PRIMARY CERVICAL NEUROBLASTOMA IN CHILDREN: CLINICAL CASE AND REVIEW OF THE LITERATURE

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ABSTRACT

Cervical neuroblastomas are rare tumors in children and are associated with poor outcomes. Their diagnosis requires clinical and paraclinical examinations. Imaging is key in determining the operability and search for metastases. Treatment depends on disease stage. Of note, there is limited existing data on this entity. We report a case of a 4-year-old female with cervical neuroblastoma stage 2B as per the International Neuroblastoma Staging System (INSS). She underwent surgery and adjuvant chemotherapy with favorable outcomes. Cervical neuroblastomas should be suspected in patients with a neck mass and constitutional symptoms. Treatment depends on the tumor stage.

Keywords: *cervical, neuroblastoma, child, surgery, computed tomography.*

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1. INTRODUCTION

Neuroblastomas are the most frequent tumors in children arising from primitive neuroectodermal cells and accounting for approximately 8–10% of all childhood malignancies and 15% of cancer mortality in children [1]. Cervical neuroblastomas are rare, accounting for 3-5% of neuroblastomas [2], and are usually metastases rather than primary tumors. They present as a smooth, solid, rubbery, painless mass in the neck, Horner's syndrome, ophthalmoplegia, conjunctival or eyelid edema, or papilledema. Diagnosis is based essentially on imaging and biopsy. Treatment depends on numerous factors (age, location, and staging) and includes surgical resection, chemotherapy, radiation therapy, bone marrow transplant, and immunotherapy [1]. We present the case of a 4-year-old child with cervical neuroblastoma and characterize this rare location, which will help increase knowledge about this entity.

2. Case Presentation

A 4-year-old girl presented with a rapidly progressing cervical swelling for 1 year. On examination, the child was emaciated and asthenic. Ear, nose, and throat (ENT) examination revealed a 10-cm long, painless right lower jugulo-carotid and right supra-clavicular mass, mobile on the superficial and deep planes and covered with healthy skin on cervical palpation. The rest of the ENT examination was normal, with normal tonsils, nasal endoscopy findings, and an unobstructed cavum. The patient had no symptoms of paraneoplastic syndrome. Standard preoperative laboratory work-up findings were normal. Cervical ultrasonography confirmed the presence of a mass with no collection. The thyroid gland and salivary glands were normal. Cervico-thoraco-abdomino-pelvic CT revealed a 10.5 x 6-cm long left jugulo-carotid nodal mass at the cervico region (Figure 1), displacing the vascular axis and extending into the upper mediastinum. There was no mass in the abdominopelvic region. Cervicotomy performed under general anesthesia revealed a multilobular mass (Figure 2). Histology examination revealed an undifferentiated cervical neuroblastomawithout amplification of the N-Myc oncogene (Figure 3). On immunohistochemistry, the atypical cells with hyperchromatic nuclei, chromatin with a pepper-salt appearance, positive for Vimentin, and chromogranin, synoptophysin, G-FAP. The tumor was classified as stage IIb according to the INSS (International Neuroblastoma Staging System) classification (Broudre 1993) and 05 sessions of chemotherapy based on Cyclophosphamide were decided: Endoxan 300 mg/m2 IVL Day2 to Day5, Adriblastine : 30 mg / m2 in IVL on Day6, Oncovin: 1.5 mg / m2 in IVL on Day1, followed by a assessment. the interval between treatments is 21 to 28 days.A surgical revision of the lymph nodes that appeared shortly before the start of the treatment made it possible to finish with 05 other sessions of chemotherapy based on VP16-CBDCA (etoposide carboplatin) followed by two CADO courses (cyclophosphamide, adriamycin, vincristine). The patient is in partial remission. The outcome was entirely favorable with a child who is doing well, without recurrence 1 year after the intervention.

3. DISCUSSION:

Neuroblastomas are the most frequently occurring extracranial childhood tumor. They arise from neural crest progenitor cells and occur along the sympathetic nervous system. One-third of patients are diagnosed before 1 year old, and two-thirds are diagnosed before 5 years old [2,3]. The incidence of the disease among children under 15 years is 10.5/1 million/year. The median age of presentation is approximately 18-24 months, with male predominance [2,4]. Cervical neuroblastomas are rare, accounting for 3% of neuroblastomas [1,4].

Neuroblastomas present as a palpable, multiple, indolent mass on the neck that may cause respiratory manifestations from snoring to severe respiratory distress, dysphagia, or food aspiration if compression of the pharynx and Horner's syndrome [1]. Our patient presented with a palpable cervical mass. However, there were no complications secondary to the compression of a neighboring organ.A "surgical" biopsy is recommended if the tumor is inoperable, so as to specify histological and cytogenetic analysis. Surgery is indicated only for tumors localized, resecable without noble organ sacrifice [5].

Diagnosis is based on paraclinical examination with computed tomography (CT) or magnetic resonance imaging and ultrasonography, meta-iodobenzylguanidine scan, neuron-specific enolase level, urine levels of serum catecholamines and metabolites (vanillylmandelic acid and homovanillic acid), and excisional biopsy. They produce catecholamines because they originate from neural crest cells destined to differentiate into sympathetic peripheral neurons. The urine metabolites are raised in most patients (80–95%) [5]. However, catecholamines are only positive in 30 to 40% of cases, and are very frequently negative in cervical locations as in the present clinical case [2]. CT is also useful in determining other locations In our case, CT helped characterize the tumor: we found no metastasis.

Histologic confirmation is necessary using light microscopy, immunohistochemistry, or electron microscopy. Histology reveals small round pale blue cells known as Homer-Wright pseudorosettes. Moreover, DNA ploidy and N-myc proto-oncogene (*MYCN)* gene status should be assessed on the sample [1]. In our patient, immunohistochemistry staining was positive for vimentin, chromogranin, synaptophysin, and glial filament acidic protein, confirming its neurogenic nature.

Treatment of neuroblastoma is risk-based and is largely determined by tumor stage in conjunction with other factors including patient age, tumor location and resectability, tumor histopathology, and *MYCN* gene amplification [2]. Low-risk patients are observed or treated with surgical resection only; intermediate-risk patients require surgical resection with or without chemotherapy; and high-risk patients require multiple modalities including surgical resection, chemotherapy, radiation therapy, and immunotherapy [1]. Our patient was an intermediate-risk patient according to the classification, and therefore underwent surgery with adjuvant chemotherapy according to the usual protocols; the patient had a favorable response. Trends are in the search for new chemotherapeutic agents, new forms of retinoid for the treatment of residual disease, immunotherapy for a more selective approach without cross-reaction with chemotherapy, meta-iodobenzylguanidine (MIBG) for the evaluation of residual tumors and antiangiogenic agents [12].

4. CONCLUSION:

Cervical neuroblastomas are common tumors in children that should be suspected before a neck mass and constitutional symptoms. biopsy and/or surgery is important in confirming the diagnosis, and treatment depends on the stage.

Parental consent

The patient's family authorizes the anonymous publication of the patient's information in a scientific journal.

Conflicts of interest

The authors declare no conflict of interest.

Author contributions

All authors read and approved the final version of the manuscript.

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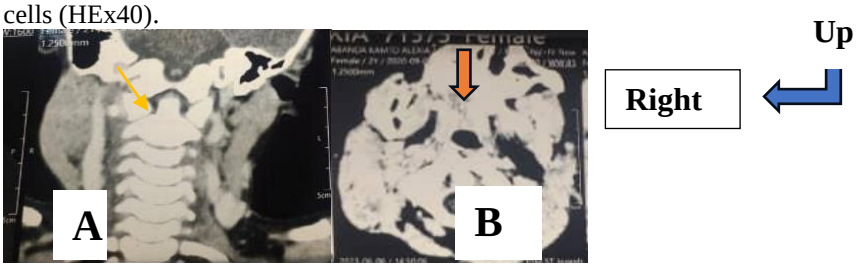

Fig. 1: CT coronal and axial section: Right heterogeneous cervical tissue mass capturing the contrast product. sheathing the vessels of the neck.

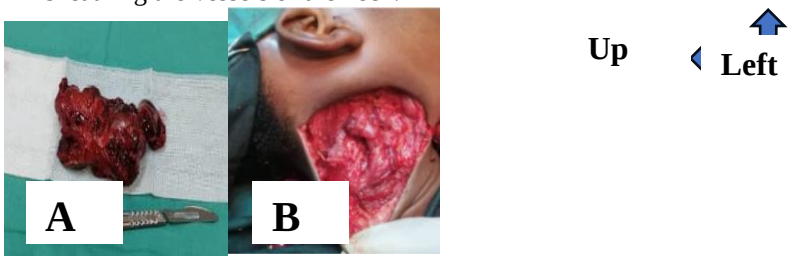

Fig. 2: Intraoperative view (A) of the mass, (B) operating site after removal of the mass.

Fig. 3: Parenchyma of the primary cervical tumor is the site of tumor proliferation undifferentiated small round blue cells (HEx40).

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