



Lymphatic Leaks: Decoding Retroperitoneal and Inguinoscrotal Lymphangiectasia through Imaging

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Citation this Article: Dr. Esha Saruparia, Dr. Pratik Patil, Dr. Devanshi Shah, “Lymphatic Leaks: Decoding Retroperitoneal and Inguinoscrotal Lymphangiectasia through Imaging”, IJMSIR - April – 2025, Vol – 10, Issue - 2, P. No. 147 – 150.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Retroperitoneal lymphangiectasia is a very rare benign lymphatic malformation characterized by abnormal enlargement of lymphatics. Lymphatic malformation in the inguinoscrotal region is rare, occurring in about 0.2% of patients with groin and scrotal swellings.¹

USG and CT scan are the commonly used imaging modalities to diagnose, evaluate and see extent of intra-abdominal lymphangiectasia. It appears as multilocular cystic non enhancing lesion. They are trans fascial and involve multiple compartments. The diagnosis is confirmed by aspiration of chylous fluid.²⁻⁷

We present a case of a 25-year-old male with a retroperitoneal lymphatic malformation that was causing right scrotal swelling, which was diagnosed as retroperitoneal low flow vascular malformation i.e. retroperitoneal lymphangiectasia and right inguinoscrotal lymphangiectasia by laboratory tests and Ultrasonography and CECT.

Retroperitoneal and inguinoscrotal lymphangiectasia is a type of slow flow lymphatic malformation with clinical presentation of scrotal swelling. Ultrasonography and cross-sectional imaging are helpful in diagnosis and differentiation of these lesions from hernia/ varicocele and filariasis. Further CT and MRI help with the anatomical extent of the lesion.¹

Keywords: Retroperitoneal lymphangiectasis, lymphatic malformation, inguinoscrotal region.

Introduction

Retroperitoneal lymphangiectasia is a very rare benign lymphatic malformation characterized by abnormal enlargement of lymphatics. Lymphatic malformation most commonly occurs in the head and neck, but it can involve any area of the body. Lymphatic malformation in the inguinoscrotal region is rare, occurring in about 0.2% of patients with groin and scrotal swellings.¹

USG, CT scan and MRI are the commonly used imaging modalities to diagnose, evaluate and see extent of intra-

abdominal lymphangiectasia. It appears as multilocular cystic non enhancing lesion. They are trans fascial and involve multiple compartments. The diagnosis is confirmed by aspiration of chylous fluid.²⁻⁷

Presenting a case of a 20-year-old male with a retroperitoneal lymphatic malformation that was causing right scrotal swelling, which was diagnosed as retroperitoneal low flow vascular malformation i.e. retroperitoneal lymphangiectasia and right inguinoscrotal lymphangiectasia by laboratory tests, Ultrasonography and CECT.

Case Report

A 25 year old man was referred with history of trauma to abdomen. Patient also had scrotal swelling on right side since birth. No other significant medical history. Emergency laparoscopy with ileostomy was done in view of transverse colon perforation. Three weeks after surgery, stoma closure was planned hence patient was advised USG.

Ultrasonography: Multiple tortuous, cystic, anechoic spaces seen in the right inguino-scrotal region. Bilateral testes appear normal. On doppler study, the tortuous spaces showed no flow. There was no evidence of herniation of bowel loops from inguinal canal or moving linear echoes within for filarial larvae and abnormal filarial dance.

On CECT, non enhancing cystic lesion of average attenuation +15 seen in the retroperitoneum surrounding the aorta, inferior vena cava extending from mid pole of bilateral kidneys further inferiorly along the right common iliac and external iliac vessels with extension into the right side of scrotum. It was causing displacement of the right testis anteriorly. No significant mass effect was seen.

On CECT the diagnosis of retroperitoneal slow flow vascular malformation i.e. retroperitoneal

lymphangiectasia and right inguino-scrotal lymphangiectasia was made.

Further intra operative during stoma closure, lesion was aspirated and confirmation of chylous fluid on laboratory tests was made.

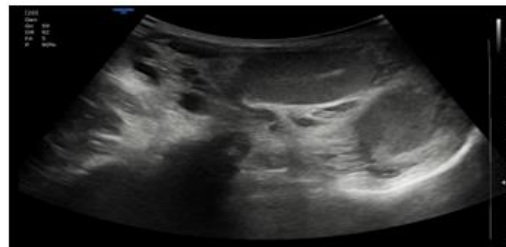


Figure 1&2: Multiple tortuous, cystic, anechoic spaces in right inguinoscrotal region. Normal Bilateral testes

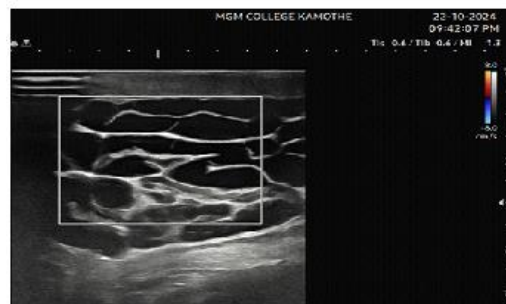
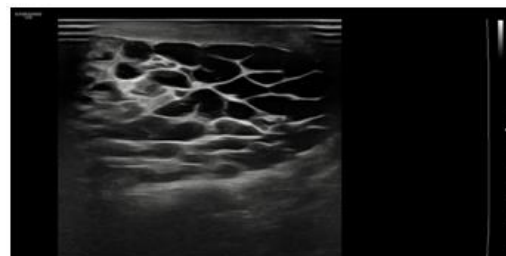


Figure 3&4: Multiple tortuous, cystic, anechoic spaces with no internal echoes or vascularity on Doppler study

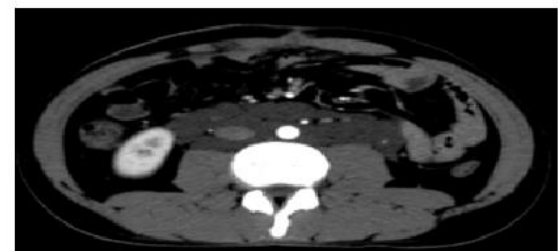


Figure 5&6: CECT A+P Axial cuts at infrarenal and scrotal level: Non enhancing cystic lesion in retroperitoneum surrounding Aorta & IVC extending into right inguinoscrotal region with no mass effect.

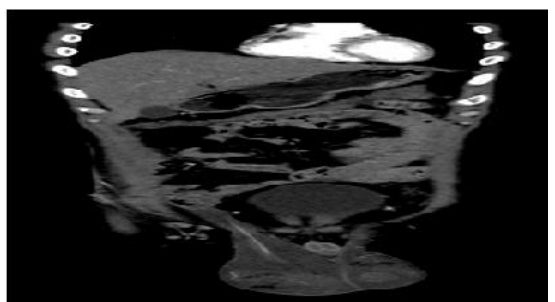
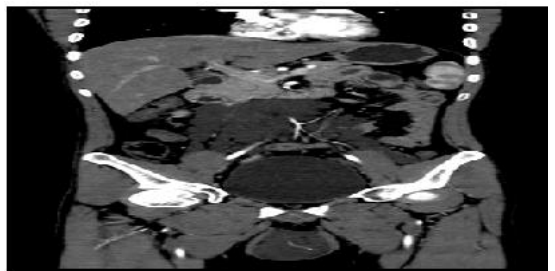


Figure 7&8: CECT A+P Coronal cuts: Non enhancing cystic lesion in retroperitoneum extending into right inguinoscrotal region along the right common and external iliac vessels with no mass effect.

Discussion

International Society for the Study of Vascular Anomalies (ISSVA) classification divides vascular

anomalies into 2 categories: tumors and vascular malformations.⁸⁻¹⁶

The vascular malformations can be simply capillary, venous, lymphatic malformations, arterio-venous malformation, arterio-venous fistula or combined. Lymphangiectasia is a type of lymphatic malformation classified under slow flow vascular malformation mostly occurring 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.⁸⁻¹⁶

Head or neck region vascular anomalies can be observed superficially and clinically. However, retroperitoneal lesions are deeper therefore incidental and are difficult to evaluate and differentiate without imaging. Lymphatic malformation or lymphangiectasia unlike other lesions demonstrates insignificant mass effect and tends to insinuate along anatomical planes. USG and cross-sectional imaging are useful in evaluation of retroperitoneal vascular lesions.⁸⁻¹⁶

Ultrasonography is the initial imaging but is not able to evaluate the entire extent of large, deep lesions. Multiple cystic, anechoic channels can be seen in majority of the 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 or no flow.

CECT and MRI are helpful to evaluate the extent of the lesion and relationship of the lesion with adjacent soft tissue. It appears as multilocular non enhancing cystic lesion with trans fascial multi-compartment involvement. On MRI, they are iso to hypointense on T1W and hyperintense on T2W images. Some show thin septations which are better characterized with ultrasound or T2W MR images. Enhancement of these septations may also be seen in T1 post contrast images.

Conclusion

Retroperitoneal and inguinoscrotal lymphangiectasia is a type of slow flow lymphatic malformation presenting as scrotal swelling. Ultrasonography helps in diagnosis and differentiation of these lesions from hernia/ varicocele and filariasis. Further CT helps with the anatomical extent of the lesion.

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