

CLINICAL CASE

GIANT GANGLIONEUROMA IN PEDIATRIC PATIENT

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Abstract

Ganglioneuroma is a rare, benign neuroblastic tumor, arising mainly from the central or peripheral autonomic nervous system, especially the sympathetic system. We present the case of a 7 year old male patient, B.S., with a giant asymptomatic ganglioneuroma located in the retroperitoneum.

Keywords: *ganglioneuroma, asymptomatic, prenatal diagnosis*

Introduction

Ganglioneuromas are rare, slow growing, peripheral neuroblastic tumors arising from sympathetic ganglion cells. They are fully differentiated neuronal tumors and are benign by nature. These tumors tend to occur more frequently in females than in males and more often before the age of 20, with an average size of 7 centimeters. The most affected anatomical sites are the posterior mediastinum, retroperitoneum, adrenal gland and the soft tissue of the head and neck. They are often asymptomatic, thus making the diagnosis incidental, but when the symptoms do appear, they are due to local mass effect [1].

Imaging is the most valuable tool of identifying the ganglioneuroma, the MRI being the most helpful modality in denoting the presence of the mass, determining whether the mass is intrinsic or extrinsic to the nerve and delineating involvement of adjacent structures. Surgical biopsy is also important in its evaluation, as pathology will determine further surgical options [2].

Ganglioneuroma, which is composed of gangliocytes and mature stroma, is generally considered to be a benign tumor and potentially surgically curable. However a case of a 21-year-old woman who underwent resection of a retroperitoneal ganglioneuroma and developed spinal neuroblastoma 11 years later, does exist. To the author's knowledge, this is the first report of malignant transformation of a ganglioneuroma into a neuroblastoma. This case raises the possibility of a dedifferentiating potential for ganglion cells in a ganglioneuroma or the presence of a long-term, quiescent form of neuroblastoma.

Treatment for ganglioneuroma is largely passed on surgical removal, but not all patients will need surgery as some may best be treated conservatively with observation. Tumor excision is associated with postoperative complications which can be persistent and may affect the quality of life of survivors. The capsule may itself be adherent to important structures and total excision though desirable may not be possible. Still, tumor progression was not observed in spite of incomplete excision [3].

Case presentation

B. S., a 7 year old male was transferred from another hospital after being operated on for what was initially thought to be a hepatic tumor. Physical examination revealed an underweight, afebrile patient with a relatively good general status. The following are worth mentioning: highly enlarged abdominal mass on the right side, mostly on the upper right quadrant and right flank, with relatively high consistency, no tenderness of the abdomen and preserved transit for stools.

Notable laboratory results are:

- GPT: 64 mg/dl (7 – 56 mg/dl)
- GOT: 95 mg/dl (5 – 40 mg/dl)
- Creatinine: 0.7 mg/dl (0.5 - 1 mg/dl)
- Hb: 9.4 g/L(11.5 - 14.5 g/L)
- LDH: 222 IU/L (120 - 300 IU/L)
- VMA/24h: 5,82 mg/24h (1 - 11 mg/24h)
- NSE : 24.91 ng/mL (< or = 15 ng/mL)
- Ferritine: 53.9ng/mL (14 - 124 ng/mL)

The test results show altered hepatic function due to the mass effect on the parenchyma and also suggest the diagnosis later confirmed through biopsy, allowing the differential diagnosis with other peripheral nerve tumors such as neuroblastoma, ganglioneuroblastoma and other adrenal gland or liver tumors.



Figure 1 - Abdominal CT scan – coronal plane

The contrast CT scan reveals a giant tumor mass (150 / 133 mm) in the right hemi-abdomen

that displaces the surrounding organs (liver, portal vein, pancreas, aorta, left renal vein and artery and the inferior vena cava – which cannot be fully viewed) (Figure 1-3).

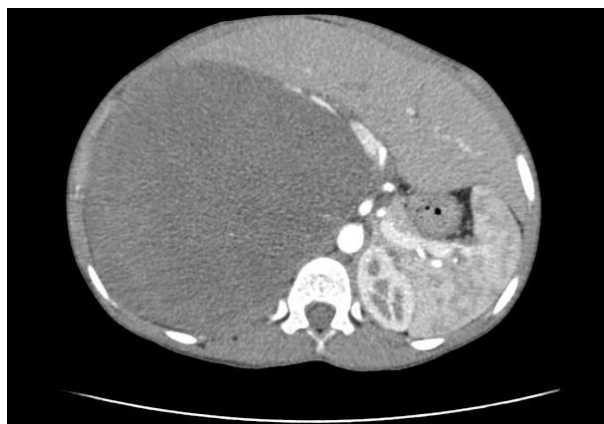


Figure 2 - Abdominal CT scan – axial plane (maximum diameter)



Figure 3 - Abdominal CT scan – axial plane



Figure 4 - Postoperative aspect of the excised tumor along with the right kidney and ureter

The patient underwent surgery which consisted in excision of the tumoral mass, along with right ureteronephrectomy. Although treatment consists of surgical excision, care must be taken for adjoining structures and nerves.

The final pathology report provided the diagnosis of ganglioneuroma, revealing an encapsulated tumor consisting of mature aggregates of ganglion cells and mucoid areas surrounded by an edematous Schwann-cell stroma; perivascular chronic inflammatory infiltrate; adjacent renal tissue showing blood stasis, discrete chronic interstitial inflammatory infiltrative tissue and tubular hyaline cylinders.

The postoperative evolution of the patient was favorable, hemodynamically stable and with no incidents.

Immunohistochemistry tests are recommended.

The patient is discharged in a relatively good shape and will be followed up in 3 months.

Discussions

Previous studies have shown that ganglioneuroma excision is associated with postoperative complications which may be

persistent and may affect the quality of life of survivors, as in this case (right ureteronephrectomy). However, the size of this tumor required surgical intervention.

Conclusions

This case presents several unique features: the patient is a male (female predominance), he is asymptomatic, and the large dimensions require surgical excision and also suggest a congenital tumor. For this reason we strongly advise prenatal investigations such as ultrasonography.

References

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