CASE REPORT

Retroperitoneal Lymphangioma with Poly Cystic Ovary Disease: A diagnostic dilemma

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Introduction:

Retroperitoneal cyst is a very rare, benign slowly growing developmental anomaly of lymphatic channel within the retroperitoneal space which commonly (>90%) manifests before the age of 2 years1. Approximately 1/6000 live born is affected with cystic lymphangioma where retroperitoneal space accounts only 5% cases[2]. The incidence of affecting retroperitoneal space is less than 1% of all lymphangiomas. The most frequently involved retro peritoneal cysts are in neck region (75%) & intra abdominal region is affected fewer than 5% in mesentery, Gastro intestinal tract (GIT), pancreas, spleen and liver[3].

The patient may pose no symptoms most often. When cysts are large enough they produce abdominal distension, pain, fatigue, weight loss, hematuria or they can show features of complication (acute abdomen, pain, fever). Cysts are palpable when they are large and it is very difficult to diagnose a retroperitoneal cyst from other differentials by clinical examination. So imaging study is essential for getting some ideas about the space involvement and extension of the swelling and exploratory laparotomy with histopathology is done for confirmatory diagnosis.

Case Study :

A case of 16 years old girl who was normotensive, nondiabetic and nonasthmatic presented with the complaints of pain in the lower abdomen for 15 days and changed bowel habit for last 5 months. The pain was mild, continuous, dull, diffuse, radiating to back and not subsided with analgesic. There was no history of fever, pedal edema, GIT bleeding, decreased urine output. She also complained of constipation for last 5 months. For relief she was regularly taking osmotic laxatives. She had a history of mild form of pain 1 month prior to last attack. Her menstrual abnormalities (dysmenorrhoea and passage of blood clots during menstruation) were also documented.

On examination, she was found obese with abdominal distension and striea. There were no other findings except tenderness in the hypogastrium and right iliac fossa. There was no inguinal lympadenopathy. Digital Rectal Examination findings were normal.

Some basic investigations were done 1st. Haemoglobin-13.8%, Erytrocyte Sedimentation Rate-50mm in 1st hour, White blood cell-8100/cubic mm, Platelet- 229000/cubic mm,

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Serum Electrolytes - within normal limit, SGPT- 21 IU, Serum Creatinine - 0.8 mg/dl. To evaluate differential diagnosis some more investigation were advised. Ultra Sono Graphy (USG) showed - fairly large multisepted cystic lesion measuring about 18.1×8.7 cm in size involving right lower abdomen and right iliac fossa extend beyond the midline upto the left para vertebral region at the level of umbilicus suggesting mesenteric cyst.

Computed Tomography(CT) scan findings were large non enhancing relatively hypodence (density value +22 to +24 HU) mass was seen in the mid abdomen in pre aortic and precaval area apperenly in retroperitoneal space with well defined imperceptible lobulated margin measuring 150×60×140mm. No septa or loculi and calcification were detected. Adjacent bowels were mildly displaced.

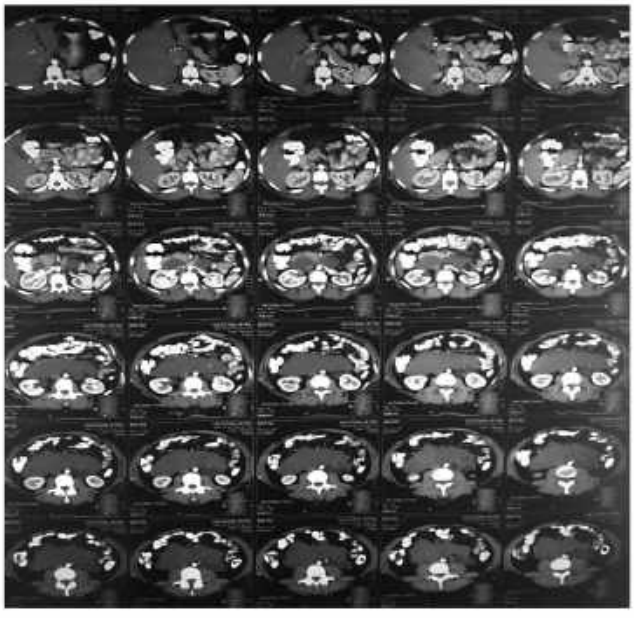

Figure-1: CT scan showing a mass in the retro peritonel space

CT guided FNAC showed scanty cellular material containing lymphocyte and histocyte with clear background.no granuloma and malignant cell involved.

Gynecology department was given consultation for proper management of the patient. Decision for doing exploratory laparotomy was taken in collaboration with them.

Per operative findings: a grape like cystic lesion was found in retroperitoneal para aortic region which was extending up to the mesentery. Opinion was taken from the Gynecology department and PCOD was diagnosed per operatively by the department. Then total excision of the cysts were done. Whole procedure was performed under general anesthesia .

Histopathology report showed fibro fatty tissue with dilated vascular channel (Lymphangioma). The stroma was filled with lymphocyte. No evidence of malignancy was present .

Discussion :

Cystic lymphangioma is a unusual malformation of lymphatic system. They are familiar in children with M:F = 5:2[4]. They can present the symptoms in infancy but 50 to 60% cases by the 1st year of life and 90% by the year 2nd. It develops due to sequestration of lymphatic tissue that fail to communicate normally with the lymphatic system at about 14 to 20 weeks of intrauterine life or from abnormal budding of the lymphatic endothelium[5] or due to abnormal communication between iliac and retroperitoneal lymphatic and venous channel that lead to lymphatic stasis. Other potential cause may be abdominal trauma, localized lymphatic degeneration, radiation, lymphatic obstruction[6].

They are mostly asymptomatic but show symptoms when they are large enough to mimic a spectrum of disease like - intestinal

obstruction, ascitis, bleeding, rupture, ureteric obstruction, haematuria or clotting disorder[7,8]. Complications of retroperitoneal cysts are bleeding, infection, cyst rupture, volvulus, compression to other structures[7,8]. Differential diagnosis are cyst of urothelial and foregut origin, microcystic pancreatic adenoma, retro peritoneal hematoma, abscess, duplication cysts, ovarian cysts, pancreatic pseudocysts and other malignant tumors like undifferentiated sarcoma, cystic mesothelioma, teratoma, lymphangiosarcoma, malignant mesenchymoma, cystic metastasis from ovary and gastric primaries

A contrast x-ray demonstrates water density fluid within multilcular or unilocular cysts. Magnatic Resonance Imaging(MRI) shows delineate additional lesions. USG is used to demonstrate internal structure like septa (unilocular or multilocular) and fluid (clear or chylous) of cysts. CT is essential for excluding differential diagnoses and it give detail information of the space involvement, size, presence of calcification, central scar, extension with attenuation value in the range of fat to water. CT of the cystic lymphangioma shows well circumscribed, encapsulated water dense poly cystic tumor within septa which are hard to differentiate from other cystic lesion. MRI is needed for demonstration of additional lesions, abnormal communication and delineates their boundaries. None of the imaging studies can confirm the diagnosis. It is mainly confirmed by histopathology and immunohistochemistry after laparoscopy or laparotomy. On histological diagnosis lymphangioma shows well circumscribed cystic lesion with or without endothelial lining, a stroma composed of a meshwork of collagen and fibrous tissue and a wall containing focal aggregation of lymphoid tissue.

Treatment of choice is total surgical excision[9] in order to avoid recurrence or complications[10]. Aspiration or sclerosing agents can be used in unresectable cases when involvement of the vital structures have occured[11] laparoscopic procedure is used in some selected cases[12] Hauser et al suggested that isolation and ligation of the cystic lymphangiomas peduncle as well as ligation of lymph channel can prevent recurrence and chylascos[13]. Immunohistochemistry shows positive result of CD31, CD34 and D2-40, factor viii related antigen,endothelial receptor 1,vascular endothelial growth factor-3,prox-1 expression and negative result of cyto keratins in case of lymphatic endothelium[14].

Outcome after complete excision is good. Recurrence is the result of incomplete excision is 50% while in complete excision it is only 7%[15,10].

Conclusion :

Retroperitoneal lymphangioma is a result of misshapenness of the lymphatic channel. It is a challenging and inconclusive case preoperatively by using imaging studies as most of the cystic lesions in retro peritoneal space show same features. Though tough, complete excision of cysts followed by histopathology and immunohistochemistry is compulsory to confirm and cure of the disease. This rare case has a tremendous prognosis after surgery.

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