General Surgical Guidelines

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12.1 Introduction

Surgical resection in Wilms' tumor (WT) is the backbone of multidisciplinary regimen for achieving the objective of complete cure in the child. The first successful extirpation of a WT in a child was performed by Thomas Richard Jessop in 1877 [1, 2]. However, it was not until the beginning of the twentieth century that surgery became the effective therapy for this tumor. The concepts about the extent of surgical expatriation have been forever changing. Both Ladd [3] and Gross [4] recommended simple nephrectomy for WT. Gross [4] suggested removing only fat clinging to the tumor with the affected kidney. It was Robson [5] who championed radical nephrectomy (RN), which includes excision of the entire kidney with the tumor, Gerota's fascia, adrenal gland, and ureter (Fig. 12.1). Most of the cooperative consortia globally consider RN with lymph node (LN) sampling (selective lymphadenectomy) as the benchmark for surgical excision of pediatric renal tumors including Wilms' tumor (WT) and anything short of it is taken as protocol violation. The only concession that is made nowadays is avoidance of excision of the adrenal gland, if possible.

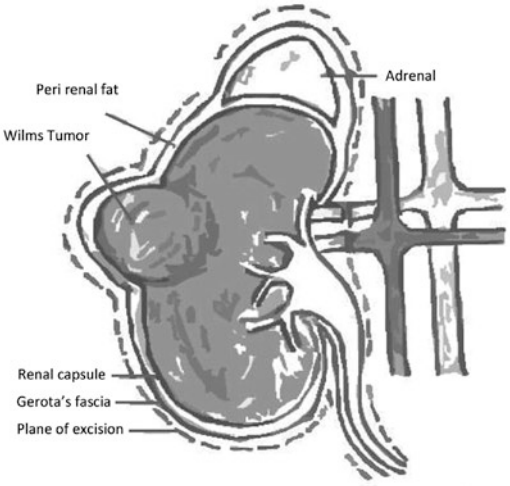

Fig. 12.1 Radical nephrectomy: note the plane of excision is outside that of Gerota's fascia

Notwithstanding this, dissenting voices have been raised from different parts of the world for as long as one third of the century. Philadelphia group first indicated in 1985 that there was no data supporting the need of RN in children with WT [6]. Afterward, Kelalis and Mesrobian [7] also made observations that in children with WT simple nephrectomy (SN), that is, excision of kidney with tumor and perirenal fat, but nonremoval of adrenal gland and Gerota's fascia (Fig. 12.2), may be associated with good overall survival (OS) rates, similar to those obtained

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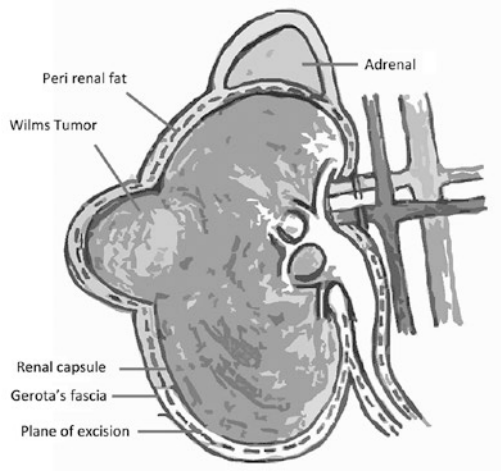

Fig. 12.2 Simple nephrectomy: note the plane of excision is outside the renal capsule; the perirenal fat attached to the kidney is removed along

with RN. Ramon et al. [8], an adult urology group, had begun to doubt the usefulness of RN even in adults with renal cell carcinoma (RCC) and have not found any statistically significant difference between the group of patients with RCC treated with SN and the group treated with RN. Zani et al. [9] justified leaving Gerota's fascia and perirenal fat behind in stages I and II; they had only two patients in stage III so didn't make any clear recommendation for that stage. They felt that as such WT is too large, the distinction between RN and SN is often irrelevant. Szymik-kantorowicz et al. [10] from Poland believed that surgical extent should be also risk stratified, similar to the way it is done for chemotherapy (ChT) and radiotherapy (XRT). They felt RN was non-compulsory in Stage I WT wherein majority of these children could be managed with either simple SN, or nephron-sparing surgery (NSS), based on the size of the tumor. They prescribed SN for tumors of more than 5 cm and NSS for tumors less than 4 cm of diameter. Umbrella protocol of RTSG of SIOP recently legitimized NSS as an acceptable surgical treatment of small volume localized tumors [11].

In this chapter, general recommendations for unilateral nephroureterectomy and surgery for horseshoe kidney with WT are mentioned.

12.2 General Surgical Guidelines for Nephroureterectomy for Unilateral Wilms' Tumor

12.2.1 Access

The patient is placed supine with a rolled towel or bolster placed under the loin on the side of the tumor. Access through a generous transverse abdominal incision is the preferred option. The thoracoabdominal approach may be useful in huge masses located high in the abdomen, but a few authors have reported a higher complication rate with this incision [12]. Whatever the incision, LN sampling must be done. The flank incision, the paramedian incision, and midline incisions are to be avoided; the flank incision doesn't allow adequate LN sampling [13], and the other incisions have been known to be associated with higher rates of intraoperative spill (IOS) [14].

12.2.2 Inspection of the Abdominal Cavity

A self-retaining retracting system is an essential aid to adequate exposure. To start with, any peritoneal fluid, especially hemorrhagic, should be collected for malignant cytological examination. The next step is to inspect and examine the entire abdominal cavity including the liver, LN, and peritoneum for the presence of metastatic lesions, which if present, should be excised (in resectable lesions) or biopsied (in unresectable lesions) and sent for histopathology in a separate container with a clear mention of its origin.

If preoperative imaging is completely normal, exposure of the contralateral kidney is not required except in syndromic WT (risk of bilateral tumors is high) or in WT with high-risk tumor biology [15]. However, if contralateral kidney lesion is diagnosed on preoperative imaging, then the assessment of the contralateral kidney gets precedence over the ipsilateral nephrectomy. Some authors suggest use of intraoperative ultrasound to localize the lesion [16]. Resection of large WT >12–15 cm diameter

increases the risk of IOS due to added on vascularity and intra-tumoral necrosis in large areas [13, 17], and thus, careful handling of the kidney having WT is mandated. Apart from the regular techniques for dissection, bipolar diathermy, ultrasonic dissector can be used (Harmonic®, Ethicon). To increase the intra-abdominal working space and exposure of the tumor, dissecting and transferring all the bowel to bowel bag is suggested by some authors. This technique also helps to prevent any inadvertent bowel injury, heat loss from bowel surface, and also postoperative bowel adhesions [16].

12.2.3 The Procedure

RN including Gerota's fascia, perirenal fat, and adrenal gland should be achieved en bloc (Fig. 12.1). The dissection begins with mobilization of the colon medially to expose the retroperitoneal structures. The colonic mesentery may be left attached on the tumor with preservation of marginal arcade of colonic vessels [16]. In left-sided tumors, the spleen and pancreas may also be mobilized and retracted medially for better access and exposure of the tumor. On the right side, Kocher's maneuver is helpful in exposing the inferior vena cava and renal vein. There has been no consensus regarding the extent of lateral mobilization of the tumor. The "*purists*" believe in no handling or mobilization of the kidney harboring the tumor until after the vessels have been ligated at the hilum (Fig. 12.3a). There are others who choose the middle path and tend to dissect laterally, mobilize, and retract the mass with the kidney partly out of the incision [16, 17]. But few including the principal author believe in delivering the entire tumor outside the abdominal cavity after thorough dissection and mobilization followed by ligation and transaction of renal vascular pedicle for complete en bloc excision (Fig. 12.3b) [18]. This goes against the traditional teaching of control of renal hilar vessels first. In very large or infiltrating tumors, primary ligation of the renal vessels may be in fact difficult or risky, resulting in major vascular complications such as injury to mesenteric arteries/celiac vessels/aorta/IVC, etc. [19, 20]

The ureter is identified at the pelvic brim/pelvic-ureteric junction (depending on tumor size) after mobilization of the kidney and is divided as close to the bladder as possible after division of gonadal vessels and securing all the blood supply of the ureter. In cases of extension of tumor into the ureter (botryoid WT) [21] with-

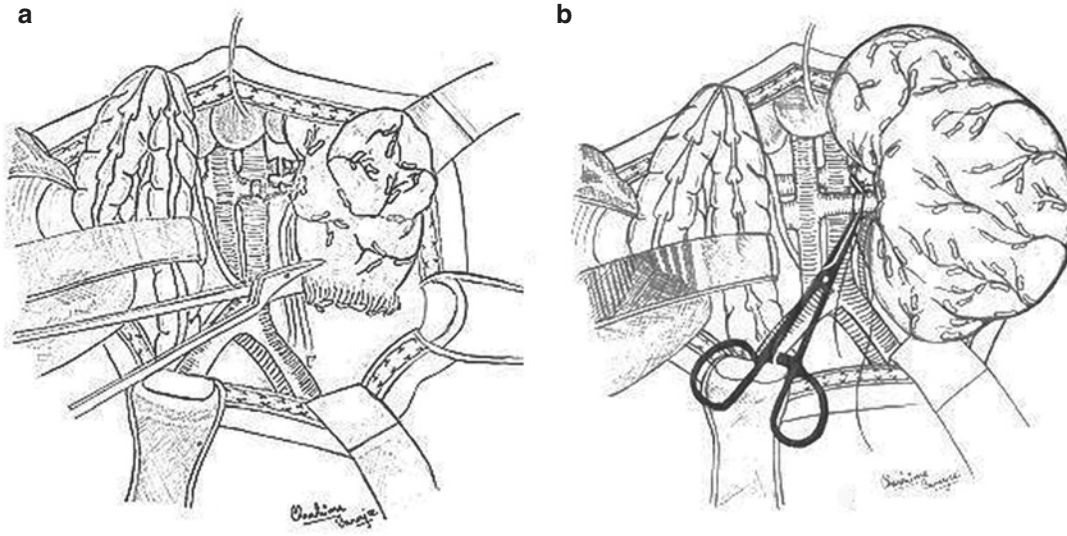

Fig. 12.3 (**a**) Traditional "pedicle first" technique and (**b**) "tumor delivery" technique, where the pedicle is divided the last

out extension into the bladder, the entire length of the ureter up to the ureteric orifice in the bladder needs to the excised taking care of IOS during dissection, ligation, and division of the ureter. All patients presenting with gross hematuria should have cystoscopy just before the surgery to rule out extension up to or beyond ureteral orifice, and in doubtful cases, a cuff of bladder should also be excised along with the ureter, using the distal end of upper divided ureter for traction to expose the renal hilum from below upward.

The *"purists"* believe that when tackling the hilum, the sequence of ligation of vessels is first artery (to avoid venous congestion and possible tumor rupture) and then renal vein. Both artery and vein should be ligated individually to avoid the probability of high output cardiac failure due to renal vessel vascular shunt in the future. Another important feature to be kept in mind is tenting of IVC during renal vein ligation, which could lead to elliptical IVC breach after release of the traction following RN. Before (double) ligating and dividing the renal vein, it is important to palpate it so as not to cut through the intravascular tumor and causing IOS. Extension of tumor in the posterior abdominal wall/diaphragm would require attentive excision and adequate repair of the muscles of these structures [22].

Very extensive and mutilating resections of surrounding organs (e.g., pancreatectomy) are not recommended [23]. Infiltrations into adjacent tissue, affected LNs, macroscopic residues, and macroscopic IOS should be detailed in the operative notes.

As dictated by intraoperative findings, the tumor bed could be prepared for future XRT by marking the site with titanium clips.

12.2.4 Tumor Thrombus in the Renal Vein and Inferior Vena Cava

Preoperative evaluation, by MRI, CT, or ultrasound scan, should state the patency of the renal veins and inferior vena cava (IVC). However, intraoperative examination of renal vein and IVC is suggested. Several surgical options exist depending on the extent of tumor thrombus cranially such as simple thrombectomy for renal vein thrombus with or without complete excision of renal vein, inferior vena cavectomy for extension in IVC below the hepatic veins, and resection without bypass or on cardiopulmonary bypass (CPB) for thrombus extending above the hepatic veins into the IVC/into the atrial chamber [24] and finally staged resection.

For vena cavotomy, the contralateral renal vein as well as IVC on both sides of the thrombus have to be looped with vascular loops before proceeding further. If the defect in the IVC is large, simple closure may cause constriction in which case autologous graft of saphenous or internal iliac vein may be required. In cases where tumor thrombus is densely adherent to the IVC wall, inferior vena cavectomy is the only option which is safe due to the development of multiple alternate collaterals [16].

Cardiopulmonary bypass will be required in the case of intra-atrial thrombus. It may also be very useful in case of a longer thrombus, extending to or above the level of the hepatic veins [16, 24]. Details of these sophisticated procedures are beyond the scope of this chapter.

12.2.5 Adrenal Gland

As per evidence, removal of adrenal gland as a routine has been challenged and rejected by some authors as the involvement of adrenal gland is rare [25, 26]. In situations wherein WT is arising from upper pole of the kidney increasing the risk of local infiltration as well as in difficult dissections wherein risk of tumor rupture increases during attempts to save the adrenal gland, adrenalectomy is advised [17]. van Waas et al. also favored adrenalectomy quoting that one adrenal gland is enough to maintain normal function and does not lead to adrenal insufficiency [27].

12.2.6 Lymph Nodes

Even when LN do not seem involved on gross examination, at least seven LNs have to be excised and sampled for histological examina-

tion; the chances of finding a positive LN increase when more than seven LNs are biopsied [28–31]. The areas of LN biopsies are paracaval supra-hilar, paracaval infra-hilar, paraaortic supra-hilar, paraaortic infra-hilar, right iliac, left iliac, and mesenteric (1 LN from each site) [32]. Appropriate labeling of site and character is crucial before sending the samples for histopathology. Unlike in RCC, radical LN dissection is not recommended for WT as there is no benefit in terms of overall survival.

12.2.7 Translocation of Ovary

The principal author believes in surgically translocating the ipsilateral ovary in girls to the contralateral side with preservation of its blood supply, lest the patient is staged III necessitating ipsilateral flank XRT.

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