Laparoscopic excision of a retroperitoneal lymphatic malformation in a newborn☆

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Abstract Abdominal lymphatic malformations may be challenging to eradicate. Retroperitoneal lesions may be more difficult to resect than mesenteric ones; however, the latter may predispose to intestinal volvulus, leading to calls for their prompt excision. Such lesions identified perinatally may pose particular challenges: in one case, respiratory failure caused by abdominal distension required emergency drainage followed by later laparoscopic excision; laparoscopy has also been used promptly to diagnose and resect neonatal mesenteric lymphatic malformations with their inherent volvulus risk. We illustrate that even if neonatal laparoscopy identifies a retroperitoneal rather than mesenteric lymphatic malformation, curative endosurgical excision remains feasible.

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1. The case

A male patient was admitted within 24 hours of birth with an antenatally identified intraabdominal cystic mass. Inspection demonstrated a large left flank swelling which palpation revealed to occupy much of the left side of the abdomen. The ipsilateral testis was palpable. No other anomalies were identified, and the baby fed and passed both urine and meconium normally. Postnatal ultrasound confirmed a large multicystic lesion occupying much of the left side of the abdominal mass.

Although most lymphatic malformations present around head, neck and axillae, abdominal lesions do occur and vary between retroperitoneal and mesenteric lesions [1]. Prenatal lymphatic malformations may present symptomatically on antenatal scans or be associated with stillbirths or life-threatening respiratory failure, infection, and chylous leak [2]. Management options include observation, drainage ± sclerotherapy, laser therapy, and surgical excision [3–5]. However, for mesenteric lymphatic malformations, expeditious removal has been recommended to minimize risks of intestinal volvulus [6]. Surgery for large neonatal abdominal lymphatic malformations has generally required laparotomy [7]. However, in older children, several centers have reported laparoscopic excision [8–10]. In neonates, laparoscopic-only approaches appear rare and confined to mesenteric lymphatic malformations [2,11–13]. We show that even if laparoscopy identifies a retroperitoneal rather than mesenteric lymphatic malformation, curative endosurgical excision remains feasible in the newborn.
abdomen but failed to identify the impalpable testis. The renal tract appeared normal. α-Fetoprotein and β-human chorionic gonadotrophin levels were not abnormally elevated. Similarly, computed tomography was not particularly suggestive of a germ-cell tumor (Fig. 1). Diagnostic and therapeutic laparoscopy was performed at 2 weeks of age. A large multilocular lesion with a dominant cyst was identified extending from the left flank to groin (where palpation under anesthesia had revealed it to be fixed). Second and third ports were therefore introduced in the left upper quadrant and right iliac fossa to allow the anterolateral surface of the lesion to be fully mobilized. At this stage, to allow a safe dissection of the posterobasal aspects of the lesion, it was electively drained via a small window directly into an endoscopic sucker. This step allowed the lesion to be gradually mobilized off its origins: the ipsilateral common iliac vessels, ureter, and external iliac vessels. The lesion was delivered via a port site. At this first surgery, a small atrophic-looking left testis was also identified within the lower abdomen. Excision of small residual cystic elements surrounding the testicular vessels and vas deferens was deferred until a repeat laparoscopy several months later, which was planned to see if a first stage Fowler Stevens orchidopexy would be worthwhile (when in fact the small and atrophic-looking left testis was excised with accompanying residual cysts). Postoperative recovery was uneventful, and histology confirmed a lymphatic malformation. Follow-up ultrasounds now at more than 24 months have shown no recurrence.

2. Discussion

Management of large abdominal lymphatic malformations can vary from straightforward to extremely challenging [14]. In contrast to lymphatic malformations elsewhere, the risk of volvulus with mesenteric lesions led to a recommendation that early surgery be pursued for presumed abdominal lymphatic malformations [6]. In our case, the impalpable ipsilateral testis raised additional concerns that the sonographically identified lesion might represent a neoplastic abdominal testicular lesion. Following the antenatal diagnosis, this baby’s parents were also keen to have the lesion removed. For these reasons and after careful discussion with the parents, surgery was selected over sclerotherapy and other modalities in this case. In contrast to observation or sclerotherapy, minimally invasive surgery offered the parents the prospect of a histologic diagnosis alongside assessment of the impalpable testis. With a lesion extending from the left kidney and adherent to the ipsilateral iliac vessels at the level of the inguinal ligament, a conspicuous laparotomy incision would have been needed to allow safe clearance. In contrast, laparoscopy provided excellent visualization of the lesion, the vasculature, and the impalpable testis while avoiding laparotomy. Furthermore, after optical trocar placement, we were able to position the subsequent trocars to optimize dissection of the lymphatic malformation from its roots around the ipsilateral common and external iliac vessels.

Neonatal laparoscopy permits prompt diagnosis and treatment for prenatally suspected abdominal lymphatic malformations. While avoiding laparotomy, this approach can promptly treat mesenteric lymphatic malformations at risk of intestinal volvulus [6]. Moreover, if laparoscopy rules out a mesenteric location, it is nevertheless feasible to excise the retroperitoneal lymphatic malformation endoscopically.

References