Case study

A Huge Benign Cystic Lymphangioma of The Epididymis – An AtypicalCaseof ScrotalSwelling andLiteratureReview

Abstract

Lymphangioma of the epididymis is a rare condition that originates from the sequestration oflymphatic tissue, leading to a lack of normal communication with the lymphatic system. It consists ofunilocular or multilocular lymphatic cysts lined by a single layer of endothelium. This benign tumor istypically identified at birth, during childhood, early adolescence, and occasionally in adulthood, withthe most common locations being the head, neck, and axilla. Lymphangioma of the epididymis isexceptionally uncommon, with the first reported case by Thompson in 1936. Only six cases of cysticlymphangiomaoftheepididymis inadults havebeendocumented inthe Englishliteraturetodate.

Here, we present an atypical case involving a 35-year-old male with a huge benign cysticlymphangioma of the right epididymis, manifesting as a painless inguinoscrotal cystic massextending up to umbilicus. A complete surgical excision was performed, to minimize the risk ofrecurrence.

Keywords

benign,Cysticlymphangioma,epididymis.

Introduction

Lymphangiomas represent congenital lymphatic abnormalities primarily developing in the headand neck (75%), axilla (20%), and more rarely in other locations such as the liver, spleen, kidney,mediastinum, mesentery, retroperitoneum, inguinal region, and scrotum (5%). They account forapproximately 26% of all benign vascular tumors in children, with 50% present at birth, and 90%emerging within the first two years of life. Inguinal and scrotal localization is exceptionallyuncommon. Traditionally classified as capillary, cavernous, and cystic lymphangiomas. In adults mayarise as primary benign cystic neoplasms or secondary to lymphatic obstruction associated withherniasurgeryortrauma. [1,2,3,5]

Microscopically, cysts in lymphangiomas are lined by flattened endothelial cells. The primarymanifestation is a gradually painless mass. However, differentiating scrotal lymphangiomas fromother scrotal pathologies, such as hernia, hydrocele, haematocele, varicocele, epididymal cysts,spermatic cord lipoma, teratoma, dermoid cyst, and epidermoid cyst, can be challenging due tosimilarclinicalfindings. [2,4,5]

Ultrasonography with colour Doppler is a valuable diagnostic tool, and CT and MRI play crucialrolesincaseswithsuspectedpelvicorretroperitonealextension.Adefinitivediagnosisisestablished through histopathological examination. Surgical treatment involves the preferred option of totalexcision of the mass, considered the gold standard in the pediatric age group. Total orchiectomymaybenecessaryfor lesionsinseparablefromthetestisandspermaticcordinadult.[4,5,6]

Casereport

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On March 17, 2020, a 35-year-old male was admitted to our centre complaining of a substantialright inguinoscrotal swelling that had gradually extended up to the umbilicus over the last 2 years.There was no history of trauma or surgery in the past in the inguinoscrotal region. Upon physicalexamination, a painless right scrotal swelling was observed, extending through the right inguinalregion up to the umbilicus. The swelling, measuring 15x10 cm, was tense and cystic, while theopposite testis appeared normal in size and shape. Scrotal ultrasonography revealed a thick-walled,massive solitary cystic lesion arising from the right testis, extending up to the umbilicus. The cysticlesion was filled with internal echoes and debris, and colour Doppler study confirmed it as anavascularcysticlesion.

CT scans of the abdomen, pelvis, and scrotum showed a large thick-walled solitary cystic lesionarising from the right testis, extending intraabdominally up to the umbilicus, measuring 15x10 cm.The patient denied any history of trauma, surgery or pain. The patient's general condition washealthy, and he had two children's. All laboratory investigations showing normal results. TheultrasonographyandCTdiagnosisconfirmedamassivecysticlymphangiomaoftherighttestis. Surgical exploration through the right inguinal route revealed a surprisingly large cystic massoriginatingfromthe righttestis andextending uptoumbilicus,measuring15x10cm.

Right side Orchidectomy was performed along with the removal of the massive cyst. Upon grossexamination, the huge mass weighed approximately 3 kg, and upon cutting the cystic lesion, 2 litersofdirty,yellowish-brownfluidweredrained.Histopathologicalexaminationrevealedamassivecystic mass filled with small cystic spaces lined by flattened endothelium and filled with lymph,suggestinga primarybenigncysticlymphangiomaoftheepididymis.**(Fig1-10)**

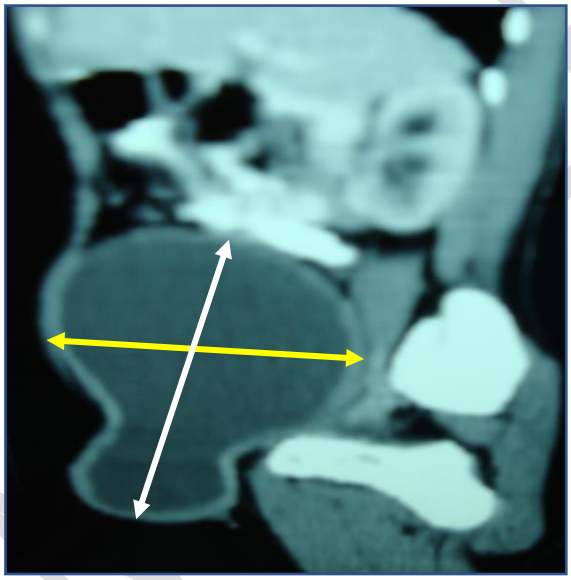

Fig-1CTpelvisandscrotumshowedalargeuniloculcarcysticlesion15x10cmwith extrascrotalpelvisextension

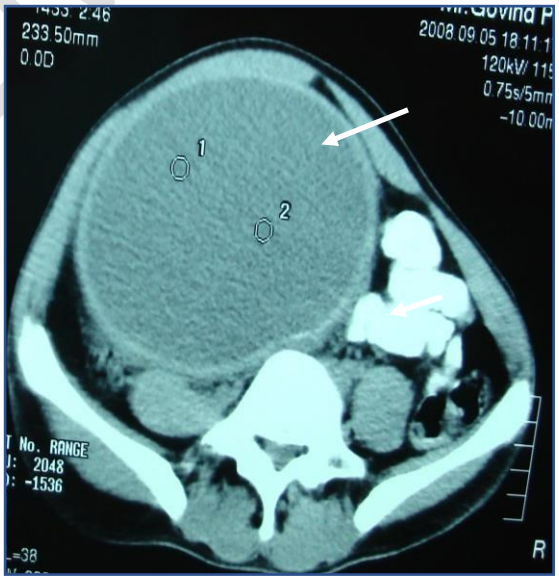

Fig-2 CT pelvis showed a thick intraabdominalcysticswelling

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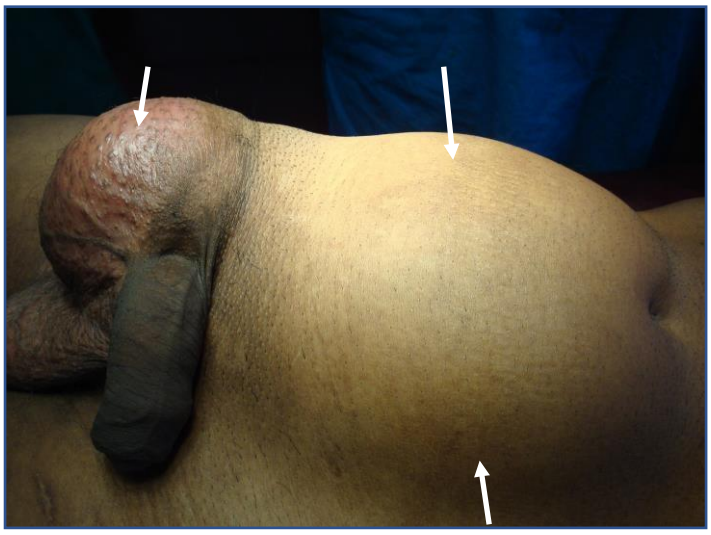


Fig- 3Photographsshowinginguinoscrotals wellingextendingupto umbilicus

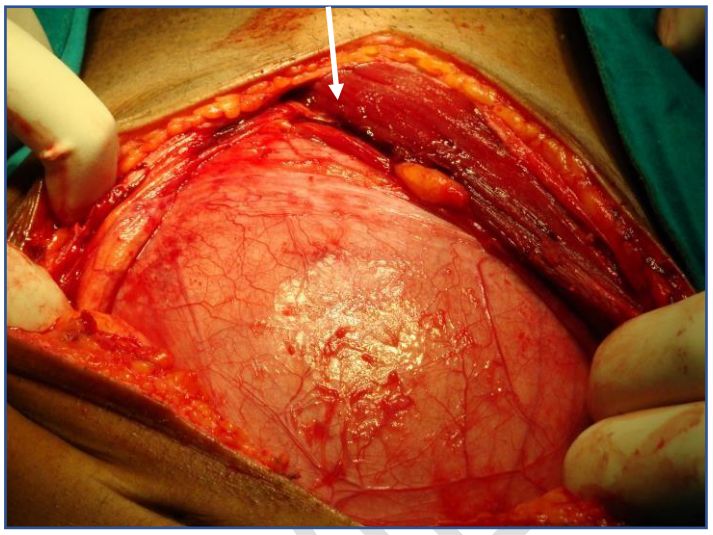


Fig-4 Intraoperative photographs showing a huge cystic massatright inguinalregion

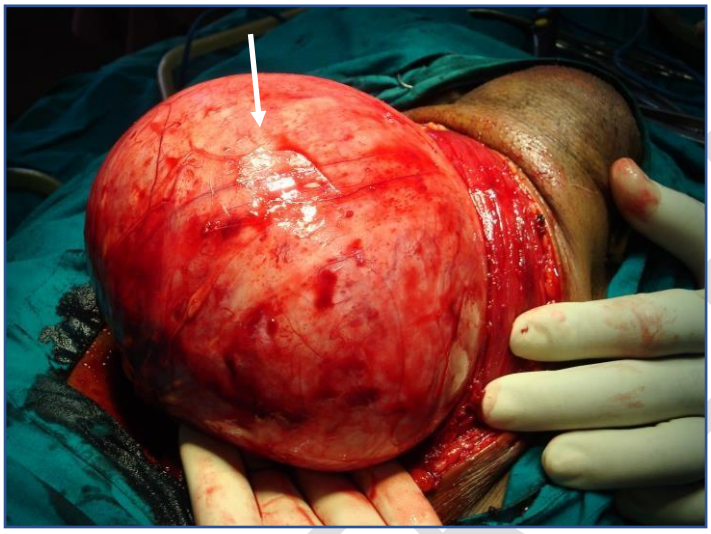


Fig- 5Intraoperativephotographsshowingahug ecysticmassatright inguinalregion

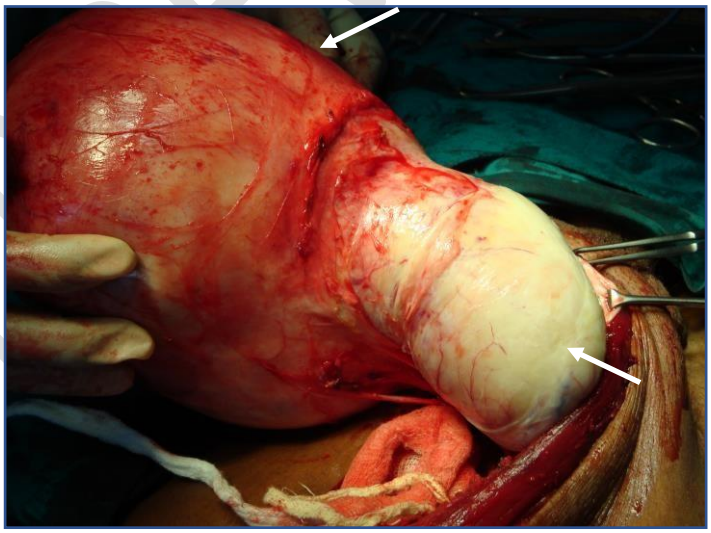


Fig- 6Intraoperativephotographsshowingahuge cysticmassalongwithright testis.

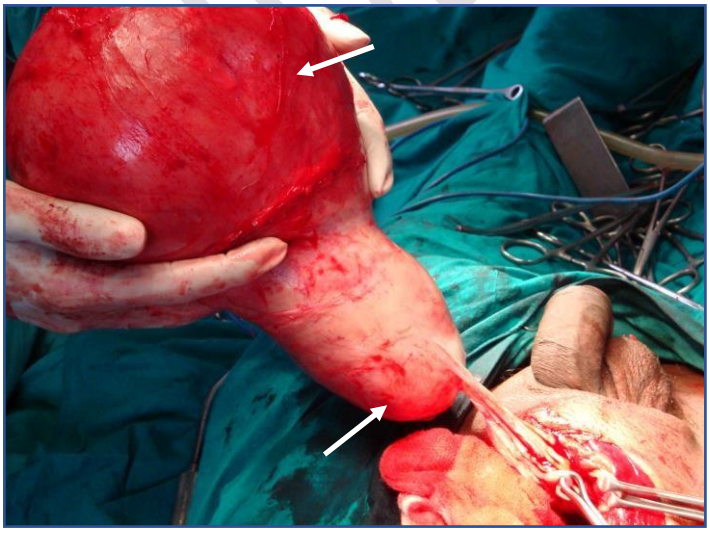
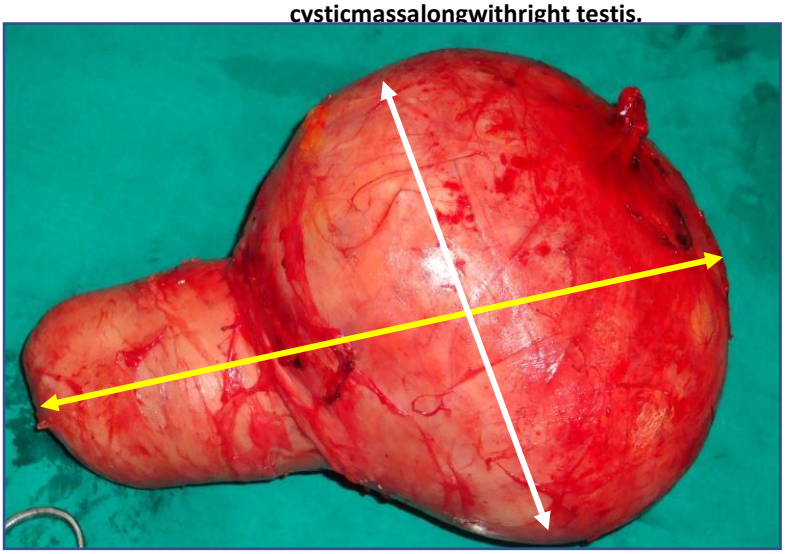


Fig-



7Intraoperativephotographsshowingahugecyst

icmassalongwithright testis.

Fig-8 Photograph showing well circumscribed solitary cystic massofsize15x10cmarisingfromrighttestisandweighing 3kg

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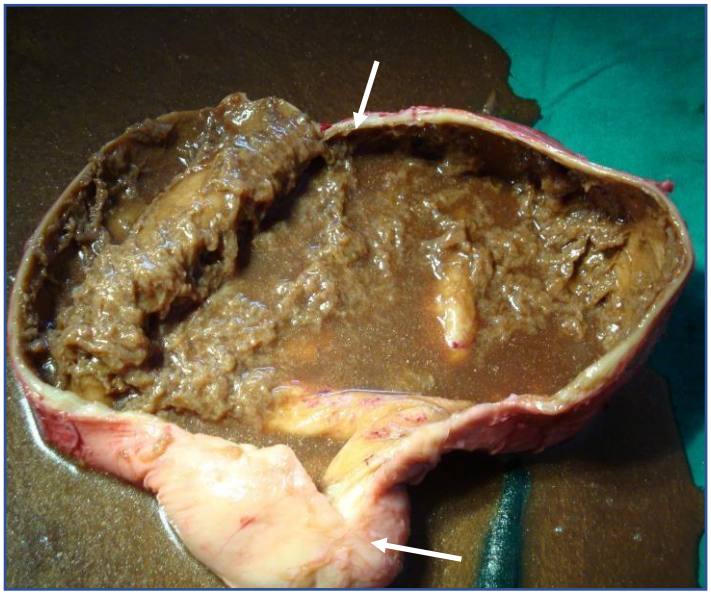

Fig-9 Photographs showing cyst containing brownish, muddymaterialwiththickwallalongwith righttestis

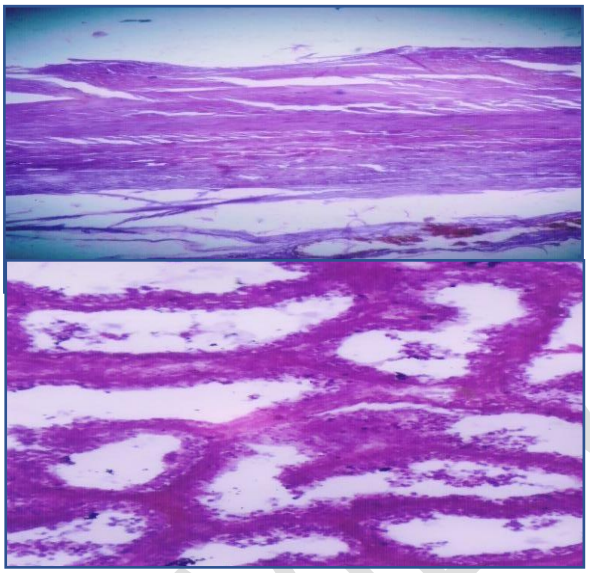

Fig-10Histopathologicalexaminationrevealedmultipledilatedspacesli nedbyasinglelayerofepitheliumfilledwithlymph

Discussionandliteraturereview

Cystic lymphangiomas can either be congenital tumors or arise secondary to factors such aspost-inguinal hernia surgery or trauma. The majority of these lesions (90%) manifest during the firsttwo years of childhood, and occurrences in adulthood are uncommon. Typically, lymphangiomasprimarily affect the head, neck, and axilla (95%), with a rarer (5%) occurrence in the liver, spleen,kidney, mesentery, and inguinoscrotal region. Traditionally, lymphangiomas are classified into threetypes:capillary, cavernous,andcystic lymphangiomas.[2,4,5]

The scrotum and inguinal canal are unusual sites for the development of lymphangiomas, andepididymal lymphangiomas are particularly rare. To the best of our knowledge, only six cases havebeen reported to date, with the initial case documented by Thompson in 1936. Among thepreviously reported cases, one resulted from secondary lymphangiomas post-herniorrhaphy, whiletheremainingfiveweretrueprimary lymphangiomas.[5,6,7]

In 1976, Whimster characterized lymphangiomas as congenital abnormalities stemming fromthe abnormal development of lymphatic channels. These channels originate from sequesteredlymphatic tissue, lacking normal communication with the lymphatic system and presenting as acystic mass. Singh et al reported 32 cases of cystic lymphangioma in children, with only one locatedin the scrotum. Loberant et al reported fewer than 50 cases of scrotal cystic lymphangioma until2002,andHurwitzet al documented seven casesovera 10-yearperiod. [4,5,8]

Intra-scrotal cystic lymphangiomas can be mistaken for other extra-testicular conditions, bothcommon and uncommon, such as hydrocele, varicocele, haematocele, inguinal hernia, epididymalcyst, spermatocele, lipoma of the spermatic cord, and hydatid of Morgagni. Patients with suspectedcystic lesions that extend to the abdomen or pelvis can benefit from ultrasonography and a CT scanof the abdomen or pelvis. Ultrasonography in conjunction with Doppler evaluation can provideimportant information for differential diagnosis and surgical treatment of certain condition. Forscrotal lymphangiomas, biopsies confirm the diagnosis, ultrasonography determines the cysticnature andfluidcomponent andguidesthesurgicalstrategy. [4,5,6]

Scrotum isaveryuncommonlocationforcysticlymphangioma,anditshouldbeconsideredinthedifferentialdiagnosi sofmultiloculated,benign-appearingextratesticularmasseswithinsufficient

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vascular supply in an adult. Lymphomography is not advised because cystic lymphangiomas do notcommunicate with the lymphatic system. Surgical excision of the entire mass is used to treat thedisease.Othertreatment techniques, suchassclerosantinjections,extensive fulguration, and cryotherapy, have failed miserably. Because of the lack of availability in our hospital of othertreatmenttechniques,such assclerotherapy, wewerechosenforsurgicalexcisionofthe entiremass. The cyst lymphangioma must be removed completely to prevent a recurrence. Completesurgical excision is considered the gold standard treatment modality, and in some cases,orchidectomy may be necessary in adults with extensive cystic lymphangiomas at the epididymis andcomplete orchidectomymayberequired.[2,4,5]

In our reported case, the patient presented at 35 years of age with two children's and amassive benign cystic lymphangioma extending from the inguinoscrotal region up to the umbilicus.We performed a complete orchidectomy along with the removal of the cystic mass. This caserepresents the first documentation of such a sizable benign cystic lymphangioma of the epididymis,weighing3kg, intheliterature.

Conclusion

Primarybenigncysticlymphangiomaoftheepididymisisanextremelyrarecondition.In children's the scrotal cystic lymphangioma is completely excised while preserving the testis and toprevent recurrence. It is the gold standard treatment. So preoperative a proper diagnosis of thescrotal lymphangioma and its extent using the ultrasonography, CT and MRI is essential to planninganappropriatesurgicalapproach.

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