Case Reports in Oncology

Case Rep Oncol 2023;16:1041–1047
DOI: 10.1159/000533829
Received: May 23, 2023
Accepted: August 24, 2023
Published online: September 26, 2023

Case Report

Large Retroperitoneal Teratoma Presenting with Unilateral Hydronephrosis in an Infant: A Case Report and Review of the Literature

Negar Safaei[a] Hooman Kamran[a] Mohammad Hossein Anbardar[b] Mohammadreza Bordbar[c] Omid Reza Zekavat[c] Mehdi Forooghi[d]

[a]Student Research Committee, Shiraz University of Medical Sciences, Shiraz, Iran; [b]Department of Pathology, Shiraz University of Medical Sciences, Shiraz, Iran; [c]Hematology Research Center, Shiraz University of Medical Sciences, Shiraz, Iran; [d]Department of Pediatric Surgery, Shiraz University of Medical Sciences, Shiraz, Iran

Keywords
Hydronephrosis · Immature teratoma · Obstructive uropathy · Retroperitoneum space · Teratoma

Abstract
Teratoma is a type of germ cell tumor layer that appears in the gonadal, sacrococcygeal, mediastinal, and retroperitoneal regions. Primary retroperitoneal teratoma is rare and asymptomatic but can present with symptoms due to a mass effect on neighboring organs. These tumors have to be considered in the differential diagnosis of a mass in the abdominal cavity of children to distinguish between Wilms' tumor, neuroblastoma, and other intra-abdominal lesions. We presented an infant boy with protrusion of the left upper quadrant of the abdomen and a palpable abdominal mass that had progressively enlarged. An abdominal computed tomography scan revealed a large retroperitoneal cystic, solid mass on the left side of the abdominal cavity, causing pressure on the left ureter. Also, hydronephrosis of the left kidney was seen with a decreased enhancement of the left kidney due to obstruction uropathy. The mass was suspicious on imaging for a retroperitoneal teratoma. The patient underwent laparotomy, and excision of the huge retroperitoneal mass was done. The final diagnosis was an immature teratoma grade 3, and the patient was discharged in good condition. Retroperitoneal teratomas are rare tumors in infants. These tumors would be an incident diagnosis or diagnosed by the mass effect of giant tumors on other organs. They must be considered in the differential diagnosis of intra-abdominal tumors in children. Hydronephrosis and obstructive uropathy can be rare consequences of the mass effects of these tumors.

Correspondence to:
Mehdi Forooghi, dr.forooghi@yahoo.com

Case Rep Oncol 2023;16:1041–1047
DOI: 10.1159/000533829

Introduction

Teratoma is a type of germ cell tumor containing an embryonic germ cell layer that appears in the gonadal and sacrococcygeal, mediastinal, intracranial, cervical, and retroperitoneal regions [1]. Primary retroperitoneal teratoma is rare, accounting for 4% of all primary teratomas, and is usually asymptomatic [2, 3]. However, in some cases, it can present with different symptoms based on the mass pressure effect on neighboring organs. Because of this mass effect of tumors in patients with teratoma, there is a risk of urologic comorbidities, such as bladder dysfunction or hydronephrosis [4]. These types of germ cell tumors may be cystic or solid, mature or immature, or mixed germ cell tumors [5, 6].

Retroperitoneal cystic immature teratoma rarely occurs in children (less than one percent of all childhood tumors) and is histopathologically composed of mainly immature neural tissue [7]. These tumors be an issue in the differential diagnosis of a mass in the abdominal cavity of children to distinguish between Wilms' tumor, neuroblastoma, and other intra-abdominal lesions [8]. Here, we will discuss a rare case of huge retroperitoneal immature teratoma in a 6-month-old infant that presented with abdominal mass and hydronephrosis. The CARE Checklist has been completed by the authors for this case report, attached as supplementary material, available online at https://doi.org/10.1159/000533829.

Case Report

Chief Complaint and Admission

The patient was a 6-month and 29-day old boy without any significant past medical disease who presented with irritation and protrusion of the left upper quadrant of the abdomen. Also, an abdominal mass was palpated by his mother, which had progressively become enlarged. So, the patient was referred to a pediatrician, and abdominopelvic ultrasonography was requested. Abdominopelvic ultrasonography showed hydronephrosis of the left kidney due to the pressure effect of a large septate cystic lesion (91 × 72 mm) without a solid part on the left side of the abdomen. He was referred to our hospital for further evaluation.

Physical Examination, Paraclinical Tests, and Imaging

He was afebrile with stable vital signs during the physical examination. In the abdominal examination, a left-sided abdominal mass was palpated. Laboratory data showed elevated C-reactive protein and erythrocyte sedimentation rate (C-reactive protein = 67 mg/dL, erythrocyte sedimentation rate = 21 mm/h) and mild leukocytosis (white blood cells = 13,000 per μL). The kidney function tests, such as blood urea nitrogen and creatinine, were in the reference range, and the urine analysis showed alkaline urine with a specific gravity of 1.005.

An abdominal computed tomography (CT) scan revealed a large cystic, solid mass (90 × 88 × 118 mm) on the left side of the abdominal cavity arising from the retroperitoneum causing anterior deviation of the pancreas and lateral deviation of the left kidney. The mass caused pressure on the left ureter, and moderate hydronephrosis of the left kidney was seen with a decreased enhancement of the left kidney due to obstruction uropathy. Also, internal calcification and fat density in the lesion were seen. The mass was reported to be in favor of retroperitoneal teratoma. Besides, the left adrenal gland could not be delineated because of the pressure effect of the mentioned mass. The deviation of the abdominal aorta to the right side was also present due to the mass (shown in Fig. 1).

Case Rep Oncol 2023;16:1041–1047
DOI: 10.1159/000533829

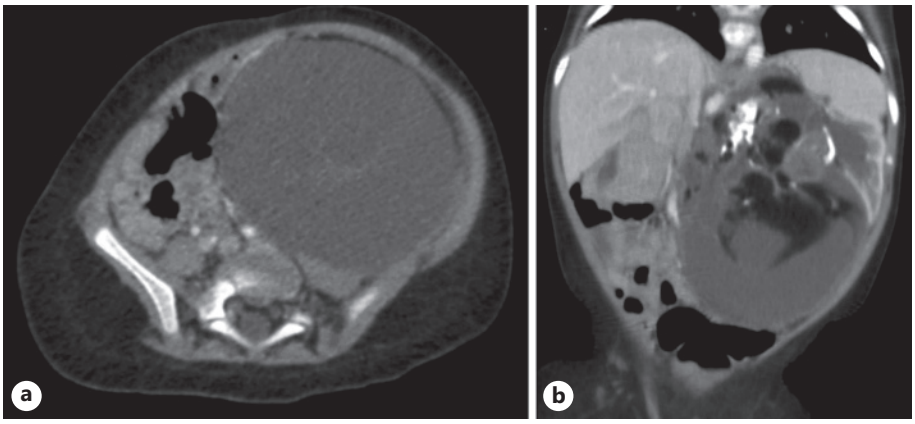

Fig. 1. Preoperative computed tomography (CT) scan of a giant retroperitoneal teratoma; axial view (**a**) and coronal view (**b**).

Surgical Procedure

The patient underwent exploratory laparotomy, and excision of the huge retroperitoneal mass was done. Intraoperative findings showed a solid cystic mass on the left side of the abdominal cavity from the upper to lower quadrant with transposition of the small bowel, large bowel, and major vessels to the right side. However, the left ureter and left kidney were transposed to the posterolateral, leading to hydronephrosis (shown in Fig. 2).

Pathological Findings

An oval mass with a size of 12 × 8 × 7.5 cm was received in the macroscopic examination. The external surface was bosselated and creamy pink in color. Cut sections showed multicystic areas filled with clear fluid, mucinous material, and some nodularity and bony areas (shown in Fig. 3). The final diagnosis was an immature teratoma grade 3 (20% of teratoma composed of immature elements).

Post-Operation and Follow-Up

The patient was discharged in good condition 6 days after the surgery with suspensions of cefixime and metronidazole. The patient was visited at our clinic postoperatively 1 week later, with no signs and symptoms presented. Also, the patient was referred to a pediatric hematologist-oncologist for possible chemotherapy sessions. The pediatric hematologist-oncologist evaluated the patient following the operation. After performing chest and abdominopelvic CT scans, whole-body bone scans, and laboratory factors, including alpha-fetoprotein and beta-human chorionic gonadotropin, and consultation with the patient's parents, it was decided that the patient could be followed with routine imaging and laboratory data (with no need for chemotherapy) due to no sign of metastasis and normal imaging and laboratory factors. The patient had been visited in an outpatient setting every 4 months in the first year of post-operation, and, currently, he is being followed every 6 months as we are in the second year of the follow-up (current follow-up: 1.5 years). The patient is well with no signs and symptoms or metastasis/recurrence.

Discussion

The most common type of germ cell tumor is teratoma, divided into three types: mature, immature (or malignant type), and monodermal. The mature type of teratoma is the most common type, which is a benign solid or cystic tumor [9]. While most teratomas are present in the gonadal

Case Rep Oncol 2023;16:1041–1047
DOI: 10.1159/000533829

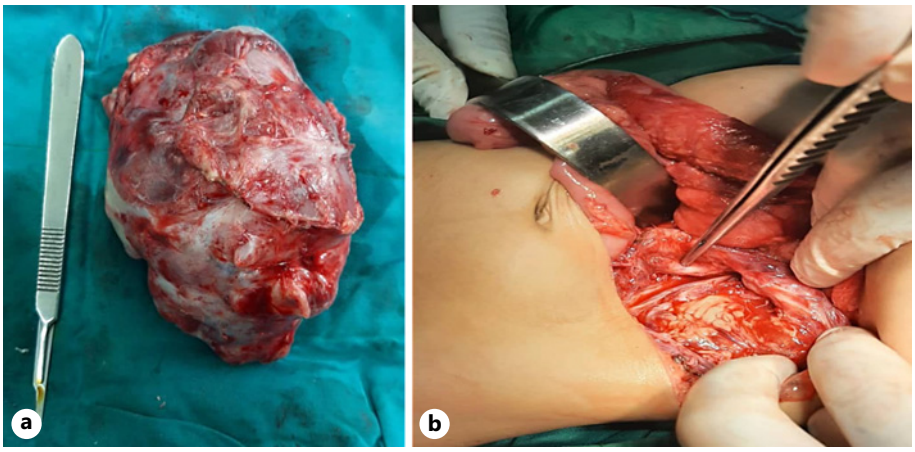

Fig. 2. a Gross appearance of the excised teratoma. **b** Intraoperative image showing dilated ureter due to pressure effect of the huge retroperitoneal teratoma.

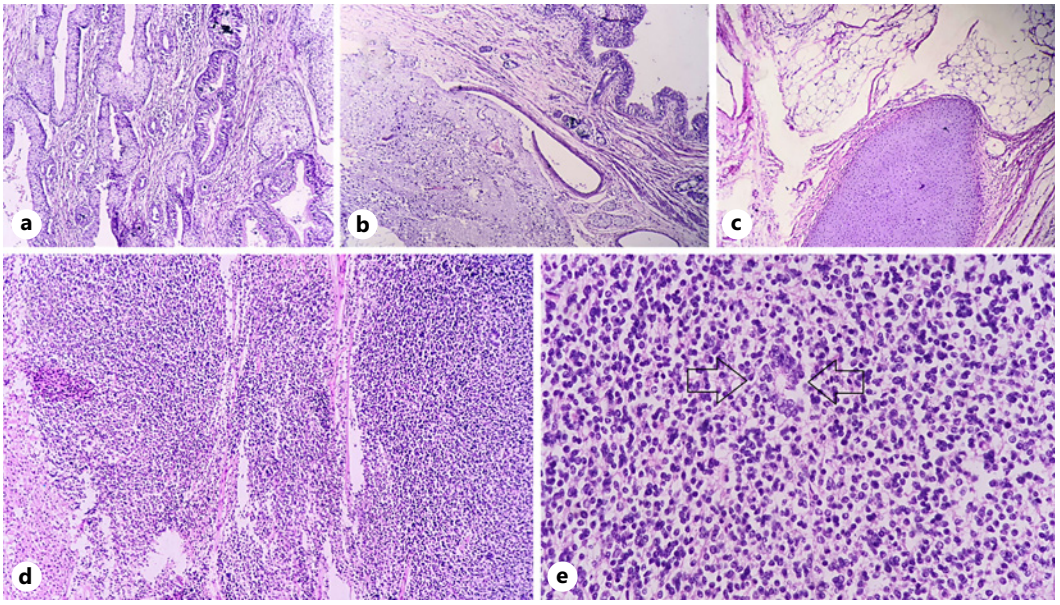

Fig. 3. a–c Microscopic sections show mature elements. **a** Squamous and respiratory epithelium. **b** Neurogenic mesenchymal tissue and respiratory epithelium. **c** Mature cartilage and adipose tissue (hematoxylin and eosin, ×40). **d**, **e** Microscopic sections show immature neuronal component. **a** Low magnification of primitive neuronal areas (hematoxylin and eosin, ×100). **b** Primitive cells with scant cytoplasm, hyperchromatic nuclei and rosette formation (arrow) (hematoxylin and eosin, ×400).

region, the extragonadal teratomas are in the sacrococcygeal area, mediastinum, intracranial, and retroperitoneal area. Retroperitoneal teratomas, after Wilms tumor and neuroblastoma, are the most common retroperitoneal tumors in children accounting for about 3.4–4% of all germ cell tumors in children. About 4–6% of all teratomas are primary retroperitoneal teratomas, usually occurring in the suprarenal area and are more commonly present on the left side [10]. One of the problems of these tumors is differentiating them from Wilms tumors and neuroblastomas or other intra-abdominal tumors [8]. Our case describes a giant immature retroperitoneal teratoma, which is rare in this size in children under 1 year of age. Besides, this tumor was large enough to lead to a mass effect on the left ureter in a way that the patient developed hydronephrosis.

Case Rep Oncol 2023;16:1041–1047
DOI: 10.1159/000533829

Table 1. Review of four previously reported pediatric cases with retroperitoneal and renal teratomas causing hydronephrosis

Author (year)	Age, sex	Tumor size	Signs and symptoms	Imaging	Pathology	Treatment	Follow-up
Wolski et al. [15] (1981)	3 y/o, male	16 × 8 cm	N/A	Urography: tumor displaced the left kidney and induced hydronephrosis	Teratoma adultum	Tumor resection along with the left kidney	No problem after 1 year
Kobayashi et al. [12] (2014)	6 m/o, female	A round 7.5 cm tumor	A palpable mass in the upper abdomen	CT: a 5.5 cm mass with calcification adjacent to the right kidney and right hydronephrosis	Mature cystic teratoma	En bloc resection	No recurrence after 1 year
Saini et al. [13] (2020)	4 m/o, male	25 × 18 × 19 cm mass	Gradual abdominal distension	FDG-PET CT: displaced right kidney with hydronephrosis, thinned cortical parenchyma, and delayed contrast uptake and excretion	Mature teratoma	Mass excision, en bloc nephrectomy, and infrarenal caval resection without reconstruction, limited diaphragmatic and psoas muscle resection, and in continuity removal of the sinus tract	Tumor-free after 4 months
Rampersad et al. [14] (2021)	4 m/o, female	15 × 8 cm tumor	Painless abdominal distension	Ultrasound: left cystic structure, dilated left pelvicalyceal system, and left hydronephrosis	Mature cystic teratoma of kidney	Complete resection (left nephrectomy)	Disease-free over 7 years
Current case	6 m/o, male	12 × 8 × 7.5 cm	Palpable mass and abdominal distension	CT: pressure on the left ureter and moderate left hydronephrosis	Immature teratoma	Complete excision of tumor	Symptom-free after 1.5 years; referred to pediatric hematologist-oncologist for further follow-up

CT, computed tomography; FDG-PET, 18-fluorodeoxyglucose-positron emission tomography; N/A, not available; m/o, months old; y/o, years old.

Case Rep Oncol 2023;16:1041–1047
DOI: 10.1159/000533829

Teratoma in children usually has no specific characteristic signs or symptoms. There are different clinical presentations based on the size and location of the tumor, but their main presentation is abdominal distension or a palpable abdominal mass [11]. To the best of our literature search, there are only a few cases of retroperitoneal teratoma in children, large enough to cause hydronephrosis by the pressure effect of the mass [12–15] (Table 1). In our case, the tumor had a pressure effect on the internal organs and caused a lateral deviation of the left kidney and pressure on the left ureter, so the patient developed moderate left hydronephrosis. Also, the enhancement of the left kidney decreased due to obstruction uropathy of tumor effect. Of note, while the previous cases of retroperitoneal teratoma causing hydronephrosis were mostly mature teratomas (Table 1), our case had an immature teratoma grade 3.

Wilms and neuroblastoma are the most important differential diagnosis of retroperitoneal teratoma. Also, they can be mistaken for ovarian and adrenal tumors, renal cysts, or sarcomas [16]. However, most retroperitoneal teratomas have typical radiological features. CT and magnetic resonance imaging are reliable modalities that show the tumor and vessels well. CT is the most reliable imaging modality for diagnosing and evaluating retroperitoneal teratomas. A large complex mass with well-defined cystic components, fat, and areas of calcification represents the typical radiologic CT appearance of a teratoma [10]. In our case, before further evaluation and because of hydronephrosis reported in the ultrasonography, we suspected Wilms tumor and performed a CT scan, which plays an important role in diagnosing retroperitoneal teratoma. By CT scan and histopathologic diagnosis after resection, the definite diagnosis of teratoma was taken, and an immature teratoma grade 3 was diagnosed in our case.

In conclusion, retroperitoneal teratomas are rare tumors in infants that are generally asymptomatic and can be found incidentally or due to the mass effect of the huge tumors on the other organs. However, they should be considered as a differential diagnosis of intra-abdominal tumors in children. Hydronephrosis and obstructive uropathy can be rare consequences of the mass effect of teratomas resulting from very large-sized masses. Our case highlights two important issues.

1. Although immature retroperitoneal teratoma under 1 year of age is rare, it should be considered as a differential diagnosis of retroperitoneal masses in this age group.
2. This mass can be large enough to cause obstructive uropathy and hydronephrosis.

Statement of Ethics

This retrospective review of patient data did not require ethical approval in accordance with local/national guidelines. Written informed consent was obtained from the patient's parents for publication of this case report and any accompanying images. A copy of the written consent is available for review by the editor of this journal.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Funding Sources

The study was not funded.

Author Contributions

M.F. performed the surgery, supervised the work, and contributed to data gathering. N.S. drafted the manuscript and contributed to data gathering. H.K. contributed to data gathering and revising the manuscript. M.H.A. provided the pathology images. O.R.Z. and M.B. contributed to revising the manuscript. All authors have read and approved the final version of the work.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

References

1 Yuksel Y, Ozen O, Kuru A. Retroperitoneal teratoma with secondary infection. J Coll Physicians Surg Pak. 2019;29(8):778–9.
2 Sasi W, Ricchetti GA, Parvanta L, Carpenter R. Giant mature primary retroperitoneal teratoma in a young adult: report of a rare case and literature review. Case Rep Surg. 2014;2014:930538.
3 Nguyen CT, Kratovil T, Edwards MJ. Retroperitoneal teratoma presenting as an abscess in childhood. J Pediatr Surg. 2007;42(11):E21–3.
4 Cost NG, Geller JI, Le LD, Crombleholme TM, Keswani SG, Lim FY, et al. Urologic co-morbidities associated with sacrococcygeal teratoma and a rational plan for urologic surveillance. Pediatr Blood Cancer. 2013;60(10):1626–9.
5 Polsky MS, Shackelford GD, Weber CH Jr, Ball TP Jr. Retroperitoneal teratoma. Urology. 1976;8(6):618–21.
6 Rathod J, Patel S, Barot KS, Naik SH, Bhatt R, Chotaliya J. Massive primary retroperitoneal immature teratoma in an adult: a case report. Int J Surg Case Rep. 2021;81:105775.
7 Li J, Gong P, Liu F, Sun P, Wu C. Retroperitoneal cystic immature teratoma: a case report. Oncol Lett. 2015;10(2):1023–5.
8 Luo CC, Huang CS, Chu SM, Chao HC, Yang CP, Hsueh C. Retroperitoneal teratomas in infancy and childhood. Pediatr Surg Int. 2005;21(7):536–40.
9 Mathur P, Lopez-Viego MA, Howell M. Giant primary retroperitoneal teratoma in an adult: a case report. Case Rep Med. 2010;2010:650424.
10 Kawano T, Sugita K, Kedoin C, Nagano A, Matsui M, Murakami M, et al. Retroperitoneal teratomas in children: a single institution experience. Surg Today. 2022;52(1):144–50.
11 Alwakid IH, Madni M, Gupta RK, Seada LS, Alfayez AA, A tallaa AT. Giant mesenteric teratoma in a 6 month-old. J Pediatr Surg Case Rep. 2017;27:56–60.
12 Kobayashi T, Sakemi Y, Yamashita H. Increased incidence of retroperitoneal teratomas and decreased incidence of sacrococcygeal teratomas in infants with Down syndrome. Pediatr Blood Cancer. 2014;61(2):363–5.
13 Saini S, Kazi M, Qureshi SS. Miserable 9 Years of life: a misdiagnosed and mismanaged case of large retroperitoneal teratoma. Indian J Surg Oncol. 2020;11(Suppl 2):253–6.
14 Rampersad B, Raghunanan B. Renal teratoma–A rare cause of hydronephrosis. J Pediatr Surg Case Rep. 2021;69:101800.
15 Wolski Z, Jasiński Z. Retroperitoneal teratoma. Int Urol Nephrol. 1981;13(2):137–40.
16 Gatcombe HG, Assikis V, Kooby D, Johnstone PA. Primary retroperitoneal teratomas: a review of the literature. J Surg Oncol. 2004;86(2):107–13.