Elective suprarenal and infrarenal cavectomy for excision of giant retroperitoneal teratoma in infancy

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Abstract Retroperitoneal teratomas are rare, often massive tumors usually presenting in infancy; being mostly mature lesions, their treatment is surgical but may represent a formidable challenge. Major vessel displacement may not be well demonstrated on imaging: vascular injuries are well-recognized surgical complications with urgent repair, ligation, or even segmental excision of major vessels being required. However, the literature provides few suggestions to avoid these problems. In our approach, we assessed the important effaced abdominal veins on imaging and at laparotomy to allow us to electively excise the suprarenal and infrarenal vena cava (with both renal vein ostia) and thereby resect a giant retroperitoneal teratoma without inadvertent vessel injury, major bleeding, renal disturbance, or tumor recurrence. Described for renal tumors, elective cavectomy has not been reported as a technique to manage primary retroperitoneal teratomas. In selected cases, with careful preservation of renal venous collaterals, we show this can be a well-tolerated, preemptive option to reduce the high risks of surgical complications.
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Retroperitoneal teratomas are uncommon, usually benign tumors of infants and young children whose treatment is currently excision. Unfortunately, their massive size and location means that major vessel anatomy can be grossly distorted [1]. Because of rarity, most surgical series of these tumors are small; perioperative deaths and major vascular injuries remain problems even for experienced surgical oncologists, with the exception of Luo et al [2-5]. In the United Kingdom's largest contemporary surgical report, major bleeding was a problem in all 6 cases of retroperitoneal teratoma [4]. Injuries to aorta, portal vein, renal vein, inferior vena cava (IVC), esophagogastric junction, and bile ducts have also been reported [1-4]. Although preoperative imaging can be unreliable, aortic displacement is typically contralateral to the side of lateral lesions, whereas with "bilateral" or central tumors, it remains midline, with anterior displacement unreported. In contrast, failure to identify the IVC on preoperative imaging scans is frequent, and at operation, its displacement by tumor is typically anterior [1]. Vascular encasement appears less common than in neuroblastoma. Given the imaging limitations and displacement of major vessels and kidneys, it is not surprising that life-threatening hemorrhage has at times necessitated urgent repair, ligation,

and even excision of major vessels such as the infrarenal IVC [1-4].

We report a preemptive approach to a giant, bilateral retroperitoneal teratoma with gross vascular and renal displacement, where elective cavectomy with both renal vein ostia allowed excision without vascular injury or major bleeding. Although elective cavectomy has been described for renal tumors invading the IVC, we have not encountered a previous report of successful elective cavectomy (with both renal vein ostia) for excision of primary retroperitoneal teratoma [6-9].

1. Case report

A female child presented at 4 months of age with feeding difficulties and a huge nontender abdominal mass. Abdominal ultrasonography confirmed the presence of a huge, complex mass but did not reveal the distorted vascular anatomy. Contrast-enhanced computed tomographic (CT) scanning demonstrated a massive heterogeneous tumor occupying most of the abdomen, featuring scattered calcifications and gross displacement of both kidneys (Fig. 1). The IVC was not visualized. Tumor markers, serum α-fetoprotein and β-human chorionic gonadotropin, were not abnormally elevated for her age. A nasogastric tube was placed, but enteral feeds were not tolerated. A central venous line was inserted for parenteral nutrition, and a biopsy was performed. Histology was consistent with a mature teratoma. After

Fig. 1 Coronal reconstruction of the preoperative contrast-enhanced CT scan shows the bilateral abdominal tumor and gross displacements of right (oblique arrow) and left (horizontal arrow) kidneys.

multidisciplinary discussion, she was referred to the corresponding author for consideration of surgical excision. Owing to the massive tumor size and the bilateral renal displacements, the parents were counseled regarding the risks of mortality, major bleeding, unresectability, and renal failure.

2. The surgical procedure

Preoperative imaging suggested that the IVC was occluded and the kidneys had developed collateral drainage. Armed with this knowledge, a transverse laparotomy was performed and revealed a huge heterogeneous mass filling the abdominal cavity with centrifugal displacement of the intestines and other organs. Barely recognizable as tan flat structures, the IVC and medial parts of both renal veins were tautly stretched across the anterior surface of the tumor; the lateral parts of both renal veins were patent but draining into collateral vessels running toward the pelvis on the lateral surfaces of the tumor; although the aorta and renal arteries were beyond initial view, the inferior mesenteric artery was cleared to provide a guide toward the aorta. The superior mesenteric artery (SMA) was seen drawn over the upper part of the tumor in the midline. Dissecting the renal collateral veins away from the tumor allowed them to be preserved. To safely deliver the entire tumor, we confirmed that the IVC was chronically occluded before deciding to electively resect it from an infrahepatic to suprailiac level. We also ligated and divided both renal veins in their medial occluded portions, carefully preserving the collateral venous drainage from the lateral parts of the renal veins. This allowed the tumor to be dissected off the posteriorly located aorta, completing the resection without inadvertent vascular damage or major bleeding. Careful posterolateral mobilization, mindful of the renal arteries and ureters, allowed both kidneys to be freed from the tumor without injury to either. On the left side, the spleen and pancreas had been rotated upward by the tumor such that the splenic vessels lay posteroinferior to the pancreas. Tracing these medially allowed the celiac axis and SMA to be cleared and the tumor to be removed. Postoperative recovery was uneventful, and renal function remained normal. Histology confirmed a mature teratoma including an eyelike structure. Slow to feed orally at first, the child is now thriving. Now, more than 3 years later, no recurrence has been demonstrated clinically, on magnetic resonance imaging or ultrasound scanning (Fig. 2). Similarly, serum tumor markers, α-fetoprotein and β-human chorionic gonadotropin, remain normal.

3. Discussion

Large retroperitoneal teratomas in infancy represent a continuing surgical challenge [4]. Gross anatomical distortion predisposes to life-threatening injuries to major blood

Fig. 2 Postoperative magnetic resonance imaging showing normally located right (oblique arrow) and left (horizontal arrow) kidneys.

vessels and adjacent organs [1]. In contrast to the relatively predictable vascular encasement seen in neuroblastoma, the vascular anatomy of retroperitoneal teratomas can be highly variable [1]. Worse still, preoperative imaging may be unable to predict accurately the positions of the major vessels [1]. Although less invasive therapies would be welcome, alternatives such as tumor embolization appear unpromising because, unlike sacrococcygeal teratomas, retroperitoneal teratomas typically lack a dominant feeding vessel to target [1]. In light of this, we have reviewed the limited surgical literature on this difficult tumor and demonstrated an approach to excision that allowed successful resection of a massive bilateral tumor in a 4-month old without inadvertent vessel injury or major bleeding.

Preoperative scans provided some useful information. Ultrasound scanning confirmed a huge, complex mass but did not reveal the distorted vascular anatomy. Highly compressed major veins may have little or no flow to visualize on Doppler study and may also be obscured by the anteriorly displaced and gas-filled bowel. Computed tomography was preferred over magnetic resonance imaging in our case because of rapid access and short scanning times without general anesthesia; published experience does not indicate a major advantage of either for this tumor [1]. In keeping with previous descriptions, CT showed that the aorta remained posterior to the tumor and, as with other central or bilateral tumors, in the midline. Sagittal CT reconstructions revealed the SMA coursing above the tumor in the midline. Unraveling the venous anatomy was more difficult, but some clues were helpful. First, failure of the IVC to opacify and the presence of potential collateral vessels on contrast-

enhanced CT suggested that, given a tumor of this size, the IVC was occluded. Second, the literature demonstrates that such effacement and occlusion is typically associated with anterior caval displacement [1]. Third, consideration of primordial germ cell (PGC) development can help: aberrant PGCs are held to be precursors for retroperitoneal teratomas. Normally, PGCs migrate from the midline mesentery toward the developing gonad near the primitive kidney [10]. This helps explain not only these tumors' proximity to mesenteric vessels but also the intimate renal involvement often observed. In our case, the displaced renal hila reflected, in microcosm, the features of the vascular displacement in general: each was expanded by tumor with the artery pushed posteriorly, vein anteriorly and the ureter effaced on the lateral tumor surface. Thus, PGC development predicts that the kidney(s) will often be displaced by these retroperitoneal teratomas and prioritizes CT estimation of the renal positions. The surgeon can then estimate where on the tumor each renal vein is stretched and in which directions they run. Tracing the medial parts of the renal veins even allows an effaced, empty IVC to be identified stretched over the tumor (and avoids inadvertently mislabeling a sizeable venous collateral as the IVC).

Approaches for neuroblastic tumors use systematic display of tumor-encased vasculature: vessel clearance is achieved by bivalving the tumor along the course of each involved vessel [11,12]. However, in several reported retroperitoneal germ cell tumors, as well as the present case, the venous network is instead effaced over the anterior tumor surface, "pinning" it in place [13]. In the presence of well-developed venous collaterals, effaced major veins could be overlooked and, therefore, injured during the vessel display as used for neuroblastoma. Instead, our technique prioritized determination of the venous anatomy prompted by the preoperative scans. This allowed the chronic occlusion of the IVC and medial parts of both renal veins to be noted early with 2 important consequences. First, it indicates that cavectomy to release the tumor may permit tumor clearance without incising it as per neuroblastoma; with large non–chemotherapy-treated lesions, avoidance of such incision may decrease blood loss independently of reducing vascular injuries. Second, it emphasizes the need to preserve venous collaterals particularly from the kidneys; where such collaterals are absent, it is likely that the cava is less tightly drawn over the tumor, caval obstruction is incomplete, and its resection is unnecessary; although temporary caval division may still be considered in this scenario, we delivered a similar tumor by careful caval mobilization alone [13].

In contrast to the vascular display for neuroblastoma, our approach is focused on (1) early assessment of the typically anterior and unique venous anatomy (comprising occluded vessels and important collaterals), (2) resection of occluded veins where necessary (if redundant), followed by (3) centripetal tracing of vessels from peripherally displaced organs that can be more readily located once the tumor is unbound from the overlying venous network. In our case, therefore,

elective cavectomy taking both renal vein ostia facilitated complete tumor excision without the inadvertent vessel injury and major bleeding that have accompanied several reported excisions.

Acknowledgments

References

[1] Hart J, Mazrani W, Jones N, et al. Upper abdominal teratomas in infants: radiological findings and importance of the vascular anatomy. Pediatr Radiol 2008;38:750-5.

[2] Chaudhary A, Misra S, Wakhlu A, et al. Retroperitoneal teratomas in children. Indian J Pediatr 2006;73:221-3.

[3] De Backer A, Madern GC, Hazebroek FWJ. Retroperitoneal germ cell tumors—a clinical study of 12 patients. J Pediatr Surg 2005;40: 1475-81.

[4] Jones NM, Kiely EM. Retroperitoneal teratomas—potential for surgical misadventure. J Pediatr Surg 2008;43:184-7.

[5] Luo CC, Huang CS, Chu SM, et al. Retroperitoneal teratomas in infancy and childhood. Pediatr Surg Int 2005;21:536-40.

[6] Coelho S, Goncalves A, Carvalho RV, et al. Cavectomy for treatment of clear cell renal sarcoma. Pediatr Blood Cancer 2009;53:882-3.

[7] Renaud EJ, Liu D, Pipe SW, et al. Inferior vena cavectomy for nonexcisable Wilms' tumor thrombus. J Pediatr Surg 2001;36:526-9.

[8] Ribeiro RC, Schettini ST, de Campos Vieira Abib S, et al. Cavectomy for the treatment of Wilms tumor with vascular extension. J Urol 2006;176:279-83.

[9] Vicente Prados FJ, Tallada Bunuel M, Pastor J, et al. Renal adeno-carcinoma invading vena cava: state of the art of its diagnosis and treatment by total segmental cavectomy. Arch Esp Urol 1998;51:35-41.

[10] West J, Viswanathan S, Yabuuchi A, et al. A role for Lin28 in primordial germ-cell development and germ-cell malignancy. Nature 2009;460:909-13.

[11] Kiely E. A technique for excision of abdominal and pelvic neuroblastomas. Ann R Coll Surg Eng 2007;89:342-8.

[12] Jawaid W, Solari V, Mahmood N, Jesudason EC. Excision of ganglioneuroma from skull base to aortic arch. J Pediatr Surg 2010;45: E29-E32.

[13] Jawaid W, Solari V, Howell L, Jesudason EC: Excision of extensive metastatic dysgerminoma to control refractory hypercalcaemia in a child at high risk for tumour-lysis syndrome. J Pediatr Surg, (in press).