Case report

Cervical ganglioneuroblastoma. a case report

Abstract

Neuroblastoma is the most common solid extracranial tumor in childhood, that arises from neural crest cells. The cervical location is the least frequent and its clinical manifestations are diverse, from cervical mass, dyspnea, stridor, Horner's syndrome or dysphagia. In this case report, we present a male infant with an atypical clinical symptoms, with upper airway obstruction that can occur in multiple contexts, whether malformative or neoplastic masses; Simple and contrast-enhanced neck tomography revealed a lesion in the right posterior cervical triangle with involvement in the retropharyngeal region and significant displacement of the airway. The tumor was successfully excised. The histopathological diagnosis was mixed ganglioneuroblastoma, rich in Schwannian stroma, treatment was started with the COG protocol with 4 cycles of chemotherapy, at the end a control MRI was performed without evidence of tumor recurrence. The importance of recognizing primary tumors with atypical signs and symptoms is highlighted, given that the frequency of neuroblastomas, statistically speaking, is the second cause of childhood cancer, early diagnosis must be made in order to achieve successful treatment.

Keywords: Ganglioneuroblastoma, cervical mass, tumor, pediatric oncology

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Introduction

Neuroblastoma is the most common solid extracranial tumor in childhood, that arises from neural crest cells. A study of 8,369 case reports with neuroblastoma revealed primary tumor sites where 47% were adrenal glands, 24% abdomen/retroperitoneum, 15% chest cavity, 3% pelvis, 3% neck, and 8% unspecified site. It usually presents as an abdominal mass, the cervical location is one of the least frequent and its symptoms are diverse, from a palpable mass, dyspnea, stridor, Horner's syndrome and dysphagia. The incidence in Mexico with data obtained from the IMSS during 1996-2005 was 3.8 per 1000 cases/year, however only 12.1% were on the face and neck; in other figures it corresponds to 4.1% of cancers in children, and is responsible for 4.9% of all infant mortality from cancer.[1-3]

Case report

A 1 year 8 months old male patient, was admitted to our department, who began his condition in April 2020 with the presence of nocturnal rhonchi, stridor, respiratory distress in physiological sleep, with thoracoabdominal dissociation, as well as dysphagia, weight loss, ptosis and anhidrosis (Horner Syndrome). No apparent history of fever, weight loss, or diaphoresis. On physical examination, a mass was palpable, with no lymphadenopathy.

On hospital admission, imaging studies were performed, simple and contrast-enhanced neck tomography (Figure 1) finding a lesion in the right posterior cervical triangle of etiology to be determined, affecting the retropharyngeal region, ovoid, with defined borders, of heterogeneous density, with hypodense central áreas, and that after the application of contrast medium they present heterogeneous reinforcement, in the periphery with lower density, presenting dimensions of 42.92x22x36.10mm., which conditions significant displacement of the airway with a decrease in its caliber and lateral displacement of the ipsilateral carotid space causing width of the carotid bifurcation. Bone marrow aspirate and bone biopsy were performed with negative results.

Resection and right cervical lymphadenectomy were scheduled in July 202, during the initial video laryngoscopy (Figure 2) was found displacement of the trachea to the left, caused by the mass effect, the histopathology report (Figure 3) interspersed ganglioneuroblastoma, rich in Schwannian stroma with some foci of dystrophic calcification, completely resected, with a low mitosis/karyorrhexis index (<100 per 5000 cells). favorable intermediate risk group, stage I, by International Neuroblastoma Risk Group (INRGSS) L2 by IDFRS (location of the primary in the right carotid), according to the INRGSS pre-treatment classification scheme at low risk, however due to symptomatic disease at diagnosis treatment begins with the COG protocol with 4-cycle chemotherapy, with the administration of etoposide, carboplatin, dexrazoxane, and doxorubicin. At the end of the 4-cycle chemotherapy, a control magnetic resonance was performed without evidence of tumor recurrence.[4]

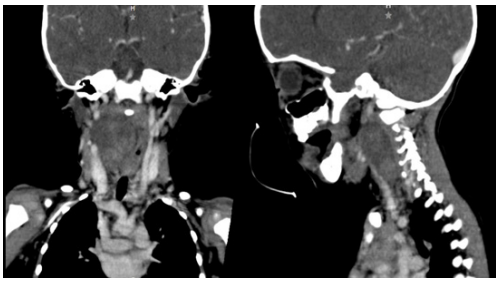

Figure 1 Simple and contrast-enhanced neck tomography. Coronal (A), Sagital (B).

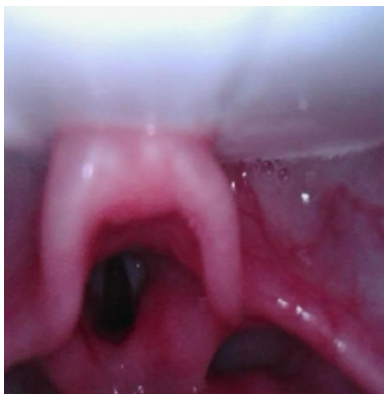

Figure 2 Initial video laryngoscopy, showing displacement of the trachea to the left, due by the mass effect of the tumor.

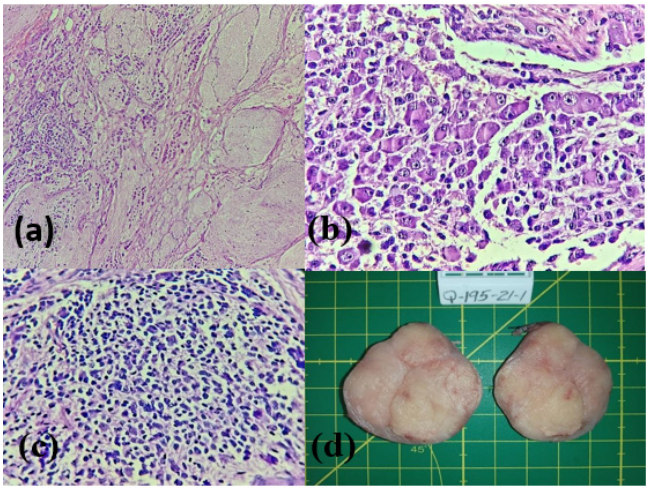

Figure 3 Histopathological study: (a) Results of hematoxylin-eosin staining (H&E) of the tumor showed a predominance of Schwannian stroma and ganglion cell maturation. (b) Ganglion cells (40x) (c) Focally distributed nests of blast cells. (d) Macroscopic image of the tumor.

Discussion

Cervical masses are very common in the pediatric population and most are benign (e.g., lipoma, fibroma, hemangioma.[5] Neuroblastoma represents a neoplastic expansion of neural crest cells in the developing sympathetic nervous system, it is approximately 97% of all neuroblastic tumors and is the third most common childhood cancer. The primary tumor originates anywhere along the sympathetic chain, usually located in the adrenal gland.[5,6] The median age at diagnosis is 19 months.[7] Primary cervical neuroblastoma should be ruled out in any child less than one year of age with a persistent neck mass.[8]

The prognosis for neuroblastoma varies widely, from tumors that spontaneously regress and require no intervention to those that present widely metastatic and resistant to therapy with resulting high mortality;[6] also varies with age, children who are less than 1 year old at diagnosis of neuroblastoma have a significantly higher 5-year survival rate compared with those who receive the diagnosis at greater than 1 year of age.[7]

The clinical presentation can be quite heterogeneous ranging from asymptomatic incidental tumors, respiratory symptoms from snoring to severe respiratory distress, and widespread metastases with systemic manifestations.[3,9] A study made in 2017, 3 of the 7 patients (43%) with primary head and neck disease presented with Horner's síndrome, they exhibited miosis and ptosis on the affected side. Horner's syndrome is caused by a disruption of the oculosympathetic system anywhere along its three neuron pathway. Cervical tumors can cause mass effect on the cervical sympathetic trunk, this was the case in our patient.[8]

The neuroblastoma primarily contains immature cells, some have a component of fully mature ganglion cells that are typically found in a ganglioneuroma. A tumor with both elements of mature and immature cells is called a ganglioneuroblastoma. Diagnosis is made on the basis of histologic confirmation combined with chemical profiling and imaging characteristics.[7] Tumors have been classified in detail according to this degree of differentiation by the International Neuroblastoma Pathology Committee, favorable and unfavorable histologic subtypes are based upon the level of Schwannian stroma present in the tumor, then further subclassified based upon the mitosis-karyorrhexis index (MKI) and patient age.[6-9]

The International Neuroblastoma Risk Group Staging System (INRGSS) was published in 2008 as a new way to stratify patients before surgical intervention. This system is now used in parallel with the older International Neuroblastoma Staging System (INSS), this one was intended for postsurgical staging, and the INRGSS emphasizes pretreatment risk stratification based on clinical criteria and image-defined risk factors (IDRFs), these ones are focused on evaluation of tumor extent to nearby vessels and adjacent structures, in the case of neck tumor the risk factors are tumor encasing carotid and/or vertebral body and/or internal jugular vein, extending to base of skull, compressing the trachea and ipsilateral tumor extension within two body compartments.[6-10]

To determine the appropriate treatment strategy, it needs to be combining the INSS/INRGSS stage with the age at diagnosis, the histologic results, and the biology and genetics of the tumor allows the patient to be placed into a low, intermediate, or high-risk group. The intensity and duration of treatment are then determined. Surgical removal of the tumor may be all that is necessary for children with low-risk disease that is localized, in intermediate-risk tumors may receive chemotherapy to shrink the tumor before surgical removal, and in higher risk, surgery may be performed to remove as much tumor as possible after the induction of chemotherapy. Chemotherapy is added for tumors that cannot be resected or for tumors that may cause spinal cord compression or respiratory compromise which this was the case.[6-9]

There is no unanimous consensus on the best surgical technique, and the operative approach largely depends on the anatomical location, the extension of the mass, and the surgeon's expertise and confidence with a technique.[4]

Conclusion

In this case report, we present a male infant with an atypical clinical symptoms, with upper airway obstruction that can occur in multiple contexts, either malformative or neoplastic masses, in addition to an infrequent location; with a lack of standard characteristics of clinical and imaging presentations, preoperative diagnosis continues to be a challenge, with the definitive diagnosis depending on histopathology. Therefore, this case highlights the importance of recognizing primary tumors with atypical signs and symptoms, but given that the frequency since neuroblastomas are statistically the second cause of childhood cancer, early diagnosis must be made in order to achieve successful treatment.

Surgery has an important role in the multidisciplinary treatment of cancer patients, where the main objective is a complete resection of the tumor (>90%) respecting its integrity, avoiding invasion, damage to surrounding organs and tissues, which still represents a surgical challenge.

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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