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Retroperitoneal Teratoma in Infants: About a Case in the Medical Imaging Department of the Mother and Child Hospital Center "Luxembourg"

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Abstract

Introduction: Retroperitoneal teratoma is a rare congenital tumour, representing only 1% to 10% of primary retroperitoneal tumors in children, far behind ovarian and testicular locations. There is a clear female predominance with a sex ratio of 3.4/1. The purpose of this study is to review the clinical manifestations, the mode of revelation of retroperitoneal teratoma, the diagnostic means and the anatomopathologic aspect of retroperitoneal teratoma, then to show the contribution and the limits of the various imaging examinations: ultrasound, CT and MRI in the diagnosis of retroperitoneal teratoma. **Observation:** We report the case of a mature retroperitoneal teratoma discovered in a 10-month-old male infant. The diagnosis evoked by ultrasound and abdominal CT was confirmed by histological study of the excised specimen. The postoperative evolution after 3 months was favorable in particular with no signs of recurrence. **Conclusion:** Retroperitoneal teratoma is a rare congenital tumour. The complete imaging assessment including ultrasound and computed tomography is necessary preoperatively in order to make the diagnosis and to clearly determine the relationship with the various organs. The anatomopathological study confirms the diagnosis of retroperitoneal teratoma. Tumor excision must be radical to avoid recurrence, which readily occurs in a malignant form. Even if the tumor appears benign, postoperative monitoring based on clinical, biological and radiological examination is essential.

Keywords

Tumours, Mature Retroperitoneal Teratoma, Child

1. Introduction

Retroperitoneal teratoma is a rare congenital tumour. Its incidence is 0.3% to 3% of all tumors and 1% to 10% of primary retroperitoneal tumors in children, far behind the ovarian and testicular locations [1].

This tumor is independent of the kidney, adrenal, excretory tract, large vessels and viscera.

The diagnosis of retroperitoneal teratoma is delayed because of its retroperitoneal site and its clinical latency. The clinical signs are the result of compression or invasion of neighboring organs.

Computed tomography is the most frequently used diagnostic imaging examination for positive diagnosis [2], characterization, preoperative assessment and follow-up of retroperitoneal tumours.

Definitive diagnosis is based on pathological examination of the surgical specimen or percutaneous biopsy.

Surgical excision is the only effective treatment, sometimes potentiated by other complementary treatments such as radiotherapy or chemotherapy.

The prognosis of retroperitoneal teratoma is related to the degree of invasion of surrounding tissues, the possibilities of complete excision and their histological nature. The local recurrence rate is around 50% [3].

In Africa in general and in particular in Mali, to our knowledge, very few cases of retroperitoneal teratoma have been reported, hence the interest of this case.

2. Observation

This is a 10-month-old male infant with no particular pathological history, resulting from a well-monitored pregnancy. He has had right abdominal swelling for 9 months, gradually increasing in size with an intermittent fever of 38˚C with no sign of deterioration in his general condition. The clinical examination found an abdominal mass, which sits at the level of the hypochondria, the flanks and the epigastrium, of mixed consistency, irregular and fixed at the deep level, not painful. Cardiovascular, pulmonary and neurological examinations are normal (**Figure 1**).

The biological blood tests did not show any notable abnormality except for hyperleukocytosis at 16.8 Mille/mm^3 with elevation of the platelet rate at 652 Mille/mm^3, of lymphocyte at 7.2 Mille/mm^3, the monocyte rate at 0.8%, 49% neutrophil, 0.3% eosinophil and 00% basophil. Urinalysis was normal as were blood urea nitrogen, electrolytes and alkaline reserve. The alfa-feto-protein assay was negative.

Abdominal ultrasound revealed a bulky heterogeneous retroperitoneal partitioned mass of 147 × 73 mm developed on either side of the spine. It presents

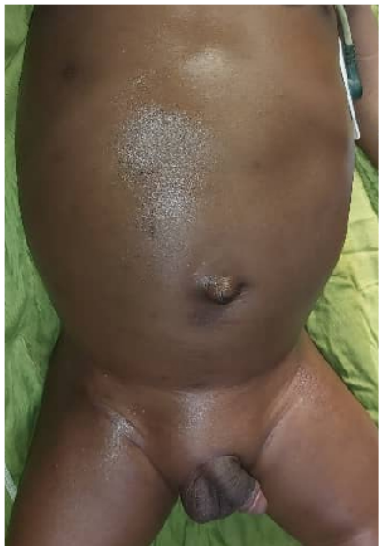

Figure 1. Photo of the infant abdomen before intervention.

cystic portions containing fine membranes, an echogenic tissue portion containing small calcifications giving shadow cones. It pushes the liver, spleen and digestive loops forward; kidneys back (**Figure 2**).

The abdomino-pelvic CT scan performed in addition to the abdominal ultrasound showed a huge bilateral para-vertebral oval partitioned retroperitoneal process measuring 147 × 73 mm, well limited containing a fatty density (−122 UH) with some internal tissue structures and bone structures. It is well limited and has a clean, slightly thickened wall. It pushes the liver, pancreas, spleen and stomach upwards, the kidneys downwards without mass effect on the vessels. There is a slight enhancement of the capsule and of the tissue portion of the mass after intravenous injection of the contrast product. Following these two radiological examinations, the diagnostic hypothesis of retroperitoneal teratoma was raised (**Figure 3** and **Figure 4**).

Surgical treatment consisted of total excision of the tumor by transverse laparotomy above the umbilical. Exploration found a voluminous bilobed mass going from the left hypochondrium to the right hypochondrium, intimately adhered to the large retroperitoneal vessels (Aorta and VCI). Careful dissection of the mass in its entirety without incident, then complete excision of the mass. Complete and satisfactory haemostasis then closure layer by layer on a suction drain. (**Figure 5**)

Macroscopically, the tumor measured 147 × 73 mm and weighed 467 grams. The postoperative course was simple (**Figure 6**).

The macroscopic study showed a yellowish piece, of firm consistency, of white-greyish colour, presenting bones, hairs accompanied by a fibrous shell (**Figure 7**).

A l'étude histologique, la paroi des kystes était bordée par un épiderme pourvu d'annexes pilo-sébacées, en profondeur s'observait des tissus osseux matures,

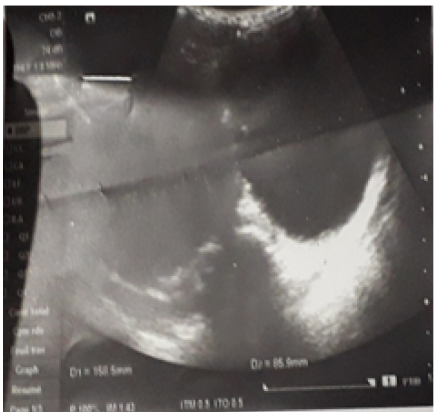

Figure 2. Ultrasound image of a rounded, bilobed mass with anechoic content with posterior cone of shadow.

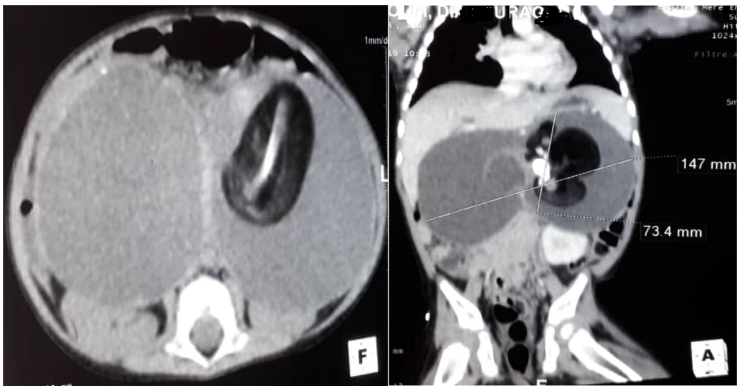

Figure 3. Abdominal CT scan, axial section and coronal reconstruction showing the retroperitoneal teratoma 147 × 73 mm, pushing back the liver and spleen above and the kidneys below.

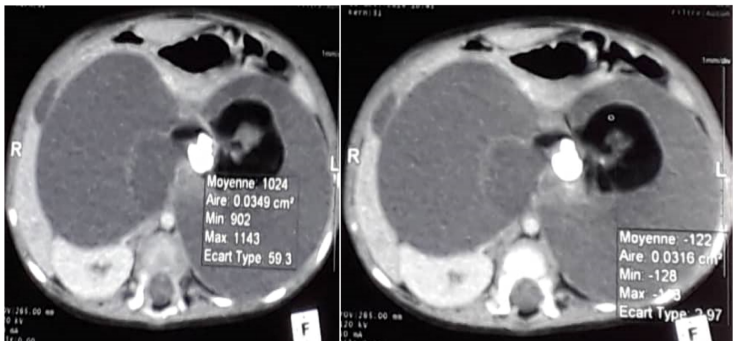

Figure 4. Abdominal CT in axial section showing the retroperitoneal teratoma with a compressive nature, with multiple components (cystic, tissue, fatty and calcium).

d'autres territoires présentaient des glandes bronchiques et du tissu glial.

On histological study, the wall of the cysts was lined by an epidermis provided with pilosebaceous appendages, in depth was observed mature bone tissue, other territories presented bronchial glands and glial tissue (**Figure 8**).

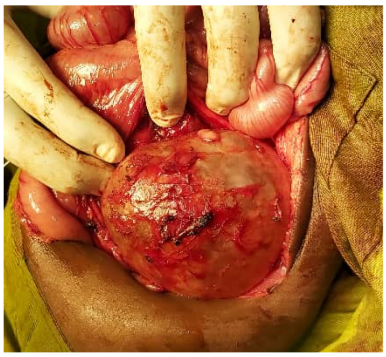

Figure 5. Intraoperative appearance of the tumor.

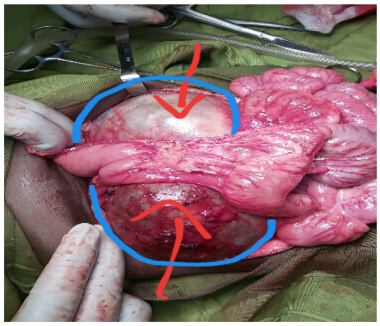

Figure 6. Intraoperative appearance of the tumor.

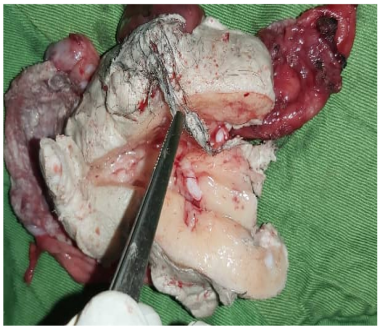

Figure 7. Operative specimen: solido-cystic mass with multiple components such as hair, fatty tissue and bone.

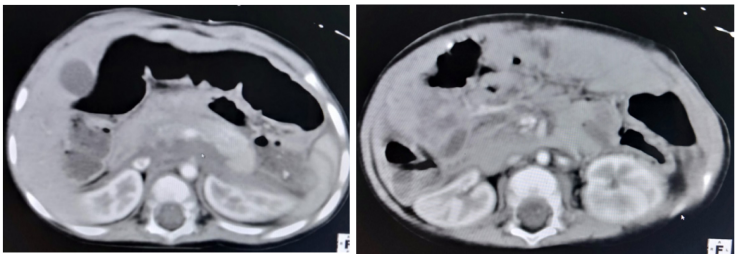

Figure 8. Abdominal CT in axial section after 3 months postoperative showing a normal aspect of the retroperitoneal compartment.

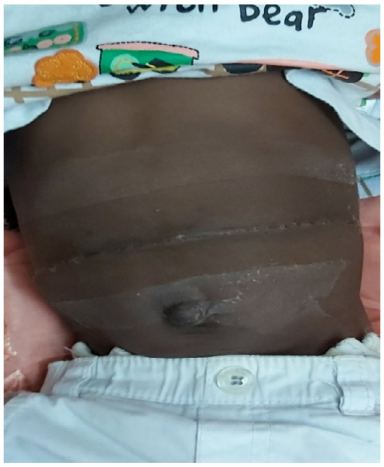

Figure 9. Postoperative photo of the abdomen showing the supraumbilical transverse laparotomy.

The anatomopathologic study confirmed the diagnosis of mature teratoma without signs of malignancy. Regular follow-up, including clinical examination, ultrasound and CT at 3 months, was unremarkable (**Figure 5** and **Figure 9**).

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4. Discussion

Teratomas are congenital tumors originating from pluripotential embryonic cells and therefore have several recognizable somatic tissues [4]. Nowadays, they represent about 1% to 10% of primary retroperitoneal tumors in children [1]. They are observed twice more in women than in men. Half of these cases are observed in female children under 10 years of age and only a fifth after 30 years. Our patient was a 10-month-old male infant with no particular pathological

history, resulting from a well-monitored pregnancy, unlike some cases reported in the literature [5] [6]. Teratomas are generally located in the ovaries, testis, anterior mediastinum or retroperitoneal region in decreasing order of frequency [4]. Retroperitoneal teratomas are often located near the upper pole of the kidney with a predominance on the left. These tumors are independent of the kidney, adrenal gland, excretory tract, large vessels and adjoining viscera. In our case, it was a retroperitoneal teratoma of bilateral paravertebral seat presenting adhesions with the large abdominal vessels. In 20% to 30% of cases, contrast enhancement of retroperitoneal teratomas is only evident in the walls and septa [7]. In our case, after intravenous injection of the iodinated contrast product, there was a slight enhancement of the capsule and of the tissue portion of the mass. Teratomas are usually benign if they are cystic and contain sebum or mature tissue, as described in our case.

The clinical signs are late and nonspecific. They are the consequence of the development of a mass in the retroperitoneum causing compression of the organs: abdominal pain, digestive signs (nausea, vomiting, constipation), and edema of the genital organs by venous or lymphatic obstruction. Contrary to what is described above, our patient only presented with an abdominal mass without any sign of compression on the neighboring organs. The discovery of a retroperitoneal teratoma of less than 5 cm is therefore rare and often fortuitous [8]. In late diagnoses, physical examination detects a large abdominal mass; which is similar to our case. The appearance of an alteration in general condition with fever and weight loss would be in favor of malignancy, unlike our case which presented episodes of fever without other associated signs.

Ultrasound is an examination that is no longer routinely performed [9]. This examination is sometimes indicated in first intention, mainly within the framework of an exploration of ¨unscrambling¨ for an ill-defined abdominal or pelvic symptomatology. It is also performed as part of a characterization, then in addition to computed tomography (CT) or magnetic resonance imaging (MRI). Its limits are the poor accessibility of the retroperitoneal space, a limited study of the anatomical relationships and the limits of the tumour, and its operator-dependent nature [10]. But in our case, it was of an important contribution for the diagnosis of the retroperitoneal mass, especially it allowed us to direct the patient for the CT scan as indicated above. Currently, CT and MRI are the examinations of choice for exploring the retroperitoneum [11].

Today, CT is the most widely used examination method for the detection, characterization, preoperative assessment and follow-up of retroperitoneal teratomas [10]. It makes it possible to confirm the retroperitoneal origin of the mass and to exclude its development from a retroperitoneal organ, to carry out a complete assessment of extension both at the supra and infra diaphragmatic level (look for metastases in the lungs, bones, hepatic and peritoneal), to ensure post-treatment follow-up, looking for example for a locoregional recurrence and finally, it allows the realization of a CT-guided biopsy [2]. In our case, the CT scan revealed a huge, well-limited bilateral paravertebral oval retroperitoneal

process containing a fatty density (density: −122 HU) with some internal tissue structures and bone structures. It has a clean, slightly thickened wall. This retroperitoneal process pushes the liver, pancreas, spleen and stomach upwards, the kidneys downwards without mass effect on the vessels. There is a slight enhancement of the capsule and of the tissue portion of the mass after injection of iodinated contrast product. With these data vessels, there is a slight enhancement of the capsule and of the tissue portion of the mass after injection of iodinated contrast product. With these data our attitude was a laparotomy in the spirit of a complete excision of the mass for the anatomopathological study. The precise diagnosis of retroperitoneal teratomas is based on anatomo-pathological examination of the surgical specimen. In our patient, the histological study confirmed the diagnosis of benign, mature teratoma and macroscopically it showed that: the wall of the cysts was lined by an epidermis provided with pilosebaceous appendages. In depth was observed mature bone tissue, other territories presented bronchial glands and glial tissue.

The prognosis of benign retroperitoneal teratomas is excellent if complete resection is possible and performed [12] [13]. Given the importance of the tumor mass, the question of the resectability of the tumor may arise; in fact, this is as often as possible. Our case benefited from the total cure of the mass. Our patient did not benefit from other treatments such as radiotherapy or chemotherapy due to the benign nature of the tumor.

Surveillance of mature retroperitoneal teratomas is mainly clinical by abdominal palpations in addition to ultrasound every 3 months for 1 year and possibly an abdomino-pelvic CT scan 6 months after the operation. The dosage of alpha feto-protein after 12 months was normal.

5. Conclusions

Retroperitoneal teratoma is a rare congenital tumour. In imaging, age, topography and sometimes the mode of installation are important elements to consider for diagnostic hypotheses. The complete imaging assessment including ultrasound and computed tomography is necessary preoperatively in order to make the diagnosis and to clearly determine the relationship with the various organs.

The anatomopathological study confirms the diagnosis of retroperitoneal teratoma. Tumor excision must be radical to avoid recurrence, which readily occurs in a malignant form.

Even if the tumor appears benign, post-operative monitoring based on clinical, biological and radiological examination is essential.

Authors' Contributions

All authors contributed to data acquisition, data analysis and interpretation, and writing of the article.

Ethical Considerations

The study was carried out with the informed consent of the parents of the infant,

the collection of data was carried out with respect for the anonymity of the patient and the confidentiality of their information.

Conflicts of Interest

The authors declare that they have no conflict of interest.

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