GAVIN PUBLISHERS

OPEN ACCESS

Annals of Case Reports

Kuypers TJL and Bemelman M. Ann Case Report 6: 596.

DOI: 10.29011/2574-7754.100596

Case Report

Chylocele-A Rare Pathology Presenting Initially as an Inguinal Hernia

Kuypers TJL*, Bemelman M

Department of Surgery, Elisabeth-Tweesteden Hospital, Tilburg, Netherlands

***Corresponding author:** T. J. L. Kuypers, Department of Surgery, Elisabeth-Tweesteden Hospital, Tilburg, Netherlands

Citation: Kuypers TJL, Bemelman M. (2021) Chylocele - A Rare Pathology Presenting Initially as an Inguinal Hernia. Ann Case Report 6: 596. DOI: 10.29011/2574-7754.100596

Received Date: 01 March, 2021; **Accepted Date:** 03 May, 2021; **Published Date**: 07 May, 2021

Abstract

We present a rare case of a young male who was initially diagnosed with an inguinal hernia that subsequently presented as a chylocele-a term which we will use in this paper as being appropriate to the circumstances. Following the initial diagnosis, during surgery we discovered a chylocele-a chylous filled peritoneal sac protruding through a normal inguinal canal. The chylocele was caused by the rare Hennekam syndrome accompanied with chylous ascites. In this paper, we share our experience and challenges with this rare pathology. To reduce the chance of chyle leakage and its corresponding morbidity, we recommend using either the Lichtenstein repair for patients with distended internal ring or a simple herniatomy for patients with a normal internal ring. We do not recommend opening the preperitoneal plane.

Keywords: Chylocele; Inguinal hernia; Hennekam syndrome; Ascites.

Introduction

The clinical diagnosis of a symptomatic inguinal hernia leads to different treatment options - open or laparoscopic. Surgeons choose the preferred technique preoperatively based on a clinical examination and adequate patient medical information. This however, may need to be changed during surgery as a consequence of unforeseen circumstances. We present a rare case of a chylocele, a chylous filled peritoneal sac protruding through a normal inguinal canal, which changed the preoperative plan to place a preperitoneal mesh.

Case

A twenty-seven-year-old male presented in our hospital with a symptomatic recurrent inguinal hernia on the right side. The relevant medical history of the patient was Hennekam syndrome (a very rare autosomal recessive inheritance of lymphangiectasia) [1]. Ventricular and atrial septum defect repair at six weeks of age. A right inguinal hernia repair at the age of twelve years, and a video-assisted thoracoscopic talcage for pleural effusion followed with a decortication complicated by a chylous leakage at the age of twenty-four years.

The inguinal hernia existed for over eight years but his complaints progressed during the several months preceding his presenation at the hospital. He experienced a stinging pain in his right groin both during exercise and while sitting. Clinical examination revealed an inguinal bulge on the right side, which progressed with valsalva. It was easily reduced with no signs of entrapment. A transinguinal preperitoneal hernia repair was proposed.

No herniation was found initially during surgery. White ellipse shaped firm structures, as large as a grain of rice, were found with dissection running along the spermatic cord. During further dissection, small amounts of white fluid were released from the tissues. With further exploration of the spermatic cord, we noticed a small indirect hernia, which filled intermittently with white fluid, depending on the abdominal pressure (Figure 1, Video 1). After opening the hernial sac, no fat or intestine was found. We suspected an indirect hernia due to chylous ascites. Samples were taken from the fluid and from the small rice grain shaped structures. Examination of the internal inguinal ring revealed a strong near to normal sized annulus.

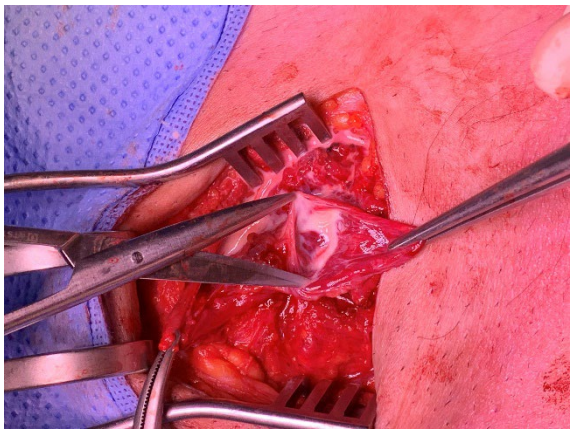

Figure 1: opened peritoneal sac, filling with chyle when abdominal pressure is applied.

We had concerns about perforating the peritoneum with a preperitoneal mesh because we suspected the patient to have a considerable amount of chyle in the peritoneum. Given the patient had an almost normal inguinal internal ring we decided to resect the primary hernial sac and reef the internal annulus instead of using a reinforcing mesh.

Cytologic analysis of the fluid confirmed our suspicion of chyle. The histologic report of the rice grain shaped hard nodule revealed lymphangiectasia. The postoperative control, four weeks later, revealed an uneventful recovery with no chyle leakage or hernia recurrence. Six months later, he presented with a completely different problem but a physical examination revealed no inguinal hernia recurrence**.**

Discussion

Chylous ascites is uncommon in young adults and could have a wide variety of causes. A Pubmed search for inguinal herniation with chylous effusion only revealed case reports in infants. The theory of these chylous ascites was that an entrapped hernia or malrotation damaged the intestinal lymphatic vessels that caused chylous effusion [2,3]. However an inguinal hernia is a known pitfall in the diagnosis of ascites, in patients with cirrhosis [4]. There are no published reports of chylous ascites causing inguinal herniation. We present a case with a recurrent inguinal hernia together with chylous ascites which were part of the patient's recently diagnosed Hennekam syndrome. We hypothesize that the hernia was caused by his chylous ascites and the corresponding hydrostatic pressure. To our knowledge, this has not previously been published.

The Hennekam syndrome is a very rare autosomal recessive inheritance of lymphangiectasia, first described by Dutch physician Hennekam in 1989 [1,5]. Worldwide there are approximately fifty patients with this syndrome [5]. It can cause lymphoedema in the legs, facial anomalies, mental retardation, intestinal malabsorption, chylous ascites and a limited life expectancy which can vary from death in childhood to survival into adulthood depending on the severity of the condition [5,6]. Due to the atypical presentation without herniated fat or intestine and a normal internal inguinal ring we did not consider the term inguinal hernia appropriate for this case. We propose a term called chylocele, previous described by Marabelli [7]. We describe the chylocele as a peritoneal sac filled with chyle protruding through a normal inguinal canal.

During surgery we doubted what the best option for repair would be. Normally, we would place a mesh preperitoneal, for repair of an inguinal hernia. There is however a significant risk of damaging the peritoneum when creating the preperitoneal space for such a mesh given our hypothesis that a considerable chyle leakage could cause significant morbidity (fistula, infection, mesh irritation). Therefore, we decided only to resect the hernia sac and reduce the diameter of the internal annulus. Our concerns about preperitoneal perforation and chyle leakage with fistula formation also apply to any endoscopic technique e.g. transabdominal preperitoneal (TAPP) repair or totally extraperitoneal (TEP) repair. Both would have a significant risk for chyle leakage. By definition the peritoneum is breached with the TAPP and with TEP the peritoneum is opened in a significant number of cases. However, there are studies stating that TEP and TAPP are safe procedures in inguinal hernia in patients with liver cirrhosis and ascites [8,9], but this needs to be confirmed in larger studies.

Another option is the Lichtenstein repair which leaves the preperitoneal plane intact and creates, with the mesh, a new internal annulus with a fibrous layer covering the inguinal canal. This seems to be the safest procedure with the least chance of recurrence. In our case we did not opt for the Lichtenstein due to the normal caliber of the internal ring. Even though the postoperative control showed a good result with an uncomplicated asymptomatic patient, we nevertheless realize that recurrence is plausible in time. Optimization of the Hennekam syndrome and the chylous ascites could minimize the risk of recurrence.

Conclusion

We present a rare case of a chylous herniation in a normal inguinal canal (referred to as a chylocele) in conjunction with the very rare Hennekam syndrome. The combination of an inguinal herniation through a normal internal annulus combined with a syndrome characterized with chylous ascites poses a challenge in the technique used to treat the hernia. In reducing the chances for chyle leakage and its corresponding morbidity we recommend using the Lichtenstein repair for patients with distended internal ring and a simple herniatomy for patients with a normal internal ring. We do not recommend opening the preperitoneal plane.

Acknowledgments

The patient in this case signed an informed consent for publication of this case report. M. Bemelman performed the operation assisted by T. Kuypers. T. Kuypers wrote the manuscript with the help of M. Bemelman. The authors received no financial support for the research, authorship and publication of this article. The authors of this manuscript declare they have no affiliations with or involvement in any organization or entity with any interest in the subject matter discussed in this manuscript.

References

1. Hennekam RC, Geerdink RA, Hamel BC, Hennekam FA, Kraus P, et al. (1989) Autosomal recessive intestinal lymphangiectasia and lymphedema, with facial anomalies and mental retardation. Am J Med Genet. 34: 593-600.
2. Santoro E, Shaw A. (1970) Chylous ascites secondary to incarcerated inguinal hernia in an infant. Am J Surg. 119: 579-580.
3. Zarroug AE, Srinivasan SK, Wulkan ML. (2010) Incidental chylous fluid during hernia repair may be a harbinger of malrotation. J Pediatr Surg. 45: E17-8.
4. Adar T, Mizrahi M. (2013) Inguinal hernia - a possible pitfall in diagnosing ascites. Liver Int. 33: 1449.
5. Lee YG, Kim SC, Park SB, Kim MJ. (2018) Hennekam Syndrome: A Case Report. Ann Rehabil Med. 42: 184-188.
6. Menon J, Venkatesh V, Thirunavukkarasu B, Lal SB. (2019) Hennekam syndrome: an uncommon cause of chylous ascites and intestinal lymphangiectasia in the tropics. BMJ Case Rep. 12: e229419.
7. Marabelli A. (1966) [On a case of chylocele. Physiopathogenetic considerations]. Arch Sci Med (Torino). 121: 83-89.
8. Ohuchi M, Inaki N, Nagakari K, Kohama S, Sakamoto K, et al. (2019) Transabdominal preperitoneal repair using barbed sutures for bilateral inguinal hernia in liver cirrhosis with ascites. J Surg Case Rep. 2019: rjz199.
9. Wang H, Fu J, Qi X, Sun J, Chen Y. (2019) Laparoscopic totally extraperitoneal (TEP) inguinal hernia repair in patients with liver cirrhosis accompanied by ascites. Medicine (Baltimore). 98: e17078.