication rate compared with laparotomy. In our case the choice of laparotomy was dictated by the enormity of the tumor and the non-availability of means technic for the laparoscopy.

CONCLUSION

The cystic Lymphangioma of the ovary is extremely rare benign tumor. The clinical symptomatology is polymorphic and not specific. The diagnosis is suggested by the imaging modalities but still requiring a histopathological confirmation. The treatment of choice is surgical and consists of a full resection of the lesion. The intracystic sclerotherapy could be used for symptomatic tumors. The present case illustrates that lymphangiomas should be included in the differential diagnosis of ovarian cystic masses. Even though, they are being benign in nature majority of the time, wide excision with clear margin and regular follow up is mandatory. The laparoscopic excision is a safe and reliable approach for the treatment.

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