www.jpnim.com Open Access eISSN: 2281-0692
Journal of Pediatric and Neonatal Individualized Medicine 2022;11(1):e110136
doi: 10.7363/110136
Received: 2021 Oct 04; revised: 2021 Nov 13; accepted: 2021 Nov 26; published online: 2022 Apr 20

Case report

Congenital retroperitoneal immature teratoma in a male neonate: a case report

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Abstract

Introduction: Sacrococcygeal teratomas are the most frequent tumors in neonates. However, a teratoma in the retroperitoneal region is quite rare.

Case: A 39-week-old baby with a cyanotic congenital heart disease was hospitalized after birth, and an intraabdominal mass was detected with an abdominal ultrasonographic evaluation, incidentally. Surgical approach was made on the 24th postnatal day, and a large mass containing solid and cystic components was completely excised from the retroperitoneal region. Pathological assessment revealed that the mass was a grade 2 immature teratoma, including neuroepithelial tissue.

Conclusion: Retroperitoneum contains main vessels, nerves, and vital organs; excision of tumors in the retroperitoneum is crucial, especially in neonates. Because of the possibility of malignant transformation, the correct timing of the surgery, meticulous excision and careful pathological evaluation should be done.

Keywords

Case report, neonate, surgery, teratoma, retroperitoneum, pathology.

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How to cite

Karaman I, Kara YA, Balci O, Ozyazici A, Keles B, Tasci-Yildiz Y, Karaman A. Congenital retroperitoneal immature teratoma in a male neonate: a case report. J Pediatr Neonat Individual Med. 2022;11(1):e110136. doi: 10.7363/110136.

Introduction

The most common congenital tumors in the neonatal period are teratomas and tend to be located in the sacrococcygeal region. Primary retroperitoneal teratomas account for 1-11% of tumors in the retroperitoneal region [1], and only a few of them are detected in newborns [2]. We report the case of a large congenital retroperitoneal immature teratoma in a newborn with cyanotic congenital heart disease.

Case report

A 39-week-old baby weighing 3,350 grams at birth was hospitalized because of cyanotic congenital heart disease. Ebstein's anomaly and atrial septal defect were detected in the patient. After detecting an abdominal mass on ultrasonography, abdominal computed tomography (CT) was performed. Thereafter, the patient was referred to Pediatric Surgery. The mass was not palpable with physical examination. CT revealed a 47 × 40 × 31 mm mass containing calcification with solid components in the abdominal midline, from the level of the superior mesenteric artery to the left of the midline (**Fig. 1**). Upper abdominal magnetic resonance imaging (MRI) showed that the mass was located in the anterior region of the abdominal aorta, between the celiac trunk and superior mesenteric artery origins. It was located behind the superior mesenteric vein, splenic artery, splenic vein, and pancreas and in front of the aorta, renal vessels, and superior mesenteric artery (**Fig. 2**). Laboratory studies revealed that the alfa-fetoprotein (AFP) (7,809.4 ng/ml; normal: 1.6-16,456 ng/ml) and beta-human chorionic gonadotropin (ß-hCG) (< 2 mIU/ml; normal: 0.5-2.5 mIU/ml) levels were normal; however, the neuron-specific enolase (NSE) (36.65 ng/ml; normal: 13.9-22.1 ng/ml) and vanillylmandelic acid (VMA) (33.1 mg/gCrea; normal: < 18.8 mg/gCrea) levels were higher than the normal range. Surgical resection of the mass was

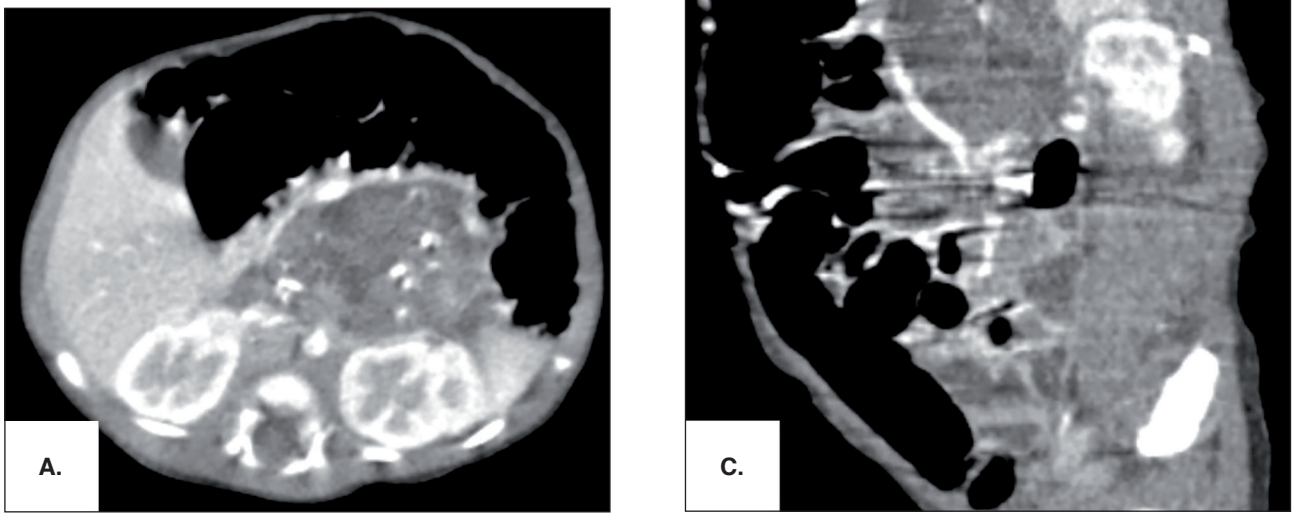

Figure 1. Abdominal computed tomography (CT) shows a 47 x 40 x 31 mm sized solid component and calcification containing mass, starting from the superior mesenteric artery level at the midline of the abdomen and extending to the left side (**A.** axial, **B.** coronal and **C.** sagittal sections).

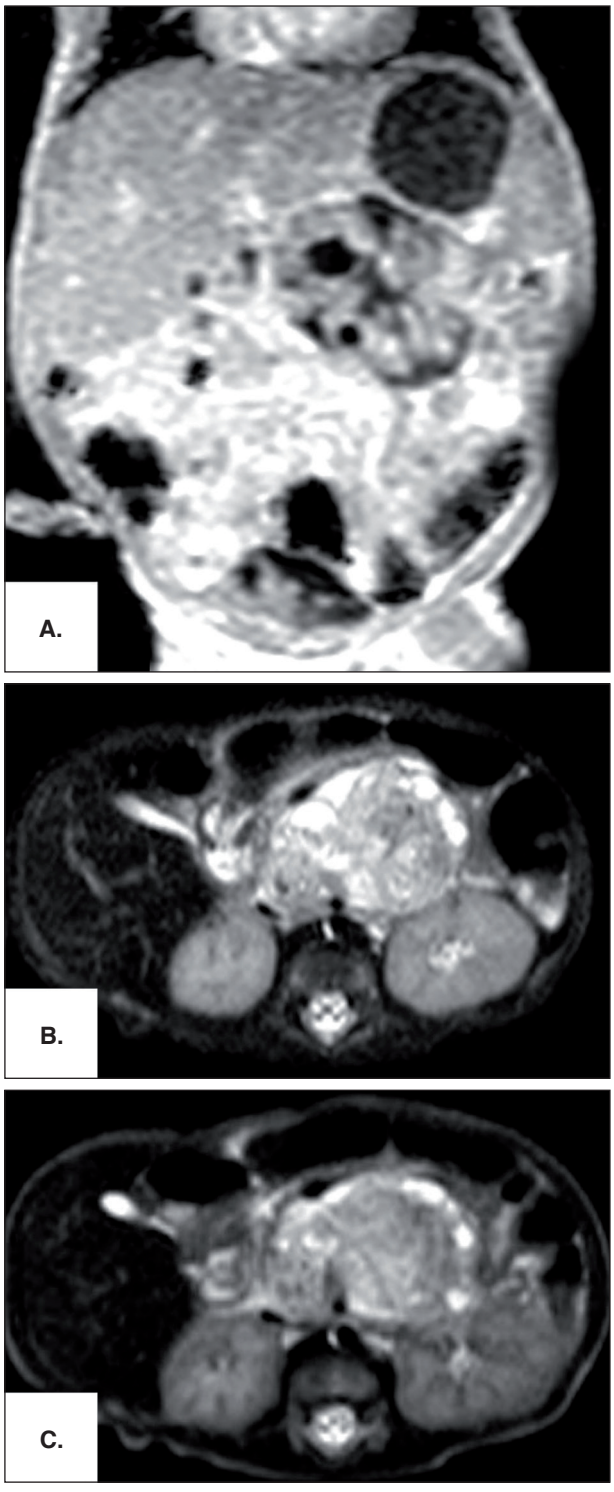

Figure 2. Abdominal magnetic resonance imaging (MRI). It is shown that the mass is located anterior of the abdominal aorta, renal arteries and superior mesenteric artery; between roots of the celiac artery and superior mesenteric artery; behind the superior mesenteric vein, splenic artery and pancreas (**A.** coronal, **B.** and **C.** axial sections).

planned with the diagnosis of retroperitoneal mass. The patient underwent surgery when he was 24 days old, and a 7.5 × 7 cm mass containing solid and cystic components was completely excised from the retroperitoneal region (**Fig. 3**). The mass

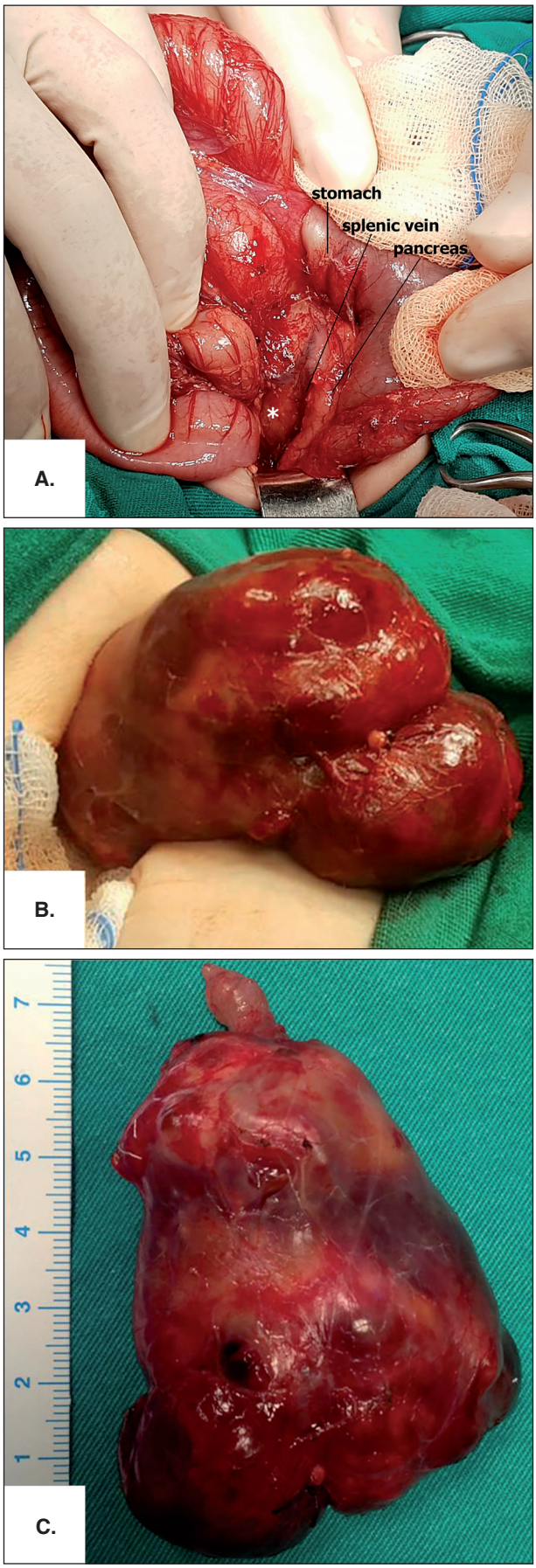

Figure 3. A. Intraoperative view (white asterisk), **B.** excision of the mass and **C.** macroscopic view of the retroperitoneal mass.

extended from the abdominal midline to the left, further extending behind the pancreas and spleen, from the back of the stomach to the diaphragm, down to the lower region of the left kidney. The patient had no problems in the postoperative period. He started being fed orally on the 3rd day and was discharged on the 7th day postoperatively. Pathological assessment revealed that the mass was a grade 2 immature teratoma, including neuroepithelial tissue (**Fig. 4**). Oncological consultation revealed no need for chemotherapy or radiotherapy in the postoperative period. The patient died at the age of 4 months due to ventricular tachycardia attack resulting from the existing cardiac anomaly.

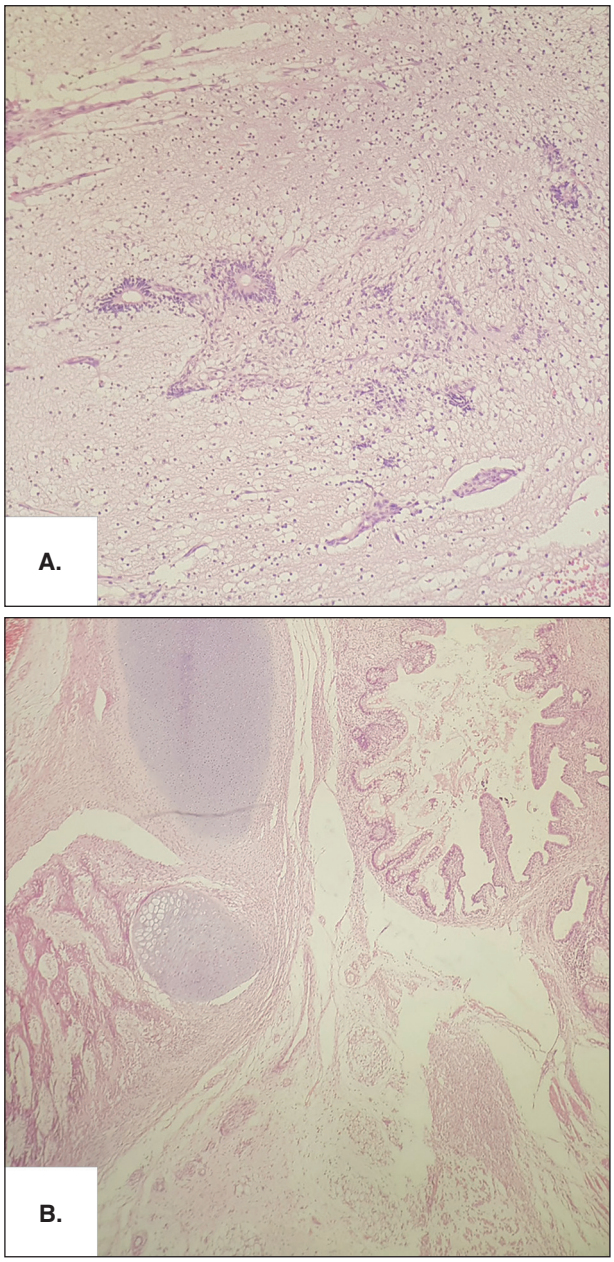

Figure 4. Histopathological appearance of grade 2 immature teratoma. **A.** Immature neuroepithelial tissue inside of immature glial tissue (H&Ex40). **B.** Intestinal, cartilaginous and osseous tissues (H&Ex40).

Discussion

Teratomas are the most common congenital tumors; they are most commonly located in the sacrococcygeal region, followed by the cranial and cervical regions, in this order. A study involving 534 fetal and neonatal teratomas reported that only 3% of teratomas are located in the tongue, liver, retroperitoneum, eye, mesentery, ileum, testicle, vulva, and anorectum [2]. Making the differential diagnosis with other retroperitoneal tumors, including Wilms' tumor, neuroblastoma, rhabdomyosarcoma, and non-Hodgkin lymphoma [3], is most challenging.

Teratomas can be benign or malignant. Benign teratomas are mostly asymptomatic and are incidentally detected, as in the present case. However, they can cause obstructive symptoms, such as nausea and vomiting, when they get bigger. Malignant teratomas grow rapidly and are mostly detected at a later stage. Patients usually experience abdominal pain and loss of weight, in addition to the presence of an abdominal mass [1].

When the diagnosis is made, surgical resection should be performed as soon as possible. Complete surgical excision is the preferred therapy for retroperitoneal teratomas [4]. However, tumor excision from the retroperitoneum is very difficult and crucial, especially in neonates. Sometimes it may be necessary to remove some of the adjacent organs such as kidney and intestine to remove the tumor completely [5, 6]. Recurrent tumors can be observed as a result of intraoperative tumor rupture [7]. To avoid such complications, after a very careful surgical excision of the tumor, a meticulous histopathological examination should be performed in order to detect immature tissues and yolk sac tissues. Postoperatively, patients should be followed closely and their serum AFP levels should be monitored [8]. Postoperative adjuvant chemotherapy may be required in cases where complete surgical excision is not possible or tumor spread during surgery [7]. Oncological consultation should be provided before and after surgery to determine the need and timing of chemotherapy or radiotherapy [8].

In conclusion, congenital retroperitoneal immature teratoma is one of the rare and surgically challenging tumors of the newborn. When complete surgical excision of the tumor is possible, close follow-up of the patient with the Oncology

Department is often sufficient without the need for any other treatment. Because of the possibility of the presence of malignant tissues, microscopic evaluation should be meticulously performed and the patient's family should be informed about the early and late complications that could arise in the follow-up period.

Informed consent

The informed consent in compliance with the Helsinki Declaration was received from the parents whose child was presented in this study.

Declaration of interest

The Authors declare that there is no conflict of interest. This case received no financial support.

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