

Cervical Ganglioneuroblastoma (About a Rare Case)

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Abstract: Neuroblastic tumors are the third leading cause of tumors in young children after leukemia and central nervous system tumors. These tumors are classified into three histological groups: neuroblastoma, ganglioneuroblastoma and ganglioneuroma. Ganglioneuroblastoma is rare at the cervical level. The authors report a new case observed in a boy aged 3 years and discuss its diagnostic, therapeutic and evolutionary features.

Keywords: Ganglioneuroblastoma, Child, Neck, Cervical mass.

INTRODUCTION

Ganglioneuroblastoma is a histological type of neuroblastic tumor of the child. Its localization at the cervical level is very rare, since only 8 cases have been found in the literature [1]. It poses therapeutic problems because of frequent invasion of the vasculo-nervous structures of the neck. From an observation of cervical ganglioneuroblastoma, we take stock of this very rare localization pathology.

MATERIALS & METHODS

Through our study, we report the case of a 3-year-old boy with no personal or familial pathological antecedents who presented a progressive right-sided cervical tumefaction for 2 years without signs of compression, evolving in a context of conservation of the general state.

The rest of the clinical examination was perfectly normal. Ultrasound showed a latero-cervical tissue mass whose etiology could not be determined.

An objective computed tomography was performed: a right lateral lesion-cervical lesion with intra-pharyngeal extension with multiple jugulo-carotid

ADP (Figure 1). Then an extension assessment made of standard chest X-ray and abdominal ultrasound were performed and returned normal.

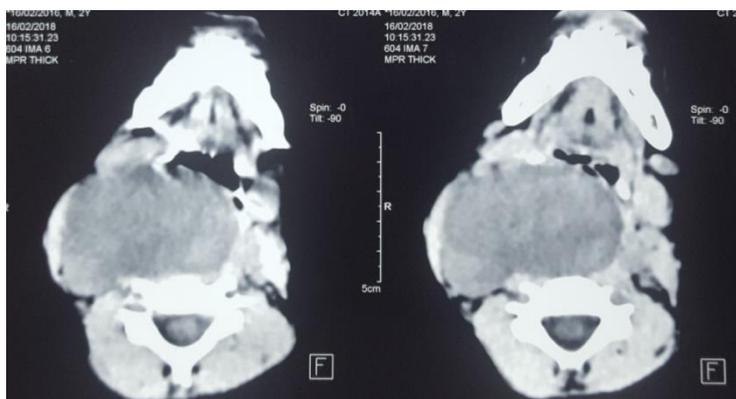


Fig-1: CT appearance showing: a right lateral lesionic lesion process with intra-pharyngeal extension with multiple jugulo-carotid ADP

Blood tests also returned normal. In view of this clinico-radiological aspect, several diagnostic hypotheses have been mentioned, notably the

possibility of tuberculous lymphadenopathy due to the endemic tuberculosis in Morocco. A malignant tumor was then suggested because of the clinical

characteristics of the mass, which may be both a metastatic adenopathy of ENT cancer or extra-ENT, a primary tumor, lymphomatous or nerve origin. This motivated a biopsy confirming the diagnosis of a cervical ganglioneuroblastoma.

RESULTS

The child received appropriate preoperative chemotherapy and then another post-chemotherapy CT

(Figure 2) was performed showing a right lateral and cervical process, with well-defined tissue densities, heterogeneously enhanced after injection with multiple hypodense lacunae areas. (necrosis) pushing back the jugulo-carotid vascular bundle, the oropharynx and the contralateral neurovascular bundle, up to the nasopharynx without invading it and back to the spine without extension, without ADP.

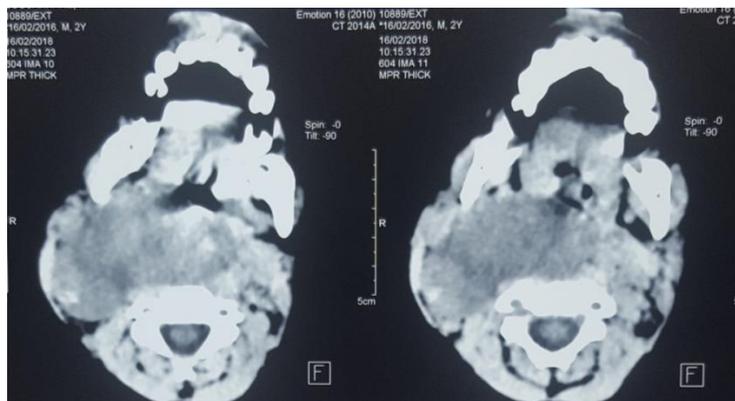


Fig-2: CT image after chemotherapy

The child was operated under general anesthesia. After incision and detachment of the cutaneo-muscular flap, a firm mass of 8 cm in diameter

was discovered, encapsulated, mobile, retro-vascular (Figure 3). Postoperative CT showed no tumor residue.

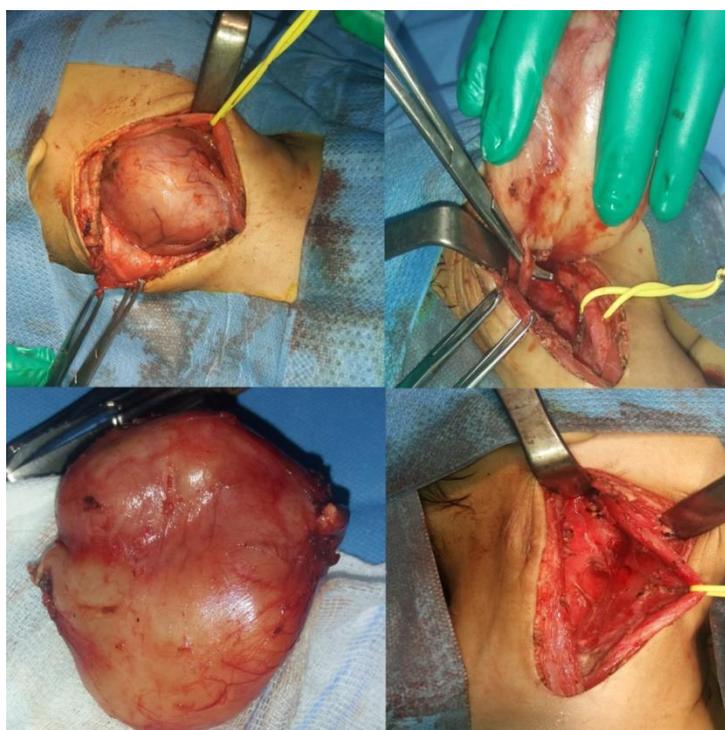


Fig-3: Intraoperative image showing a firm mass of 8 cm in diameter was discovered, encapsulated, mobile, and retro-vascular

The anatomico-pathological study confirmed the diagnosis of ganglioneuroblastoma. The evolution was favorable after adjuvant chemotherapy and presently has no sign of recurrence with a decline of 6 months.

DISCUSSION

Neuroblastic tumors are apudomas of the neural crest, representing 7 to 20% of neonatal tumors [1]. Their three histological types constitute the

different stages of the development of the same pathology and can be observed in the same tumor. Neuroblastoma is the least differentiated form with a high risk of metastasis, ganglioneuroblastoma is an intermediate form, with more ganglioneuronal than neuroblastic cells, and ganglioneuroma is the most differentiated form with a lower risk of metastasis [1,2].

Cervical ganglioneuroblastoma is the least common form. It is clinically manifested by signs of compression of the cranial nerves and the sympathetic chain. Signs of compression of the upper aerodigestive tract are common, ranging from simple drawing to true respiratory distress.

In other cases, like that of our patient, it is in the form of an isolated cervical mass. Among the specific biological signs and one of the criteria of the major diagnosis, we find the increase of urinary catecholamines in immature and / or advanced tumors.

In contrast, ganglioneuroblastoma is rarely associated with high catecholamine production [2,3]. Ultrasound can locate the tumor and differentiate it from a benign tumor, including thyroid, or a branchial cyst. CT and, increasingly, nuclear magnetic resonance imaging (MRI) are needed to determine tumor resectability, visualizing its extent and involvement of adjacent structures [2,4]. The treatment is based on surgical excision, which must be as complete as possible. However, this is not always feasible, due to the invasion of neighboring organs, including cranial nerves. About 20% of cervical ganglioneuroblastomas

are not operable. Adjuvant chemotherapy is recommended. The prognosis of these tumors is relatively good, the rate of recurrence and distant metastases is low [1-3].

CONCLUSION

Ganglioneuroblastomas represent a histological subgroup of rare neuroblastic tumors with intermediate malignant potential from neural crest progenitor cells of sympathetic nerves. Diagnosis can often be difficult on the sole basis of imaging. Pediatric surgeons and otolaryngologists must be aware of ganglioneuroblastoma when establishing the differential diagnosis of a child with cervical mass. Biopsy is recommended as a baseline investigation to avoid misdiagnosis.

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