Choroid plexus carcinoma: case report

Carcinoma de plexo coroide: relato de caso

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ABSTRACT

Aims: To describe a case of choroid plexus carcinoma which initial signs and symptoms were related to the upper respiratory tract, contrary to the classic symptomatology of this disease reported in the literature.

Case description: A two years and eight months old boy was admitted due to acute respiratory failure. He was treated with antibiotics for suspected croup without success. After two weeks he presented neurological manifestations that led to investigation and diagnosis of choroid plexus carcinoma. Resection was contraindicated due to extension of the tumor and involvement of vital areas.

Conclusions: The rapid evolution of choroid plexus carcinoma in this case points to the need for early suspicion of central nervous system disorders. This case may alert pediatricians to the need to consider the presence of a brain tumor causing injury to the vagus nerve in cases of respiratory distress resistant to treatment.

KEY WORDS: BRAIN NEOPLASMS; CHOROID PLEXUS NEOPLASMS/diagnosis; VAGUS NERVE/injuries; SIGNS AND SYMPTOMS, RESPIRATORY; NEUROBEHAVIORAL MANIFESTATIONS; CHILD; MALE; CASE REPORTS.

RESUMO

Objetivos: Descrever um caso de carcinoma do plexo coroide no qual os sinais e sintomas iniciais foram relacionadas ao trato respiratório superior, ao contrário da sintomatologia clássica da doença reportada na literatura.

Descrição do caso: Um menino de dois anos e oito meses de idade foi internado por insuficiência respiratória aguda. O paciente foi tratado com antibióticos para suspeita de crupe, sem sucesso, e após oito dias mostrou sintomatologia neurológica, que levou a uma investigação e ao diagnóstico de carcinoma do plexo coroide. A ressecção foi contra-indicada devido à extensão do tumor e envolvimento de áreas vitais.

Conclusões: A evolução rápida do carcinoma do plexo coroide neste caso mostra a necessidade da suspeita precoce de distúrbios do sistema nervoso central. Este caso pode alertar os pediatras para a necessidade de considerar a presença de um tumor cerebral levando a lesão do nervo vago em casos de dificuldade respiratória resistente ao tratamento.

DESCRITORES: NEOPLASIAS ENCEFÁLICAS; NEOPLASIAS DO PLEXO CORÓIDE/DIAGNÓSTICO; NERVO VAGO/lesões; SINAIS E SINTOMAS RESPIRATÓRIOS; MANIFESTAÇÕES NEUROCOMPORTAMENTAIS; CRIANÇA; MASCULINO; RELATOS DE CASOS.

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INTRODUCTION

Choroid plexus carcinoma (CPC) is a neoplasm of the central nervous system that originate in the epithelial lining of the cerebral ventricles.[1] These tumors are rare, accounting for 0.3 to 0.6% of brain tumors at any age, and 1 to 4% of brain tumors before the age of 15 years.[2] The incidence is higher before two years of age, and this is the age group that shows the largest number of tumors in this area.[1] Most of the choroid plexus tumors are benign. The malignant form represents only 8.1% of the total of such tumors.[3]

CPC have a unique and dangerous biological behavior and affect children mainly from two to four years old. Lateral ventricles are the most affected by CPC. Symptoms are rare, usually a consequence of hydrocephalus.[4] In children, the most common clinical findings are increased brain diameter, mental delay, vomiting, lethargy, irritability, motor and sensory disorders, convulsions, enlargement of the cranial sutures and papilledema.[5]

In cases where CPC causes injury to the lateral and third ventricles, the classical symptoms of intracranial hypertension with headache, vomiting and papilledema occur in children older than two years of age, with macrocephaly in younger children.[4] The relatively small number of cases reported in the literature, the lack of relevant epidemiological data and controversies about clinical and pathological classification of CPC make it difficult to establish a standardized therapeutic approach. Since the healing is achieved by a limited number of patients, it is important to determine the relevant prognostic factors from survival analyses of these patients.[6]

An atypical initial presentation of CPC is reported here. The study was approved by the Ethics Committee for Institutional Research of the Universidade Estadual de Montes Claros, Minas Gerais, Brazil.

CASE REPORT

A two years and eight months old boy arrived to the emergency room of the Clemente Faria's University Hospital, Montes Claros, Minas Gerais, Brazil. The patient was transferred from a small neighboring town where he was hospitalized, because of acute respiratory distress. His mother denied a history of allergy or respiratory disease and reported that he had recently fallen a few times. The patient had been hospitalized for five days in his own town with moderate dyspnea and treated for laryngitis. After antibiotic therapy (ceftriaxone) with no improvement, he was referred to Montes Claros due to worsening of respiratory distress. When the patient was admitted to the Pediatric Intensive Care Unit, assisted ventilation was started and he was treated for a suspected diagnosis of laryngitis.

Eigth days after admission the patient underwent a tracheostomy. Sedation was decreased due to clinical improvement. One day after stopping sedation, the patient began to show neurological symptoms such as facial paralysis, with labial commissure deviated to the left, dyslalia, hemiplegia and vomiting. Computer tomography (CT) revealed an intraventricular expanding lesion in the posterior horn of the lateral ventricle that was hyperdense and invading the adjacent brain wall and parenchyma. Associated vasogenic edema and intense enhancement after contrast was noted, indicating a highly aggressive tumor. Magnetic resonance imaging (MRI) showed an expansive, solid and highly aggressive lesion with an epicenter in the posterior horn of the right lateral ventricle, probably primarily of the choroid plexus, with periventricular extension to the region of the basal ganglia (Figure 1-A) and secondary lesions in the posterior fossa (Figure 1-B).

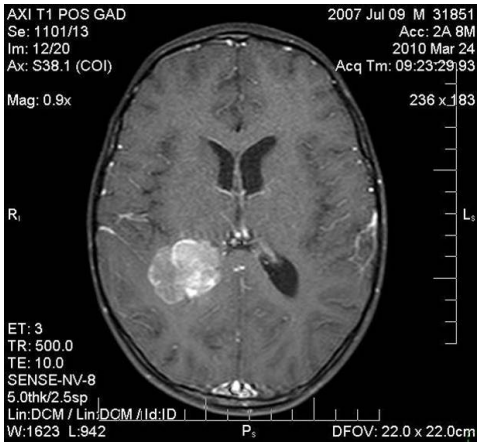

Figure 1-A. Magnetic resonance imaging, axial T1-weighted after gadolinium: bulky mass lesion located within the right lateral ventricle, showing intense enhancement after contrast medium.

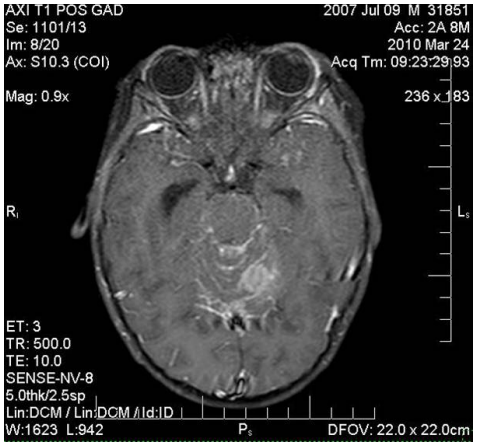

Figure 1-B. Magnetic resonance imaging, axial T1-weighted after gadolinium: an extensive lesion with intense enhancement after contrast medium located in the posterior fossa on the left.

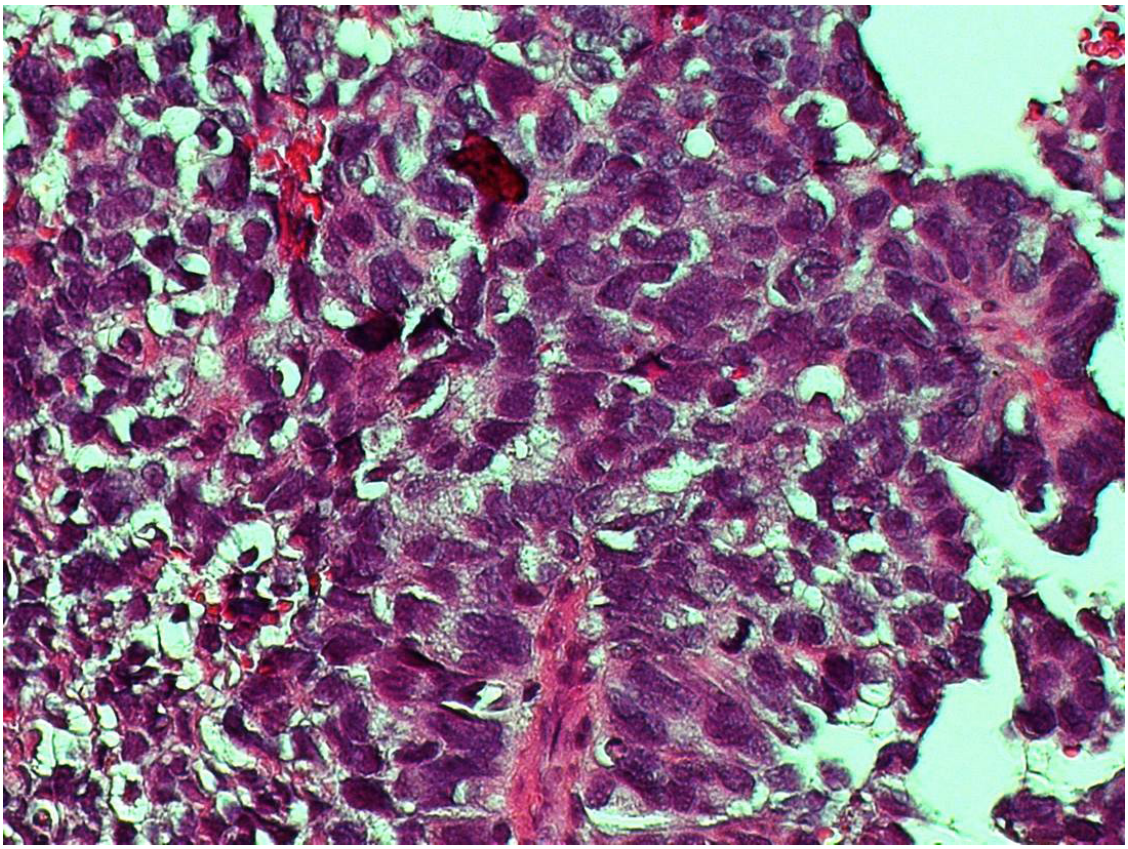

Figure 2. Epithelial cells with evident pleomorphism, conspicuous nucleolus, hyperchromasia and mitosis. Hematoxylin and eosin stain. Magnification 100 X.

After ten days the child stopped responding to commands and showed vacant eyes, strabismus, ptosis and right facial palsy. Surgical resection of the tumor was not possible due to its size and involvement of vital areas. Craniotomy was performed for pathological diagnosis. The microscopic examination of the biopsy revealed fragments of a malignant neoplasm constituted by a solid mass with moderate and marked nuclear atypia and frequent mitotic figures with atypical morphology and additional features of necrosis (Figure 2). The diagnosis of CPC was confirmed.

The choice of treatment was radiotherapy. Since there were no indications for continued hospitalization, the child was discharged. Radiation therapy was never performed because the patient died 13 days after the onset of the first neurological signs.

DISCUSSION

Cases of CPC are rare and usually occur within the first three years of life.[7] The most common site in adults is the fourth ventricle, whereas in children it is the atrium or trigone of the lateral ventricles (85%).[8-10] The incidence is higher among males.[3] This case presented some common pathological features described in the literature, but the clinical picture was not as usual.

The literature presents classical manifestations for CPC in children under two years of age. When the lateral ventricles and third ventricle are affected, CPC produces hydrocephalus 2-3 months before the diagnosis is made from the classic symptoms of intracranial hypertension, headache, vomiting and papilledema.[1,3] In addition, 70% of cases have signs of intracranial hypertension.[1]

In this case, the typical signs of CPC were not present. The patient had only fallen a few times one week before entering the emergency room with a clinical picture of respiratory distress. These symptoms, despite being attributed to a diagnosis of croup, were probably already due to the CPC. No typical manifestations related to cranial hypertension (headache, vomiting or papilledema) or seizures occurred. Hydrocephalus was only discovered by MRI. Most of the cranial nerves between the brain tumor and the brainstem were affected, including the facial nerve, with a late presentation of facial palsy, and vagus nerve, wich is responsible for innervating part of the larynx of the epiglottis and thus led to the respiratory distress. This

case has also the unique aspect of a tumor originated in the lateral ventricle and reaching the brainstem. Studies in the literature have reported that tumors in the fourth ventricle can eventually invade the brainstem, but not those originating in the lateral ventricle.[1]

CPC is diagnosed using imaging methods such as CT and MRI. Studies show that CT is sufficient to confirm the diagnosis and assess the degree of hydrocephalus, the location of the tumor and whether it is confined to the ventricular cavity or has invaded the brain parenchyma. Possible tumor dissemination in the central nervous system can also be detected. However, MRI with multiplanar high-resolution images offers additional advantages in visualization of the lesion. This facilitates the studies of ventricular anatomy, surgical planning and post-operative evaluations.[4] CT scanning of the patient showed invasion of the brain wall and adjacent parenchyma, whereas MRI clearly showed the primary and secondary lesions of the choroid plexus.

Among the available treatments for CPC, maximum surgical resection offers the best chance of survival in the long term.[11,12] However, the tumor can reach a large size and level of vascularization, making surgical treatment difficult.[6] The patient reported herein did not undergo surgery because the diagnosis was delayed and the tumor had affected vital areas.

There are two important aspects of this case report, the first one being the rapid evolution of this tumor. The patient died less than three weeks after hospitalization and two weeks after the discovery of the brain lesion. Secondly, this case shows an uncommon manifestation of CPC, with respiratory symptoms arising from the expansion of the tumor. Therefore, this report should alert pediatricians to the need to consider the possibility of a brain tumor when there are clinical manifestations of intractable respiratory distress. In this case, if the diagnosis of CPC had been made earlier, perhaps effective therapy had been possible.

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