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Case Report

Presacral ganglioneuroma in an 8-year-old child: case report, and literature review ☆,☆☆,★

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ABSTRACT

Ganglioneuromas are benign tumors of the sympathetic nervous system, rarely found in the presacral region. In this study, we report the case of presacral ganglioneuroma in an 8-year-old girl, who complained of abdominal pain with diarrhea and abdominal distension. Ultrasound showed a large hypoechoic pelvic mass complicated by right ureter hydronephrosis. the CT and MRI confirm the presence of a presacral tissue mass with heterogeneous enhancement after contrast injection. The child underwent a complete surgical resection, and the anatomopathological study returned in favor of a ganglioneuroma. The presacral ganglioneuroma is an extremely rare tumor in that only twenty cases have been reported in the literature. 3 of which were less than 8-years-old. Through our case, we will review the epidemiological, clinical, radiological and therapeutic characteristics of this type of tumor.

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Introduction

Ganglioneuromas are benign tumors, which can be found in several parts of the human body, especially in the posterior mediastinum and in the retroperitoneum [1].

They originate from sympathetic cells, which according to the degree of differentiation will lead either to a neuroblastoma where the cells are undifferentiated, or to a ganglioneuroma where the cells are well differentiated [2].

We report an additional case of presacral ganglioneuroma found in an 8-year-old girl, recalling the main epidemiological, clinical, radiological and therapeutic aspects of these tumors.

Observation

It is an 8-year-old girl, with no particular history, who had periumbilical abdominal pain 20 days before her admission, associated with diarrhea and abdominal distension; The whole evolving in a context of apyrexia and conservation of the general state. the clinical examination reveals pelvic distension, with a dullness to percussion and a right lumbar contact. The biological tests (NFS, ionogram, ECBU) was normal, and the tumor markers (alpha fetoprotein, beta HCG) were negative.

Abdominal ultrasound finds a large heterogeneous hypoechoic mass with right upstream ureterohydronephrosis (UHN). The injected thoracoabdomino -pelvic CT scan revealed a well-limited pre-sacral mass with irregular contours and heterogeneously enhanced after injection of the contrast medium, this mass pushes back the adjacent structures and measures 92 × 70 mm extended by 152 mm (Fig. 1). Pelvic MRI shows a well-circumscribed presacral mass, hypointense on T1, hyperintense on T2, hyperintense on diffusion with no clear drop in apparent diffusion coefficient (ADC), heterogeneously enhanced after contrast, displacing the bladder with right upstream UHN (Fig. 2).

The patient underwent surgical exploration by median laparotomy, which found a huge mass intimately adherent to the sacrum and coming into contact with the bladder and the uterus, which was pushed to the left, and the mass was resected completely, with reimplantation of the ureter and ligation of the hypogastric artery (Fig. 3). The postoperative follow-up was simple, and the anatomopathological study was in favor of a ganglioneuroma with an intact capsule (Fig. 4). The patient is currently doing well with a three-month follow-up.

Discussion

Ganglioneuromas are rare benign tumors belonging to the group of neuroblastomas, and developed at the expense of sympathetic ganglion cells [1], they can be located anywhere in the body: most often in the posterior mediastinum (41%), retroperitoneum (37%) and sometimes the adrenal (21%) and cervical region (8%) [3].

The presacral region is an extremely rare location, with only 20 cases reported in the literature [1]. The average age of diagnosis is around 35 years for the presacral location [4], an age of less than 8 years has been reported in only 3 cases in the literature; and the average age is 7 years for ganglioneuromas of all locations, with a slight female predominance [4].

Clinically, ganglioneuromas remain asymptomatic for a long time until they reach a certain size [2], They may be accompanied by chronic atypical pain or constipation due to rectal compression [5]; other symptoms such as amenorrhea, neurogenic bladder have been reported in some studies [4].

Biologically, most often the assessment is normal, as are tumor markers. More rarely, an increase in blood or urinary catecholamines, or an increase in the active intestinal peptide (IPV) has been reported [4] which would be responsible for high blood pressure, diarrhea or virilization [4] our patient has not received catecholamine assays. On imaging, it is a well circumscribed tissue tumor that appears encapsulated, averaging 7 cm in size, and containing punctiform calcifications in 42% of cases [1].

On ultrasound, it appears hypoechoic, homogeneous and vascularized on color Doppler. On CT scan, it is generally hypodense and heterogeneously enhances after injection of contrast medium. It may extend to the holes of conjugation and

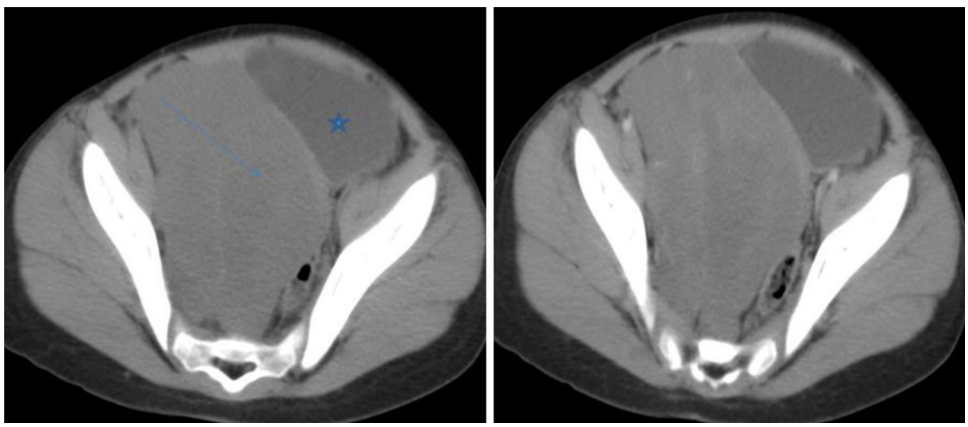

Fig. 1 – Axial plane abdominopelvic CT scan before and after injection of contrast medium showing a well-limited presacral mass, hypodense raised after injection of contrast medium (arrow), displacing the bladder (star).

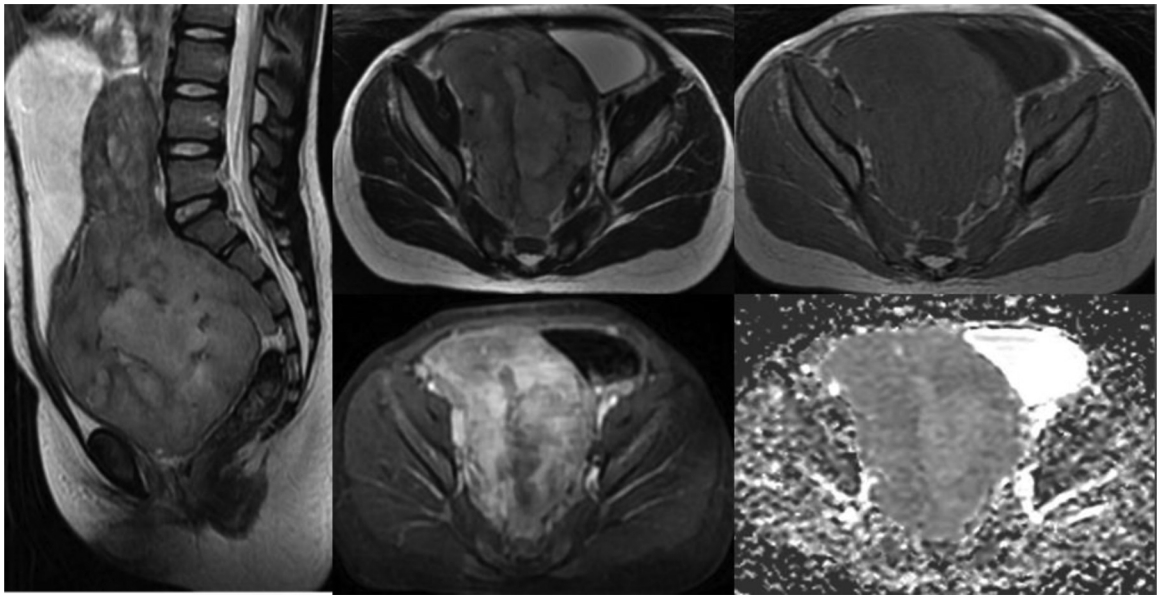

Fig. 2 – (images 3, 4, 5, 6, 7, and 8): Sagittal T2, axialT2, axial T1, axial T1 MRI after injection of contrast medium, diffusion, ADC mapping, showing a large presacral pelvic mass hypointense on T1, hyperintense onT2 and diffusion without ADC restriction, and heterogeneously enhanced after injection of gadolinium chelates.

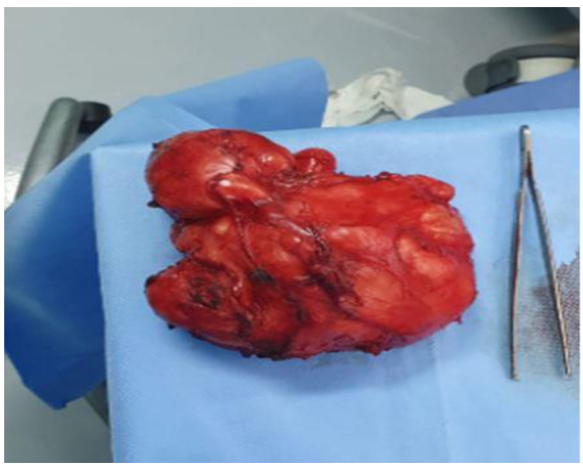

Fig. 3 – (image 9): mass operation (10 cm, 520g).

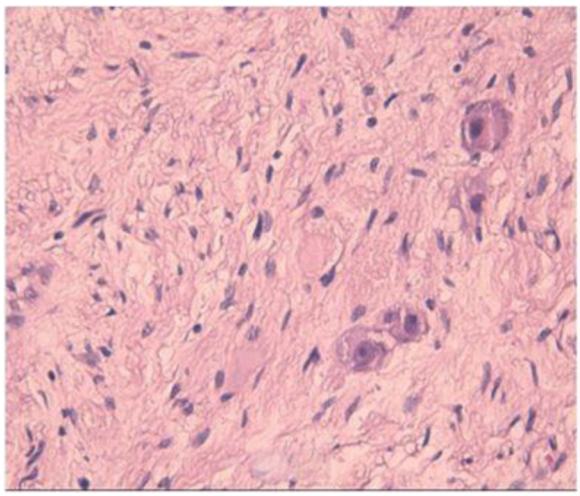

Fig. 4 – (image 10): anatomopathology specimen revealing a limited diffusive tumor proliferation, made of large cells with eosinophilic cytoplasm and irregular nuclei, arranged on a loose fibrillar background made of schwanian cells. Absence of immature neuroblastic cells.

encompasses the vascular structures without invading them. On MRI, it appears hypointense on T1, hyperintense on T2, hyperintense on diffusion with non-decreased ADC and it enhances heterogeneously after injection of contrast medium [3]. In our patient, the mass did not extend to the sacral foramen, it encompassed the vascular structures which are permeable and it pushed back the bladder causing a right UHN.

Although ganglioneuroma tends to be homogeneous, the radiological aspects are similar to those of ganglioneuroblastoma and neuroblastoma, so reliable discrimination is not possible and the definitive diagnosis remains histological [2].

The treatment of ganglioneuromas is exclusively surgical. The management includes three different approaches: a perineal approach which is performed with the patient in the prone position, the abdominal approach by median laparotomy and the combined approach which includes both a laparotomy and a sacral section [4]. The choice of the procedure depends on the level of sacral and the pelvic viscera involvement, in our case the patient had a mass extended to the abdominal region, hence the choice to perform a median laparotomy and a complete tumor resection was performed.

The recurrence rate is low for complete resections. Cases of malignant degeneration of ganglioneuromas have been described for incomplete resections in which annual monitoring by MRI is needed [5].

Chemotherapy and radiotherapy are not required, and the prognosis is good in general [4].

Conclusion

Ganglioneuromas are benign tumors of sympathetic nerve cells in which the presacral location is uncommon. Imaging cannot differentiate between neuroblastomas, ganglioneuroblastoma and ganglioneuroma. the treatment is exclusively surgical and the prognosis is generally favorable.

Authors' contribution

All authors contributed to the completion of this study.

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