Clinical challenges of scrotal lymphangioma in an adult: a rare case of scrotal swelling

Justin Loloi, MD,[1] Mustufa Babar, BS,[2] Umair Azhar, BS,[2] Jinrong Cheng, MD,[3] Amanda North, MD[1,2]

[1]Department of Urology, Montefiore Medical Center, Bronx, New York, USA
[2]Albert Einstein College of Medicine, Bronx, New York, USA
[3]Department of Pathology, Montefiore Medical Center, Bronx, New York, USA

LOLOI J, BABAR M, AZHAR U, CHENG J, NORTH A. Clinical challenges of scrotal lymphangioma in an adult: a rare case of scrotal swelling. *Can J Urol* 2022;29(4):11262-11265.

Scrotal lymphangiomas represent an extremely rare cause of scrotal swelling. We report a case of scrotal lymphangioma in an 18-year-old male who presented with painful scrotal swelling. Scrotal ultrasound revealed a complex multicystic structure in the left hemiscrotum. The patient underwent successful surgical excision of the mass. Postoperatively, he developed a hydrocele which eventually spontaneously regressed. Histopathology confirmed the diagnosis. We outline the unusual presentation, characteristic imaging and histology findings, and surgical management of scrotal lymphangiomas. With this information, urologists may exercise a heightened level of awareness for this rare cause of scrotal swelling.

Key Words: scrotum, lymphangioma, scrotal swelling, excision

Introduction

Lymphangiomas are lymphatic hamartomas that result from inadequate lymphatic drainage with subsequent pooling of lymph in a specific area of the body.[1] These benign growths are typically congenital, with half occurring at birth and 90% being evident by the age of two. Acquired lymphangiomas are less common and often occur through the obstruction of lymphatics due to inflammation, trauma, or degeneration.[2] The majority (95%) of lymphangiomas occur either in the neck or axilla.[1,3] Less common areas of occurrence are in the retroperitoneum, intraperitoneum, gluteal region, mediastinum, groin, mesentery, omentum, spleen, and the scrotum.[4,5] Scrotal lymphangioma represents an exceedingly rare cause of scrotal swelling. Thus, we report a case of scrotal lymphangioma in an 18-year-old male who presented with scrotal pain/swelling and subsequently discuss the clinical challenges associated with the diagnosis and treatment of this rare entity.

Case report

An 18-year-old male presented to the Emergency Department with a 1 day history of acute left-sided scrotal swelling and pain. He denied similar episodes of scrotal pain or swelling in the past. The patient had an upper respiratory infection 2 weeks ago and denied any recent history of trauma. Physical exam revealed bilateral descended testes and a lobular, palpable soft tissue mass independent of the spermatic cord in the left hemi-scrotum without tenderness to palpation. Laboratory studies revealed a white blood cell count of 7.7 k/uL, hemoglobin of 13.8 g/dL, hematocrit of 43.7% and creatinine 0.82.

Accepted for publication June 2022

Address correspondence to Dr. Amanda North, Department of Urology, Montefiore Medical Center, Albert Einstein College of Medicine, 1250 Waters Place, Bronx, NY10461 USA

A scrotal ultrasound (US) showed a right inguinal reducible hernia containing fat and a left inguinal nonreducible scrotal hernia containing fat and suspected bowel. A computed tomography (CT) scan of the abdomen and pelvis revealed a 1.6 cm x 2.5 cm x 3.8 cm extra-testicular elongated septate cystic lesion located superior and anterior to the left testes, Figure 1a, there was no hernia located within the inguinal region. The differential diagnosis for the scrotal mass included an extratesticular hematocele, extratesticular pyocele or a tumor. He was discharged from the Emergency Department with close outpatient Urology follow up.

The patient was evaluated in Urology clinic. The sonographic images were reviewed with the Radiology team, who described the lesion as potentially representing a lymphangioma given the presence of thin-walled cystic masses. To rule out an infectious pathology, he was treated with and completed 2 weeks of Levofloxacin. Two weeks later, repeat scrotal US showed a complex multicystic structure in the left hemiscrotum which consisted of multiple fluid-filled locules, some with low level debris and other with avascular solid appearing tissue, and intervening septations of varying thickness, Figure 1b. The imaging findings were thought to represent a lymphatic malformation versus hematocele or pyocele.

In light of the uncertainty of the diagnosis and because the patient was experiencing pain secondary to the mass, he was taken to the operating room for a left scrotal mass excision, left inguinal hernia repair, and orchiopexy. Examination under anesthesia revealed a large, lobulated scrotal mass that was separate from the testicle. An inguinal incision was performed given the concomitant inguinal hernia and the uncertainty of the etiology of the mass. Despite extensive exploration, it was difficult to deliver the mass and testicle into the field. Therefore, a counter-incision was made in the scrotum. There, the left testicle was delivered. A large, multiloculated, cystic-appearing structure was encounter superior to the testicle and adjacent to the spermatic cord. A hydrocele sac was encountered and punctured, expelling approximately 40 cc of brown hydrocele fluid which was sent for culture (grew normal skin flora). The mass was carefully dissected from its scrotal attachments and excised in its entirety. It measured up to 5 cm. The left testicle was healthy-appearing and was subsequently fixated to the dartos using a 1-point fixation via 4-0 Mersilene suture. During the scrotal portion of the mass removal, the gubernaculum was released and the testicle appeared to have increased mobility. Thus, we proceeded with a single point fixation of the testicle rather than the standard 2 or 3-point fixation orchidopexy as the patient's testicle was already in the scrotum and there was no concern for cryptorchidism or testicular torsion. The patient tolerated the surgery well and was discharged the same day.

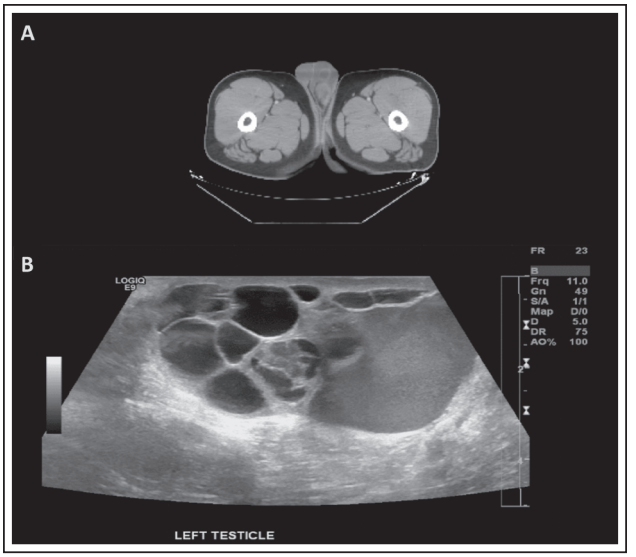

Figure 1. CT and Scrotal US prior to surgery. **(A)** CT imaging revealed a cystic mass. **(B)** Scrotal US revealed a complex multicystic structure.

At 2 weeks postoperatively, the patient reported worsening left scrotal pain, swelling, and drainage from the scrotal incision. A repeat scrotal US showed a large left hydrocele. He was seen in clinic, where the hydrocele was drained with manual pressure, eliciting approximately 100 mL of yellowish-brown fluid without purulence. The patient reported immediate improvement in his symptoms. He was prescribed a course of antibiotics. He continues to feel well with minimal scrotal discomfort and without scrotal drainage at 2 month follow up.

Gross examination of the specimen showed a 5.6 cm x 4.0 cm x 2.5 cm pink-brown and irregular disrupted cystic mass, Figure 2. The solid component was composed of white-pink and focally hemorrhagic fibrous tissue. The cysts ranged from 0.2 cm to 5.5 cm in greatest dimension and contained partially clear serous fluid and partially yellow mucinous material. The cystic component occupied approximately 80% of the total cut surface. The internal cyst wall was tan-pink and smooth and measured up to 0.2 cm in maximum thickness.

Histology of the resection showed the soft tissue lesion consists of multiple dilated cavernous vascular spaces with endothelial lining. Many of the dilated lumina contained abundant eosinophilic proteinaceous debris, and scattered lymphocytes. Those morphologic

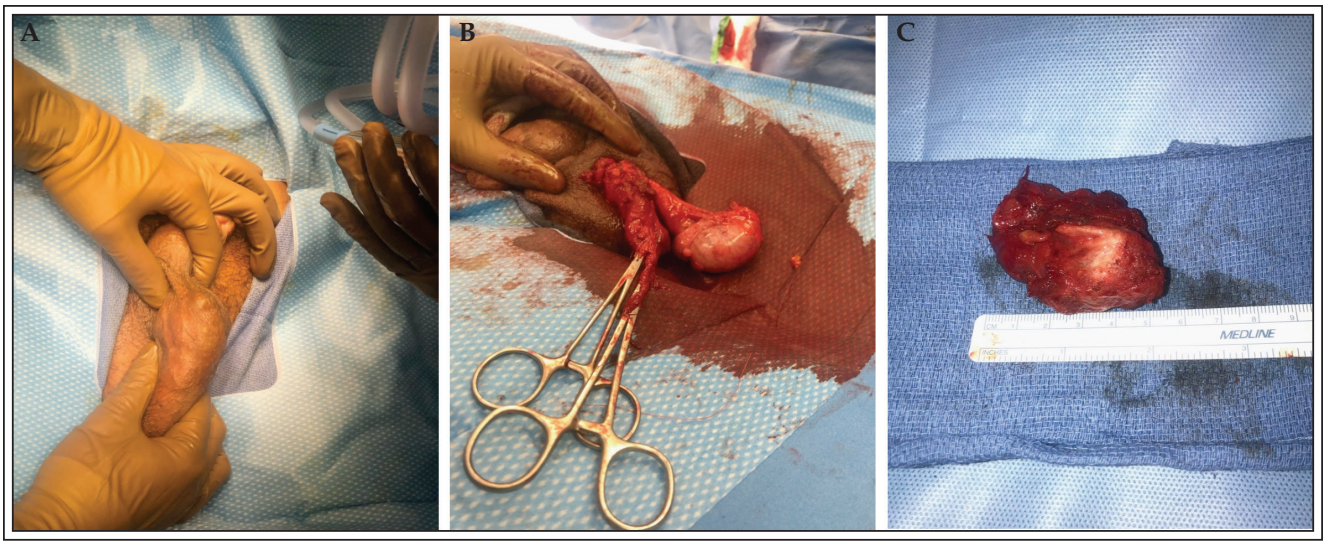

Figure 2. Gross images of the mass. **(A)** Preoperative. **(B)** Intraoperative. **(C)** Postoperative.

features of the soft tissue lesion were characteristic of a lymphangioma. Immunohistochemical evaluation showed the endothelial lining is positive for D2-40 (podoplanin), a specific lymphatic marker and CD34, a vascular marker, Figure 3. The immunoprofile of the lesion was consistent with a lymphangioma.

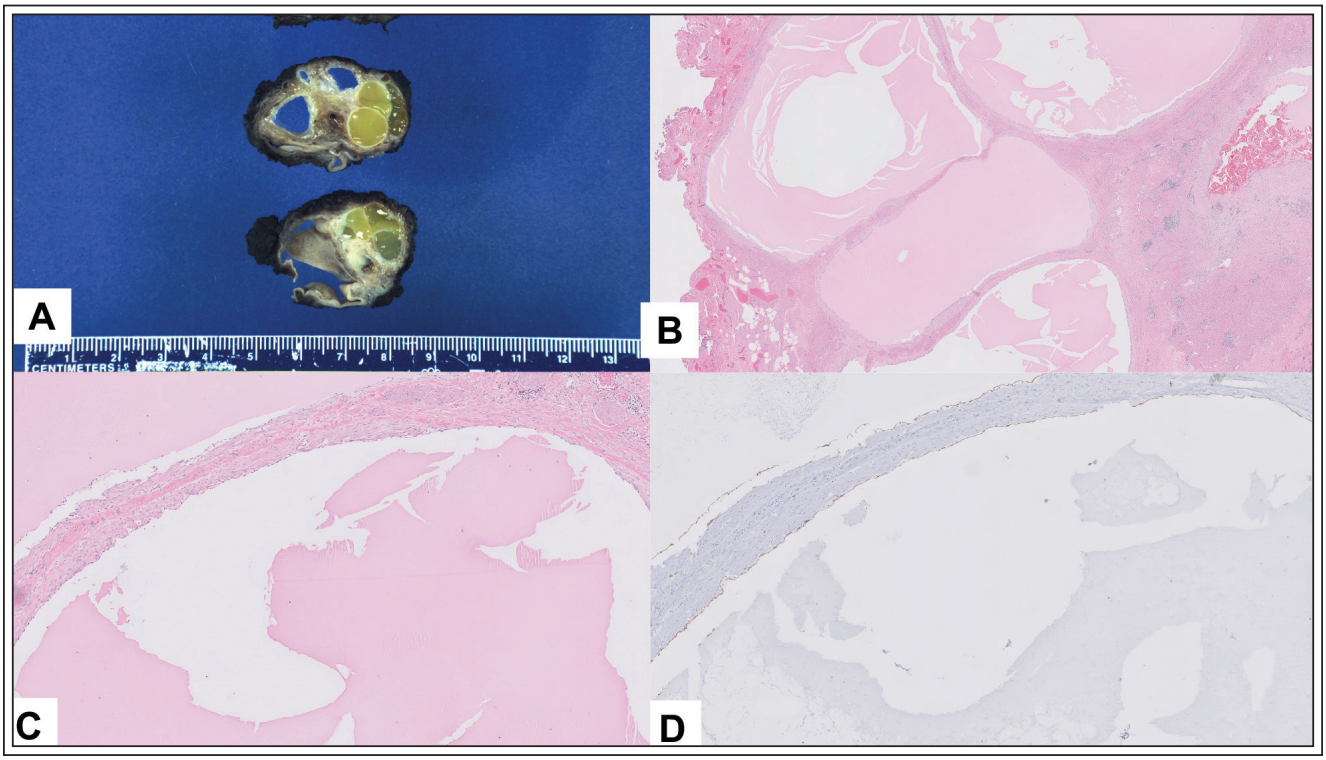

Figure 3. (A) Gross images show multiple cysts on the cut surfaces of the resection specimen. And the cysts contain clear serous fluid or yellow mucinous material. **(B)** and **(C)** Histology shows multiple dilated cavernous vascular spaces, lined by a single layer of small bland-appearing endothelial cells. And many of the vascular spaces contain proteinaceous fluid with scattered lymphocytes. **(D)** Immunostain shows the endothelial cells are positive for D2-40.

Discussion

Scrotal lymphangiomas often present as painless scrotal swelling. The presence of acute pain, as with our case, with the scrotal swelling may be attributed to hemorrhage or excessive production of lymph.[6] As with all extratesticular masses, scrotal lymphangiomas must classified as either solid or cystic through either US or CT imaging, with US often serving as the first-line imaging modality for all suspicious scrotal masses.[7] In our patient, an US was initially carried out. Given the concern for an inguinal hernia with potential bowel involvement and to better delineate the scrotal structure, a CT was subsequently performed. Imaging findings of scrotal lymphangiomas have been described as lobulated, multiseptated, multicystic, or multilocular masses.[8]

Due to the rare occurrence of a lymphangioma in the scrotal region, scrotal lymphangiomas are often misdiagnosed for hernias, varicoceles, epididymal cysts, acute scrotal cysts, or hydroceles.[6] In fact, Hurwitz et al[6] reported seven cases of scrotal lymphangiomas, all of which were misdiagnosed preoperatively. Consequently, this led to improper surgical approaches, involving incomplete excisions, and therefore recurrence in four of the seven cases.[6] Therefore, although extratesticular masses are often benign and do not require complete excision, scrotal lymphangiomas are an exception given the risk of recurrence with an incomplete excision. This further emphasizes the importance of considering scrotal lymphangiomas on the differential diagnosis prior to surgical treatment.

Through complete surgical excision, the postoperative course of scrotal lymphangiomas is often uneventful. However, our case showed the occurrence of a presumed reactive hydrocele postoperative which regressed over a period of approximately 2 months. Similarly, Grossgold and Kusuda[9] reported the occurrence of a hydrocele after complete excision of a scrotal lymphangioma. However, unlike our case, the hydrocele failed to regress and the patient underwent an orchiectomy. Therefore, patients should be counseled for the possibility of an orchiectomy after excision of scrotal lymphangiomas.

Additionally, given the uncertainty of the diagnosis prior to surgical intervention, one must consider approaching the excision via an inguinal approach, as is seen in the case of a radical orchiectomy. In this case, the mass was approached via an inguinal incision due to both the unclear nature of the mass and the presence of an inguinal hernia. However, it is important to note that, in the present case, the main indication to perform a concomitant scrotal incision was rather the sheer size and magnitude of the mass along with the fact that the mass was densely adhered to the scrotal skin, which limited complete resection solely via an inguinal incision. Thus, we had to convert to a scrotal approach to fully expose and remove the tumor.

Diagnosis is typically made on histology after surgical excision. The microscopic changes that occur alongside scrotal lymphangiomas are extra nodal involvements such as cystic endothelial-lined spaces and primitive vascular patterns.[10] Our case showed vessel walls composed of collagen and fibroblasts as well as endothelial lining positive for CD34 and D2-40, all consistent with lymphangioma.

Conclusion

This case outlines the difficult diagnosis, useful imaging modalities, histopathology, and the gold standard treatment of complete surgical excision in patients with scrotal lymphangiomas, an exceedingly rare cause of scrotal swelling. We recommend Urologists consider a lymphangioma in the differential diagnosis of a scrotal mass. Furthermore, Urologists should exercise a heightened level of awareness in the recognition of the entity's clinical presentation and the characteristic imaging findings. □

References

1. Singh S, Baboo ML, Pathak IC. Cystic lymphangioma in children: report of 32 cases including lesions atrrare sites. *Surgery* 1971;69(6):947-951.
2. Koshy A, Tandon RK, Kapur BM, Rao KV, Joshi K. Retroperitoneal lymphangioma. A case report with review of the literature. *Am J Gastroenterol* 1978;69(4):485-490.
3. Flanagan BP, Helwig EB. Cutaneous lymphangioma. *Arch Dermatol* 1977;113(1):24-30.
4. Kajal P, Nain Rattan K, V SM, Garsa V. Scrotal lymphangioma - a rare cause of scrotal swelling. *APSP J Case Rep* 2013;4(1):5.
5. Singla SL, Rattan KN, Singh S. Cystic hygroma of the gluteal region. *Indian J Pediatr* 2000;67(10):779-780.
6. Hurwitz RS, Shapiro E, Hulbert WC, Diamond DA, Casale AJ, Rink RC. Scrotal cystic lymphangioma: the misdiagnosed scrotal mass. *J Urol* 1997;158(3):1182-1185.
7. Loberant N, Chernihovski A, Goldfeld M et al. Role of Doppler sonography in the diagnosis of cystic lymphangioma of the scrotum. *J Clin Ultrasound* 2002;30(6):384-387.
8. Weidman ER, Cendron M, Schned AR, Harris RD. Scrotal lymphangioma: an uncommon cause for a scrotal mass. *J Ultrasound Med* 2002;21(6):669-672.
9. Grossgold ET, Kusuda L. Scrotal lymphangioma in an adult. *Urology* 2007;70(3):590.e591-592.
10. Johansen TL, Søndergaard G, Ebling A. Lymphangioma of the scrotum. *Scand J Urol Nephrol* 1999;33(3):205-206.