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Retroperitoneal approach in a child with ganglioneuroma: points of technique and literature review

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Abstract

Background data: Ganglioneuroma is rare in children and often incidentally discovered. The transperitoneal laparoscopic approach is the surgical choice for most of the surgeons because of the familiar anatomy and the wider working spaces. However, a posterior retroperitoneal approach started to be used in pediatric population demonstrating to be safe and effective.

Case presentation: We present a case of a child with radiological finding of a right adrenal mass. Due to the increased risk of having a malignant transformation, a surgical resection was decided using a retroperitoneal approach. The adrenal mass was recognized and completely enucleated. The histopathological examination revealed a GN. Clinical course was uneventful with hospital discharge after 3 days. The outcome was evaluated by ultrasound examination performed 3–6 to 12–24 months and then annually. After 24 months, the patient is completely asymptomatic, and no adrenal masses in both sides have been detected.

Conclusions: The posterior retroperitoneoscopy permits a more direct exposure of the adrenal gland, without violating the peritoneum, and a close vision to the main vessels. We believe that this technique is a valid option for pediatric patients with small adrenal lesions.

Keywords: Ganglioneuroma, Retroperitoneal tumor, Children, Retroperitoneoscopy, Adrenal tumor, Case report

Background

Ganglioneuroma (GN) is a rare, benign, differentiated neurogenic tumor arising mainly from primordial neural crest [1] affecting the pediatric age group. The median age at diagnosis is about 7 years old [2]. It is usually asymptomatic, until they reach large size, in which case the symptoms can be due to local expansion [3, 4]. Surgery is considered the chosen treatment for suspected GNs, and prognosis is excellent after complete surgical excision [5]. To date, laparoscopic excision is considered the preferred approach [6].

Case presentation

A 7-year-old boy with history of abdominal pain and no evidence of palpable mass in abdominopelvic examination underwent pediatric surgical consultation. The patient had a free previous medical history. Laboratory tests with urine catecholamine assay were performed, and the result was normal. The sieric values of dehydroepiandrosterone sulfate (DHEAS), testosterone, 17-hydroxyprogesterone (OHPG), adrenocorticotropic hormone (ACTH), cortisol (CORT), and delta-4 androstenedione (ASD) were into the range of normality. US evaluation showed a 32 × 5 × 36 mm right adrenal

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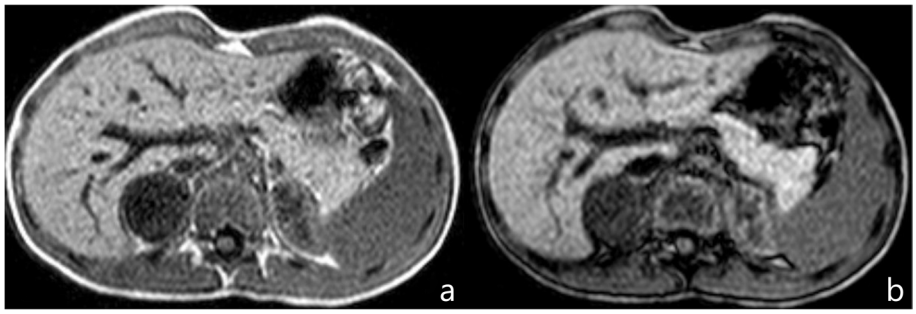

Fig. 1 **a–b** "Gradient echo in-phase (IP) and opposed-phase (OP) T1-weighted MRI axial abdominal section showing the ovalar 34 × 31 mm mass in the right adrenal gland. There is no significant loss of signal intensity in the opposed-phase (OP) images (**b**) compared to in-phase (IP) images (**a**). This behavior is related to the absence of intracellular lipids"

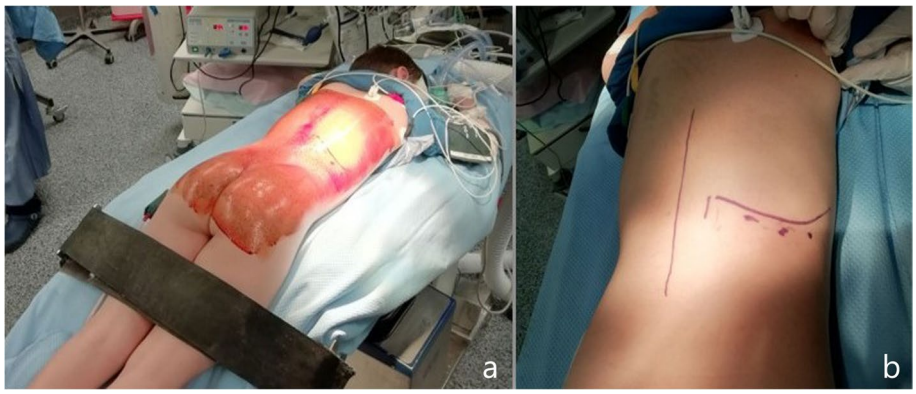

Fig. 2 **a–b** Patient in prone jackknife position (**a**). Anatomic landmarks and skin incision (**b**)

mass. The lesion was then confirmed on MRI (shown in Fig. 1a–b).

By considering the age of the patients and the suspicious of malignancy of the mass, a surgical resection was carried out. The patient was positioned prone with hip joint bended (shown in Fig. 2a). The right 12th rib and the lateral border of the right erector spinae muscle were preoperatively marked (shown in Fig. 2b). A 1.5 cm transverse incisions was made below the tip of the 12th rib. The retroperitoneal space was bluntly dissected using a finger. A small cavity was obtained digitally in order to insert a 5 mm, trocar 4 to 5 cm lateral beneath the 11th rib. A second 5 mm trocar was inserted 4 to 5 cm medially to the initial incision. Then, a blunt trocar with an inflatable balloon was placed into the transverse incision site and blocked (shown in Fig. 3). Retroperitoneoscopy was performed by a 5-mm 30° endoscope. Retroperitoneum was created by maintaining a CO2 pressure of 8 to 15 mmHg. Fatty tissue from the posterior side of the kidney was dissected to expose the posterior pole of the kidney. The superior pole of the right kidney was mobilized starting inferiorly, and the adrenal mass was recognized.

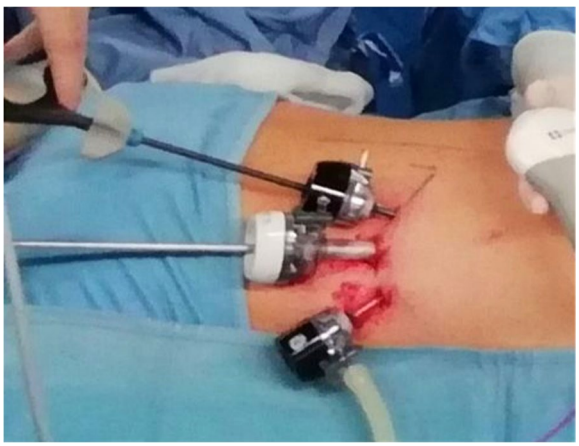

Fig. 3 Three trocars are inserted below the 12th rib

Harmonic scalpel was used to dissect the adjacent tissue around the tumor to avoid the break of the tumor capsule. Mobilization of the adrenal gland was performed by starting medially and caudally, and an endocatch bag

was used to remove the mass. Clinical course was uneventful. Patient was discharged after 3 days. US evaluation was performed 3–6 to 12 months. After 24 months, the patient is completely asymptomatic, and no adrenal masses in both sides have been detected (shown in Fig. 4). The pathological examination results for the mass of the right adrenal gland were obtained. A whitish nodular sample of 4 × 3.5 × 3 was analyzed: cut surface of yellowish white color, soft consistency, and homogeneous appearance. A neoplasm consisting of ganglion cells arranged in clusters and with swollen eosinophilic cytoplasm in a matrix represented by Schwann cells was identified. The findings were compatible with ganglioneuroma (shown in Fig. 5)

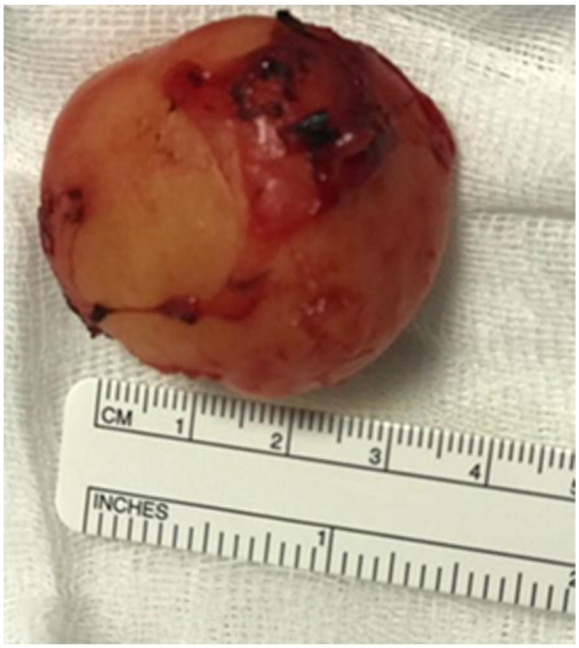

Fig. 5 A 32 × 35 encapsulated mass of firm consistency with an homogenous, solid, yellowish white cut surface, histologically confirmed to be a ganglioneuroma

Discussion

Just few papers discuss the management of GN on pediatric population [7–11]. In our patient, worried for a possible malignant behavior of the mass, having obtained parental consent, a surgical treatment was performed. Mini-invasive surgery for adrenal gland is well described in adults but less in children due to the rarity of the disease. Four approaches have been described to perform an adrenalectomy:transperitoneal, lateral retroperitoneal, posterior retroperitoneal, and transthoracic transdiaphragmatic access [12]. Walz et al. [13] firstly reported their experience with retroperitoneoscopy in children and adolescents demonstrating feasibility and safety of this procedure. Compared with the laparoscopic approach, retroperitoneoscopy offers a direct access to the gland without mobilizing abdominal organs [7]. Moreover, the anatomical features of pediatric population such as the lower muscle thickness and a lower quantity of adipose tissue make a retroperitoneal approach more feasible despite the lack of space [14]. Moreover, Walz et al. considered a mass larger than 7–8 cm, more likely to be malignant and difficult to manipulate without risk of capsular rupture, a contraindication for retroperitoneal approach [9]. Some authors prefer a retroperitoneoscopic approach for adrenal tumor on the left side, due to the fact that on the right side, the gland arteries cross the vena cava posteriorly [15]. Retroperitoneoscopy can be performed in lateral or in prone position. While lateral approach is well described in literature, less is known about the posterior retroperitoneoscopic adrenalectomy. In case of bilateral lesions, a posterior approach permits

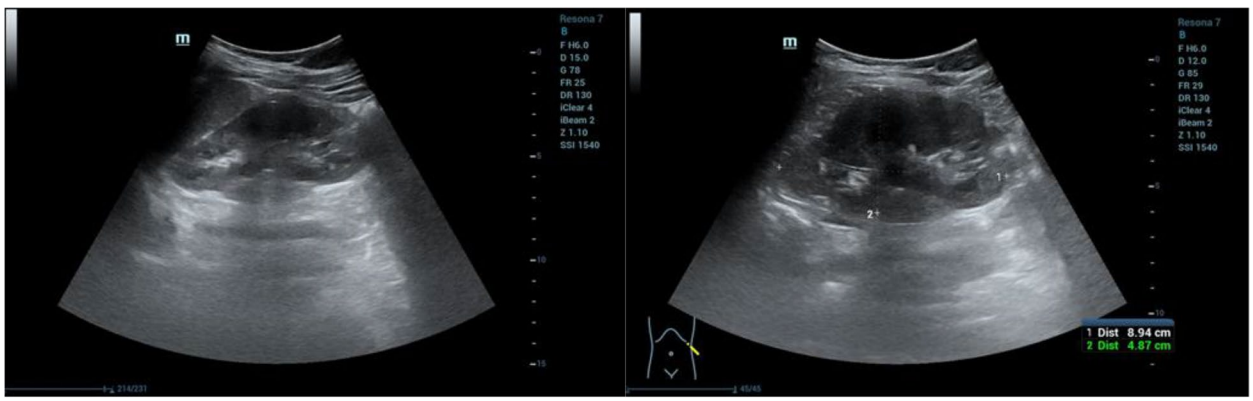

Fig. 4 Ultrasound evaluation showing no mass recurrences

Table 1 Posterior retroperitoneoscopic approach in adrenal surgery. Literature review

	Study period	*N°* patients	Median age
Walz et al. (1996) [10]	1994–1995	27	10–75 years
Walz et al. (1997) [16]	1994–1997	67	10–78 years
Walz et al. (2006) [9]	1994–2006	520	10–83 years
Walz et al. (2018) [8]	2001–2016	35	7–19 years
Vrielink et al. (2017) [7]	2008–2013	57	53 years
Lee et al. (2019) [11]	2012–2017	20	4 years
Benson et al. (2019) [17]	2019	1	10 years
Our case	2018	1	7 years

bilateral access to adrenal glands without moving the patient [7, 9]. In our case, we prefer a posterior retroperitoneoscopic approach in a prone jackknife position using a standard operating table according to the mass diameter and the preference of the surgeon. We reviewed the literature regarding the retroperitoneoscopic approach described by Waltz, the results of which are reported in Table 1 (shown in Table 1).

A complete enucleation of the mass was performed, allowing a residual adrenal gland's cortical function. Since GN can tightly adhere to, or encase major vascular structures [18], an incomplete resection is often performed. In this regard, De Carolis et al. [19] reported that an incomplete resection may be sufficient to avoid recurrence if tumor residual is smaller than 2 cm. Concerning the risk of recurrence, Spinelli et al. [20] did not experience any recurrences during follow-up period in surgically treated pediatric patients. Due to the benign nature of GN, radiotherapy or adjuvant chemotherapy is not indicated [1, 19, 21, 22]. Even if follow-up protocol after complete excision is not clearly reported in literature, a regular follow-up is suggested to exclude local recurrences [1], especially when resection was not complete [11].

Conclusion

GN is a rare lesion in pediatric patients, often incidentally discovered. Although a transperitoneal laparoscopic approach is preferred by most of the surgeons because of the familiar anatomy and the wider working spaces, posterior retroperitoneoscopic approach has gained international popularity in the past decade and proven to be reliable and safe. Main advantages of this approach are the direct exposure of the gland without the need for violating peritoneum and a close vision to the main vessels, minimizing the risks of injuries. Especially after sparing surgery, a long-term follow-up is suggested.

Abbreviations

GN: Ganglioneuroma; DHEAS: Dehydroepiandrosterone sulfate; OHPG: Testosterone, 17-hydroxyprogesterone; ACTH: Adrenocorticotropic hormone; CORT: Cortisol; ASD: Delta-4 androstenedione; MRI: Magnetic resonance imaging; US: Ultrasound.

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Authors' contributions

PI designed the study. FP and RT contributed to the final version of the manuscript. GD served as scientific advisors. CR critically reviewed the study proposal. All authors discussed the results and contributed to the final manuscript. The authors read and approved the final manuscript.

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Availability of data and materials

Available upon request.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Informed written consent was obtained from the patient for publication of this report and any accompanying images.

Competing interests

The authors declare that they have no competing interests.

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