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A Rare Intra-Abdominal Rhabdomyosarcoma in a Child (Case Report and Review of Literature)

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Abstract: Rhabdomyosarcoma is the most common soft tissue sarcoma in infants and children. Commonly, it arises from the head and neck, genitor-urinary system, trunk and extremities. Rhabdomyosarcoma arising in the mesentery is extremely rare and only few cases were reported in the literature. This is a case report of a rhabdomyosarcoma of the mesentery in an 8-year-old girl.
Key words: Rhabdomyosarcoma, mesentery, child.

INTRODUCTION

Rhabdomyosarcoma originates from undifferentiated mesenchymal cells and can arise at any site in the body. The most common sites are the head and neck, genitourinary system, trunk and extremities [1, 2]. Rhabdomyosarcoma was also reported at other unusual sites including the duodenum, fallopian tubes, skin, extraheptic bile ducts, diaphragm and ampulla of Vater [1-9]. Primary rhabdomyosarcoma arising from the mesentery is extremely rare with only one case reported in the literature in a child [10]. We report a case of rhabdomyosarcoma of the mesentery in a 8 year-old girl.

MATERIALS & METHODES

Through our study, we report the case of an 8-year-old girl with no personal or familial pathological antecedents and who presented an abdominal mass in the left hypochondrium of recent onset (1 month) after a minor trauma revealing of a fall of its height during a game.

The rest of the clinical examination was perfectly normal. Ultrasound showed an abdominal mass in the left flank of major axis of 11cm whose etiology could not be determined.

A computed Tomodensitometry (Figure 1) was completed: an intra-peritoneal lesion process, intensely and heterogeneously enhanced after injection, delineating necrosis zones without calcification and including large vessels with left uretero-hydronephrosis.

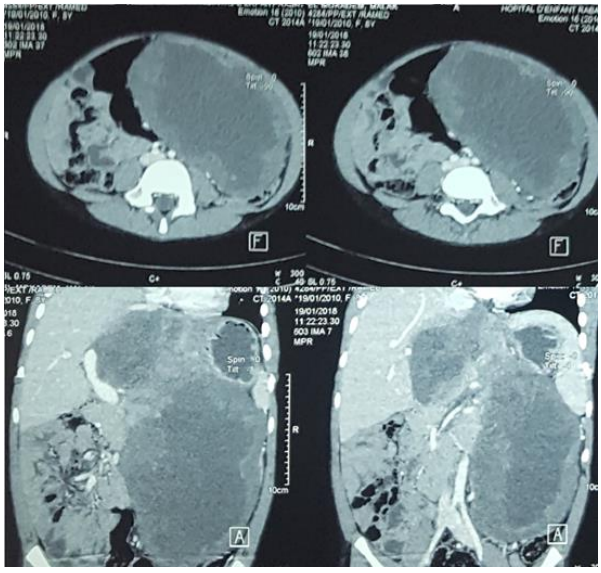

Fig-1: CT scan of the initial tumor before chemotherapy

Then an extension assessment made of standard chest radiograph initially and a thoraco-abdominopelvic computed tomography were performed and returned normal.

The alpha-foeto protein, beta-HCG and urinary catecholamine dosages also returned negative. The medullogram and the osteomedullary biopsy are without particularities.

This motivated an echo-guided biopsy after discussing with pediatric oncologists and before the diagnostic doubt, which confirms the diagnosis of a rhabdomyosarcoma. After receiving chemotherapy adapted to this invasive tumor, there is a reduction of almost 40% of the tumor. (Figure 2)

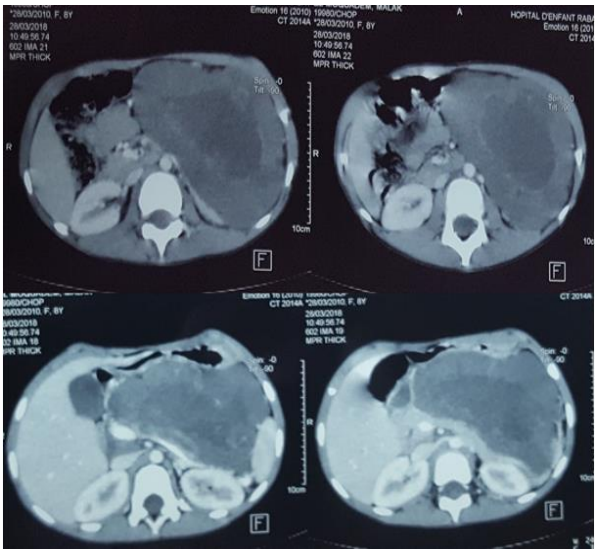

Fig-2: CT appearance of the tumor after net reduction under chemotherapy

After long discussions in a multidisciplinary staff, the decision was made to operate despite the fact that the tumor invades the large vessels in the scanner.

RESULTS

Surgical exploration, conducted by transverse left supraumbilical approach, having objectified an enormous lobulated and hard retro-peritoneal mass, infiltrating and sticking to the IVC, the VP and the celiac trunk, the liver, the duodenum and the transverse colon, the stomach (Figure 3).

Cannot resect the bulk mass, we first perform a resection of 70% of the mass initially after dissection and after section ligation of the various vascular connections between the mass and the large vessels that fed it; before starting the resection of the remainder of the mass to be oncological. Postoperative CT showed no tumor residue. Anatomo-pathological study confirmed the diagnosis of abdominal rhabdomyosarcoma (retroperitoneal).

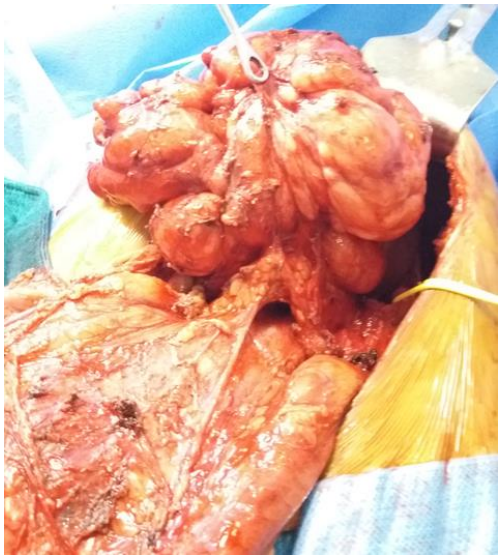

Fig-3: Intraoperative appearance of a retroperitoneal lobulated mass

DISCUSSION

Rhabdomyosarcoma is a common tumor in infants and children with a reported annual incidence of 4.5 cases per 1 million children younger than 14 years of age [11]. It represents approximately 3.5% of all malignancies in children aged 0 to 14 years [12]. Rhabdomyosarcoma commonly arise in four major sites which include the head and neck (35 to 40%), genitourinary tract (20%), extremities (15 to 20%) and the trunk (10 to 15%). It has also been reported to arise from and metastasize to nearly all body organs, but intraperitoneal rhabdomyosarcoma usually results from secondary involvement during the course of the disease. In a series of 55 children with rhabdomyosarcomas, the incidence of intraperitoneal involvement was reported as high as 11% over the course of the disease [13]. Cecchetto *et al*. [14] on the other hand, in a large series of 161 patients with nonmetastatic abdominal rhabdomyosarcomas, reported 32 intraperitoneal, 42 retroperitoneal, 75 pelvic and 12 not otherwise specified.

The exact site of origin of the 32 intraperitoneal rhabdomyosarcoma was not however specified. Rhabdomyosarcomas at these sites is considered as part of the intracavitory rhabdomyosarcoma (intraabdominal and intrathoracic rhabdomyosarcoma). Primary rhabdomyosarcoma arising in the mesentery is extremely rare. Only three cases of primary rhabdomyosarcoma of the mesentery were reported before. Agarwal *et al*. [10] reported the only child with a primary embryonal (botryoid) rhabdomyosarcoma of the mesentery. Petit *et al*. [15] on the other hand reported primary alveolar rhabdomyosarcoma of the mesentery occurring in a 68 year-old male. Seenu *et al*. [16] reported an alveolar rhabdomyosarcoma of the omentum in a 45 year-old male who presented with pyrexia. Kaplan *et al*. [17] reported an intra-abdominal embryonal rhabdomyosarcoma in a 57 year-old women but surgical exploration failed to specify the exact site of origin of the tumor. Our patient is one of the rarest cases of primary rhabdomyosarcoma of the mesentery to be reported in a child. There are four main histological types of rhabdomyosarcomas. These include embryonal, alveolar, pleomorphic and undifferentiated or anaplastic [1]. The botryoid is considered a subtype of embryonal rhabdomyosarcoma arising in mucosal lined body cavities such as the vagina, the urinary bladder, nasopharynx and extrahepatic bile ducts [1, 5, 6, 18, 19, 20]. In our patient as well as that reported by Agarwal *et al*. [10], the histology of the rhabdomyosarcoma was of botryoid type. This is unusual as these tumors are known to arise in mucosal lined body cavities, but on rare occasions they arise from unusual sites such as the conjunctiva and middle ear [1, 21, 22]. An interesting feature in our patient was the presence of four small satellite tumors in the mesentery. All showed the same histological features as the primary tumor. It is however, difficult to be certain whether these represent secondary seedings from the original tumor or they are actually multifocal tumors all arising from the mesentery. One point in favour of the multifocal origin is the absence of metastatic disease at any other sites including adjacent lymph nodes. Complete surgical excision of mesenteric rhabdomyosarcoma is the treatment of choice. This will obviate the need for local radiotherapy. In our patient, we have achieved complete excision. Regional lymph node sampling is also required.

CONCLUSION

In conclusion, rhabdomyosarcoma is one of the common tumors seen in infants and children. Rhabdomyosarcoma arising from the mesentery on the other hand is rare and should be included in the differential diagnosis of malignant primary intraperitoneal neoplasms in infants and children.

Their severity is due to an often late diagnosis due to the complacency of the space in which they develop. A complete imaging assessment including ultrasound, CT and often MRI is necessary preoperatively to determine the relationship with the various organs. The preoperative biopsy puncture is to be discussed in CPR.

The definitive diagnosis is based on surgery, which is also the most effective treatment and can be extended to neighboring organs.

Surgical treatment may be associated in case of advanced tumor with radiotherapy and chemotherapy. The high frequency of recurrence requires surveillance over several years.

Conflict d'interet

Les auteurs déclarent ne pas avoir de conflit d'intérêt et ont tous contribué à la rédaction de ce travail.

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