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Case Report

Retroperitoneal lymphangiectasia: a great clinical masquerade

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ABSTRACT

Retroperitoneal lymphangiectasia is a very rare lymphatic disorder characterized by abnormal proliferation of lymphatics. We present series of 3 cases of retroperitoneal lymphangiectasia which are diagnosed in our institute with the help of Ultrasonography (USG), Computed Tomography (CT) and Magnetic resonance imaging (MRI) of abdomen and pelvis with unusual clinical presentation. We include clinical features and imaging findings of this disorder with its pathogenesis and diagnosis. Two of the cases were clinically masquerading as hernia and one case was mimicking varicocele. Thorough clinical examination and USG, colour Doppler, CT and MRI are extremely helpful imaging investigation that aid in differentiating these lesions from hernia/ varicocele and the cross sectional imaging like CT and MRI can depict the anatomical extent of the disease.

Keywords: Computed Tomography, Contrast enhanced Computed Tomography, Magnetic resonance imaging Retroperitoneal lymphangiectasis, Ultrasonography

INTRODUCTION

Retroperitoneal lymphangiectasia is a rare benign developmental malformation of the lymphatic system characterized by abnormal focal or diffuse dilatation of lymphatics in retroperitoneal tissues, however it can occur anywhere in the body.[1] USG, CT scan and MRI are the commonly used imaging modalities to evaluate intra-abdominal lymphangiectasia. Itappears as intraabdominal cystic low attenuation nonenhancing masses encasing retroperitoneal vessels on computed tomography. The diagnosis is confirmed by aspiration of chylous fluid. Author present series of 3 cases of retroperitoneal lymphangiectasia which are diagnosed in the institute with unusual clinical presentation. Valid consent was taken from these patients.

CASE REPORT

Case 1

A 20-year-old gentleman was referred with history of right inguinal swelling, which he noticed since 4 months. It used to expand on straining. He was an otherwise healthy male with no significant medical history. Physical examination revealed an elastic soft mass without any tenderness in right inguinal region which used to increase in size on prolonged standing and Valsalva manoeuvre

(Figure 1 a, b). This patient also had dilated venous channels along the anteromedial aspect of thigh and hence associated varicose veins were suspected (Figure 1 c). Surgeons referred this patient to USG department with diagnosis of inguinal hernia with varicose veins as they wanted to rule out possibility of intra-abdominal space occupying lesion commonly associated with such clinical presentation.

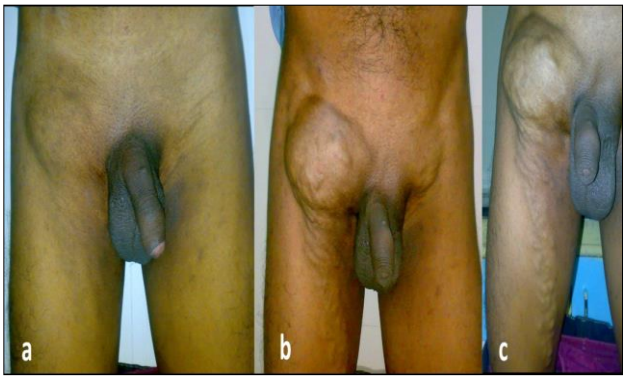

Figure 1: (a) Right inguinal swelling (b) increases in size on Valsalva maneuver (c) associated dilated vascular channels along anteromedial aspect of right thigh.

On USG and Doppler evaluation, there were multiple elongated, tortuous, cystic, anechoic spaces seen bilaterally in retroperitoneum involving paraaortic region, extending anterior to both psoas muscles. On colour Doppler examination, the tortuous spaces showed suspicious flow. There was no evidence of herniation of bowel loops from bilateral inguinal canals.

The tortuous channels were seen extending adjacent to right great saphenous vein and superficial femoral vein, however there was no direct communication seen with deep or superficial venous system of pelvis and thigh. Bilateral Sapheno-femoral junctions were patent. Lower limb venous Doppler evaluation for patency and competency of superficial and deep venous system was within normal limits. On USG and colour Doppler diagnosis of low flow vascular malformation with extension into right thigh was given.

There was no evidence of filarial larvae and abnormal filarial dance was not evident in these dilated cystic spaces. As the diagnosis based on USG and Doppler imaging were contradicting with the surgeon's clinical perspective, contrast enhanced CT (CECT) scan was advised for further confirmation and evaluation of anatomical extent of the disease.

On CECT (Figure 2 c to f), there was large retroperitoneal non enhancing cystic lesion of average attenuation +15to +19 HU seen extending into perirenal and pararenal spaces and encasing aorta, its branches and IVC. The lesion did not show any enhancement even on delayed (7 minutes post- injection) imaging. It also revealed a cystic tubular structure in the posterior mediastinum adjacent to the gastro-esophageal junction and to the right of the aorta. It extended from the mid thoracic level to the level below the diaphragmatic crura. Inferiorly the retroperitoneal lesion was extending into pelvis in perivesical as well perirectal region upto the pelvic diaphragm.

The lesion was encasing both the ureters laterally but there was no hydronephrosis seen. On right side the lesion was creeping into the right inguinal region along right iliac vessels through inguinal canal and it followed the course of GSV for short segment in upper one third of thigh. On CECT the diagnosis of retroperitoneal low flow vascular malformation i.e. retroperitoneal lymphangiectasia with dilatation of cisterna chyli and right thigh extension was kept.

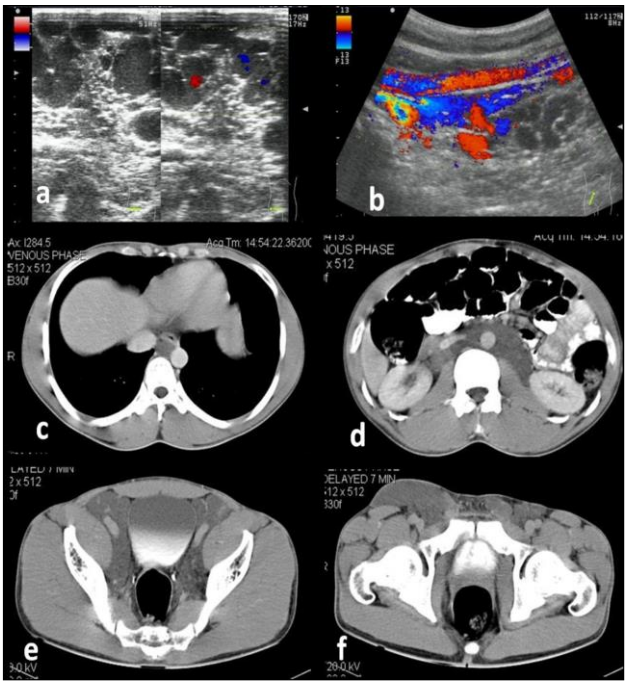

Fig. 2: (a) USG abdomen axial images in right iliac fossa region showing elongated, tortuous, cystic, anechoic spaces seen bilaterally in retroperitoneum. (b) On colour Doppler examination, the tortuous spaces showed suspicious flow seen along right common iliac vessels in oblique saggital image. (c) CT scan of abdomen and pelvis in venous phase, axial image shows dilated cisterna chyli in section at diaphragmatic level, (d) nonenhancing retroperitoneal cystic lesion in paraaortic region, (e) cystic lesions insinuates along both iliac vessels, (f) lesion in right inguinal region.

MRI of abdomen and pelvis was also performed and the findings were confirmed with better soft tissue contrast (Figure 3 a, b, c). The lesion was hyperintense on T2 and short tau inversion recovery (STIR) and hypointense on T1 with extent and morphology similar to CT scan findings.

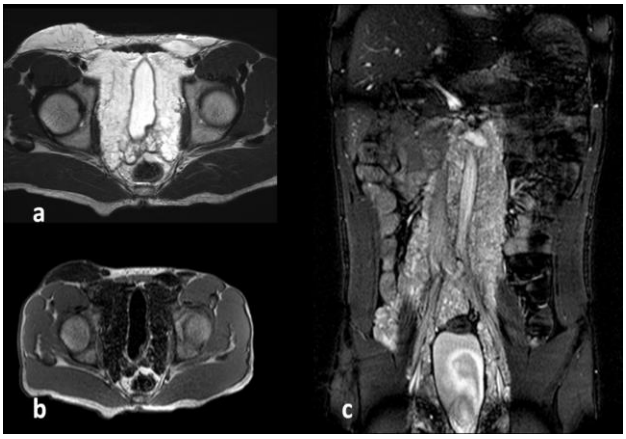

Figure 3: (a) MRI of abdomen and pelvis in axial reconstruction showing T2 hyperintense, (b) T1 hypointense, (c) lesion in pelvis in perivesical region and in right inguinal region. Short TI Inversion Recovery (STIR) coronal image showing hyperintense cystic lesion in retroperitoneum.

Case 2

A 22-year-old gentleman presented with swelling in scrotum since 6 months with a sudden onset excruciating pain 5 days back. The pain was partially relieved after medications. The swelling was progressively increasing in size and used to become prominent on standing and decreases on lying down (Figure 4 a marked with arrow). There was no other significant medical or surgical history. On clinical examination in standing position, there was increase in size of swelling on Valsalva maneuver. On palpation, a large mass was felt in scrotum with few dilated channels. There was no erythema or warmth noted over the swelling. A provisional clinical diagnosis of varicocele was made.

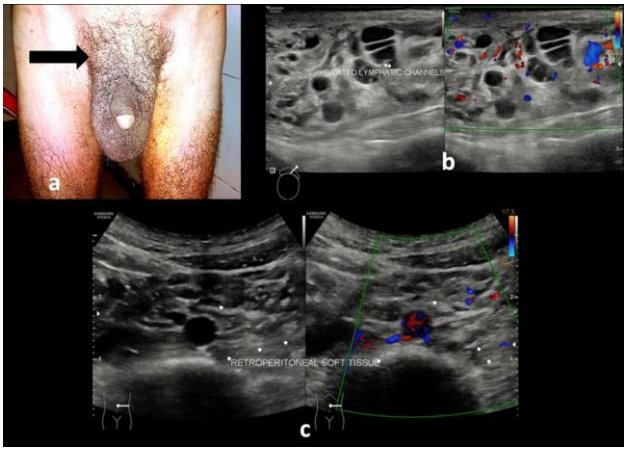

Fig. 4: (a) Swelling at scrotal root (marked by black arrow). (b) USG and Doppler axial images of abdomen showing multiple dilated and tortuous channels in scrotum with no internal vascularity, (c) similar channels are also seen in retroperitoneum.

On USG and Doppler examination (Figure 4 b and c), there were multiple dilated and tortuous channels noted in scrotum, few of them were showing suspicious venous colour flow. However, most of the dilated channels in scrotum showed no internal vascularity. Mild scrotal wall edema with bilateral mild hydrocele was noted. Bilateral testis and epididymis was unremarkable. Patient was advised CECT abdomen and scrotum for further evaluation.

On CECT (Figure 5 a to d), There was ill defined hypodense non-enhancing lesion of average HU 20-22 seen involving retroperitoneal region extending from D11 to L4 vertebral level. It was encasing aorta its branches as well as inferior vena cava and its tributaries without causing mass effect on it. It was also seen to abut adjacent structures. Multiple dilated and tortuous cystic channels were also seen in bilateral spermatic cord with no postcontrast enhancement.

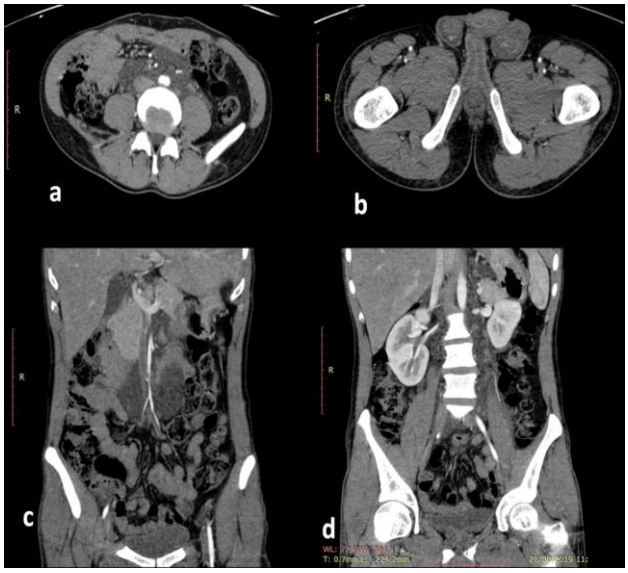

Figure 5: (a) CT scan of abdomen and pelvis in venous phase, axial reconstruction shows non-enhancing retroperitoneal cystic lesion in pre and paraaortic region, (b) bilateral inguinal region (c, d) coronal images showing lesion in retroperitoneum.

On MRI (Figure 6 a to d), There were multiple cystic lesions seen involving retroperitonium encasing aorta and inferior vena cava. They were predominantly hypointense on T1, hyperintense on T2. No postcontrast enhancement noted. Similar morphology dilated cystic channels were noted in bilateral spermatic cords. Few of the cystic lesions were T1 hyperintense suggestive of intracystic haemorrhage.

Thus, diagnosis of retroperitoneal lymphangiectasia with scrotal extension and secondary complication of haemorrhage within dilated lymphatic channels was made.

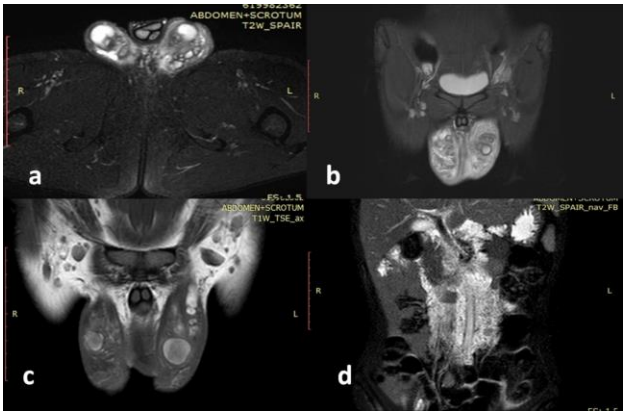

Fig. 6: (a) MRI of abdomen and pelvis showing multiple cystic lesions in scrotum appearing hyperintense on axial T2/SPAIR (spectral attenuated inversion recovery) image. (b) Coronal STIR image showing hyperintense lesion in scrotal neck. (c) few of the cystic spaces show haemorrhage in the form of T1 hyperintensity within in coronal T1 weighted image. (d)T2/SPAIR coronal image showing lesion in retroperitoneum.

Case 3

A 25-year-old gentleman was referred to our tertiary care hospital from a secondary community health centre where he was operated for bilateral inguinal swelling present since 3 months (Figure 7 a). There was no evidence of other significant medical history. Physical examination revealed an elastic soft mass without any tenderness in right inguinal region which used to increase in size on prolonged standing and Valsalva manoeuvre. Diagnosis of uncomplicated bilateral inguinal hernia was made.

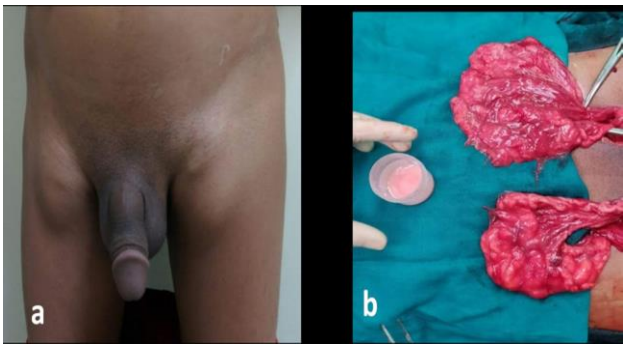

Figure 7: (a) Clinical picture showing bilateral inguinal swelling. (b) Intraoperative image showing multiple dilated cystic structures along bilateral spermatic cord with aspirated chylous fluid.

Intraoperatively, there were multiple dilated cystic structures were noted along bilateral spermatic cord. Chylous fluid was aspirated from the cysts (Figure 7 b). Patient was referred for further management and was advised CECT and MRI for further extend of disease.

On CECT (Figure 8 a and b), an ill-defined hypodense non-enhancing lesion of fluid density seen involving retroperitoneal region encasing aorta, bilateral renal arteries as well as inferior vena cava and its tributaries.

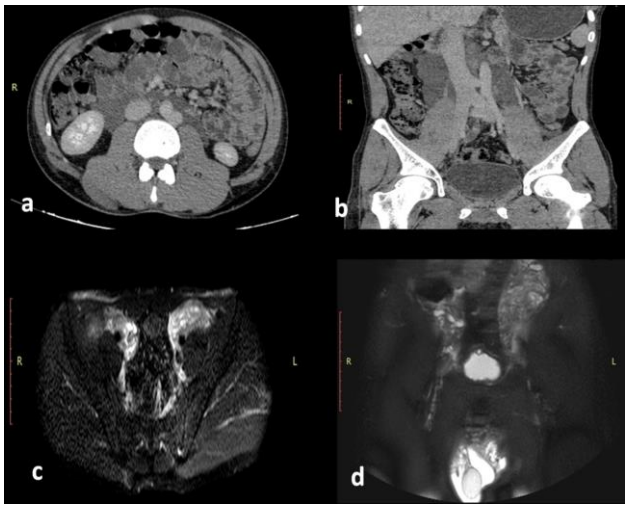

Figure 8: (a) CT scan of abdomen and pelvis in venous phase, shows nonenhancing retroperitoneal cystic lesion in pre and paraaortic region in axial (b) and coronal reconstruction. (c) MRI of abdomen and pelvis in axial reconstruction showing T2/STIR hyperintense multiple dilated cystic lesions in pelvis (d) in axial reconstruction and in retroperitoneum and scrotum in coronal reconstruction.

On MRI (Figure 8 c and d), there were multiple cystic areas seen involving retroperitoneum encasing aorta and inferior vena cava. It appears hypointense on T1, hyperintense on T2. No postcontrast enhancement noted. Thus, diagnosis of retroperitoneal lymphangiectasia was made on the basis of imaging findings.

DISCUSSION

Lymphangiectasia is a type of lymphatic malformation which is classified under low flow vascular malformation. Mulliken and Glowacki described a helpful classification scheme for vascular anomalies. They have classified vascular anomalies as either vascular tumor with endothelial hyperplasia or vascular malformations secondary to an error of embryonic development with normal endothelial turnover. This classification scheme is supported by clinical, histologic, histochemical, and biochemical differences as well as imaging features. Vascular malformations are also classified as slow-flow malformations (capillary, venous, lymphatic, capillary-venous, and capillary-lymphatic-venous malformations) and high-flow malformations (arteriovenous fistulas, arteriovenous malformations).[2]

A classification scheme for vascular anomalies based on cellular features, flow characteristics and clinical behavior was updated during the 1992 meeting of the

International Society for the Study of Vascular Anomalies (ISSVA).[3]

This classification divides vascular anomalies into 2 categories: tumours and vascular malformations. The etiology of tumors (e.g. hemangiomas) is one of endothelial cell proliferation, histologically showing endothelial hyperplasia and more mast cells during proliferative phase.

On the other hand, vascular malformations are developmental errors resulting in collections of abnormal vessels with normal endothelium and normal mast cell count. The capillary venous malformation (CVM) composed of arterial malformations are called high flow venous malformation (HFVM). In contrast, two thirds of CVM are predominantly venous, and a quarter of these lesions is completely or partly of lymphatic origin (termed low flow venous malformation- LFVM). Most of LFVM occur in the region of the head or neck, and those in the retroperitoneal region are rare. It has been reported that retroperitoneal lesions account for approximately 1% of all lymphatic malformations.[4,5]

Because most vascular anomalies occur in the head or neck region, which can be observed percutaneously, clinical behavior plays a large role in preoperative diagnosis. However, retroperitoneal lesions, being deeper, are not accessible for clinical examination and it is difficult to evaluate and differentiate them by their clinical behavior. Thus, imaging evaluation is significant in preoperative diagnosis, especially in retroperitoneal vascular anomalies. Moreover, the imaging characteristics in vascular malformations are extremely variable, relying upon its vessel components and flow characteristics.

According to the literature, USG and MRI are useful in evaluation of vascular lesions.[4] Ultrasonography is the initial imaging procedure, but its inability to evaluate the entire extent of large, deep lesions may be a disadvantage. On grey scale imaging heterogeneous lesion with anechoic channels can be seen in 50% of cases. Hyperechoic foci giving strong posterior acoustic shadowing can be seen in less than 20% of cases. Doppler USG plays valuable role as it helps to evaluate vascularity and flow pattern in the lesion. In most cases, Doppler USG demonstrates monophasic, low-velocity flow in 20%. At times, no flow is demonstrable. This absence of flow may indicate thrombosis or limitation of the equipment.[5]

MRI is extremely helpful to evaluate the extent and flow dynamics of the lesion. Because of excellent soft tissue contrast of MRI, the relationship of the lesion with adjacent soft tissue is seen nicely. LFVM are usually iso to hypointense on T1W images and hyperintense on T2W images.[6,7]

Lymphatic malformation or lymphangiectasia unlike other vascular malformations demonstrates insignificant mass effect and tends to insinuate along anatomical planes. three cases have been described of retroperitoneal lymphangiectsia in which no palpable mass was seen per abdomen despite of extensive involvement of retroperitoneal and extraperitoneal pelvic spaces. Moreover, the inguinal and scrotal extension was seen as the lymphangiectasia insinuates along the femoral vessels and spermatic cord respectively. Inguinal or scrotal swelling which increases in size on valsalva, on cough impulse and prolonged standing showed clinical behavior similar to inguinal hernia or scrotal varicocele, thus misleading the clinicians towards the common surgical diagnoses of hernia/hydrocele. Retroperitoneal lymphangiectasia thus proves to be a great clinical masquerade. Thorough and careful clinical examination with evaluation of surgical masses by clinical tests like transillumination would have helped to avoid unindicated surgical exploration as occurred in case 3.[8] Inguinal lesions of retroperitoneal lymphangiectasia are brilliantly trans illuminant and help us distinguish from hernia. Exception to this clinical test could be lymphangiectasia with secondary complication like dense internal haemorrhage. Surgeons when in doubt can always take the aid of imaging investigations.

CONCLUSION

Retroperitoneal lymphangiectasia is a type of lymphatic malformation with clinical behavior similar to inguinal hernia/ varicocele due to its characteristic features of insinuating along anatomical planes without causing mass effect. A careful clinical examination with mindful application of clinical tests like transillumination would help differentiate it from inguinal hernia. Ultrasonography and colour Doppler, CT and MRI are extremely helpful imaging investigation which aid in differentiating these lesions from hernia/ vericocele and the cross sectional imaging like CT and MRI can depict the anatomical extent of the disease.

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Ethical approval: Not Required

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