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Case Report

Microsurgical Resection of Cerebellopontine Angle Choroid Plexus Papilloma by Far-Lateral Approach. Case Report

Ressecção Microcirúrgica de Papiloma de Plexo Coroide do Ângulo Pontocerebelar Através do Acesso Far-Lateral. Relato de Caso

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ABSTRACT

Introduction: *Choroid plexus tumors (CPTs) are rare papillary neoplasms derived from choroid plexus epithelium. They account for only approximately 0.4%-0.6% of all intracranial tumors, but 10%-20% of brain tumors occurring throughout the first year of life.* ***Objective:*** *The present study describes the case of an adult man presenting a cerebellopontine angle choroid plexus papilloma (CPP) microsurgically treated through suboccipital far-lateral approach.* ***Case report:*** *A 67-years-old-man was admitted presenting progressive headaches and left lower limb weakness. Magnetic resonance images showed a large tumor on the left cerebellopontine angle with heterogeneous contrast enhancement. Total surgical resection was achieved through a right suboccipital far-lateral craniotomy on lateral position with neurophysiological monitoring of lower cranial nerves. No alterations on cranial nerves function was observed during tumor resection. The patient presented an uneventful recovery and was discharged home on the fifth post-operative day. On the six months follow-up, he was asymptomatic. Anatomopathological analysis confirmed the diagnosis of CPP, WHO grade I.* ***Conclusion:*** *Cerebellopontine angle's cpp is an extremely rare neoplasm that challenges a neurosurgeon ability to deal with tumor in close relation to lower cranial nerves. Appropriate neurosurgical route, surgeons experience and adequate anatomical knowledge of important neural and vascular structures are fundamental to safely remove CPP of the posterior fossa. Suboccipital far-lateral craniotomy gives enough view with minimal retraction to manage the tumor.*

Key-words: *Choroid plexus papilloma; Cerebellopontine angle; Occipital far-lateral approach*

RESUMO

Introdução: *Tumores do plexo coroide (CPT) são neoplasias papilares raras derivadas do epitélio do plexo coroide. Elas representam apenas 0,4%-0,6% de todos os tumores intracranianos, porém cerca de 10%-20% dos tumores cerebrais que ocorrem no primeiro ano de vida.* ***Objetivo:*** *O objetivo do presente estudo é apresentar o caso de um homem adulto portador de papiloma de plexo coroide na fossa posterior tratado com ressecção microcirúrgica da lesão através de acesso far-lateral.* ***Relato do caso:*** *Homem de 67 anos foi admitido no setor de emergência apresentando queixa de dor de cabeça e perda progressiva da força em membro inferior esquerdo. O exame de ressonância magnética encefálica registrou a presença de lesão expansiva ocupando o ângulo pontocerebelar direito e com captação heterogênea do meio de contraste. A ressecção total da lesão foi possível através de acesso subocciptal far-lateral direito na posição lateral esquerda e com monitorização contínua intraoperatória dos nervos cranianos inferiores. Não foram identificadas alterações dos nervos cranianos durante o ato operatório. O paciente apresentou recuperação pós-cirúrgica satisfatória e recebeu alta hospitalar no quinto dia pós-operatório. Em seguimento ambulatorial após seis meses, o paciente encontrava-se assintomático e com recuperação completa da função motora em membros inferiores. O estudo anátomo-patológico revelou tratar-se de papiloma de plexo coroide grau I da OMS.* ***Conclusão:*** *O papiloma de plexo coroide é uma neoplasia extremamente rara que desafia a habilidade neurocirúrgica de lidar com lesões que envolvem nervos cranianos na fossa posterior. A via neurocirúrgica apropriada, a experiência operatória e o conhecimento das particularidades anatômicas vasculares e neurais são extremamente importantes para a adequada resseção da lesão e preservação das funções neurológicas. O acesso subocciptal extremo-lateral permite visualização suficiente das estruturas e mínima retração de parênquima cerebelar.*

Palavras-chave: *Papiloma de plexo coroide; Ângulo pontocerebelar; Acesso extremo lateral*

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Introduction

Choroid plexus tumor (CPT) is a rare papillary neoplasm derived from choroid plexus epithelium[1-3]. They account for only approximately 0.4%-0.6% of all intracranial tumors, but 10%-20% of brain tumors occurring throughout the first year of life[2-5]. CPT is classified according to the World Health Organization (WHO) in three distinct categories: grade I for choroid plexus papilloma (CPP), grade II for atypical choroid plexus papilloma (aCPP) and grade III for choroid plexus carcinoma (CPC)[6]. CPP is generally a histologically benign, slow-growing tumor, and malignant evolution is uncommon occurring in 10%-30% of cases, mainly in the lateral ventricle[7, 8]. These tumors predominantly arise in supratentorial locations and only approximately 9% of all CPTs are located in the cerebellopontine angle [9,14]. Surgical resection remains the standard of care, although chemotherapy and radiotherapy may be considered for recurrent or metastatic lesions[15]. Far-lateral approach is a surgical route that allows ample visualization of posterior fossa structures, mainly those located in the cerebellopontine/bulbar angle, with minimal cerebellar retraction[12,13].

The aim of the present study is to describe the case of an adult man presenting a cerebellopontine angle choroid plexus papilloma surgically treated through an occipital far-lateral approach. We discuss the benefits of this route in order to totally resect the tumor.

Case Report

A 67-year-old-man was admitted presenting progressive headaches and left leg weakness. On the neurological examination, he presented hyperreflexia and spastic monoparesis of the left leg with muscle activation against some resistance and full range of motion. No cranial nerve deficits were observed. Magnetic resonance images (MRI) showed a large tumor on the right cerebellopontine angle with heterogeneous contrast enhancement compressing the brainstem and cerebellum. Fourth ventricle was displaced and compressed (Figure 1). Total surgical resection was achieved through a right suboccipital far-lateral craniotomy on left lateral position with neurophysiological monitoring of lower cranial nerves (Figure 2). Tumor resection was performed carefully under microscope view and utilizing microdissector most of the time. The tumor was not adhered to the cerebellum and pons which allowed easy detachment from neural structures. Basilar and vertebral arteries were not involved by the tumor and could be safely preserved during all the procedure. No alterations on cranial nerves function was observed during tumor resection.

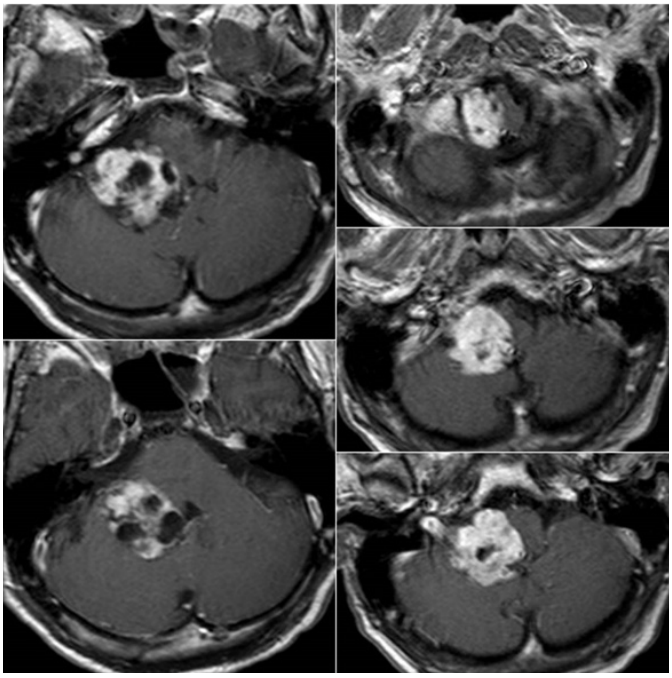

Figure 1. MRI showing a right cerebellopontine angle tumor with heterogeneous contrast enhancement.

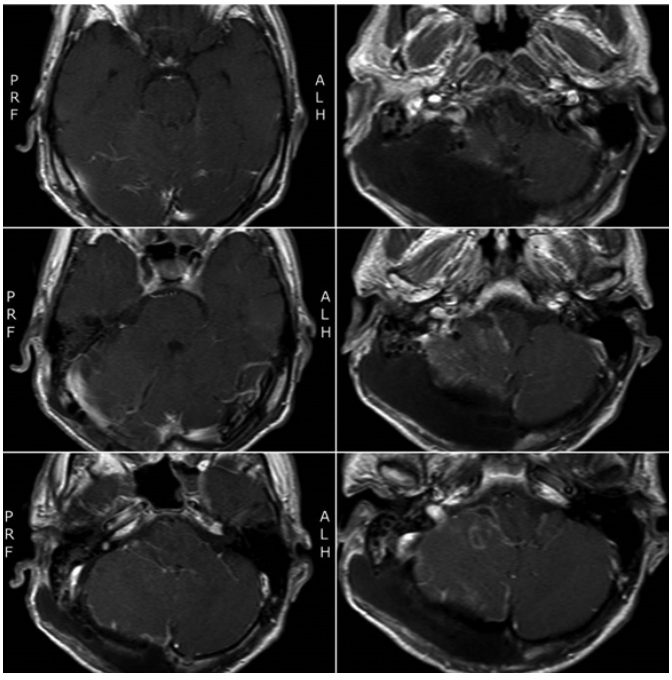

Figure 2. Post-operative MRI revealing total resection of the tumor.

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The patient presented an uneventful recovery and was discharged home on the fifth post-operative day. On the six-month follow-up, he showed improvement of the left leg weakness and was able to walk without any assistance. No shunt procedure was required. pathoanatomical analysis confirmed the diagnosis of choroid plexus papilloma (CPP), WHO grade I (Figure 3).

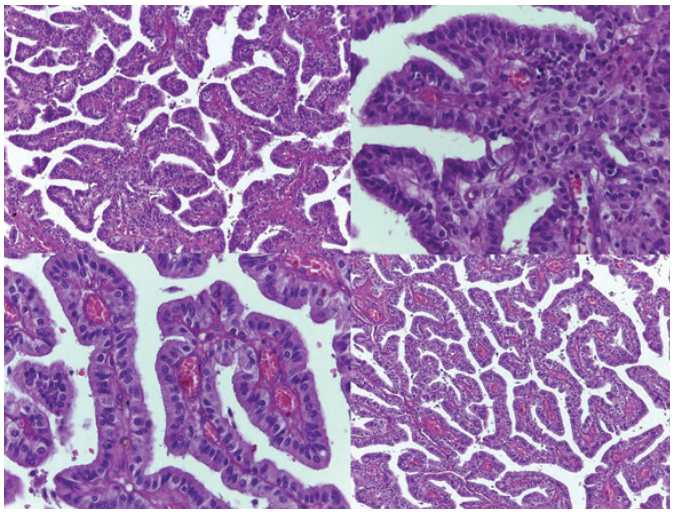

Figure 3. Hematoxylin and eosin staining showing typical papillary structure.

Discussion

CPP is a benign tumor with slow-growing rate and low malignant evolution[7,8]. The supratentorial location is the most common site of the CPP origin, mainly in children[9-14]. When all age groups are considered together, the most frequent location is the lateral ventricle (43%-67%), followed by the fourth ventricle (24%-39%) and third ventricle (9.5%-11%)[16,17]. The cerebellopontine angle is an extremely unusual location, accounting for only 9% of all CPTs and these are almost exclusively found in adult patients[9-14]. In the literature, female gender presents a predominance[14, 16, 18] and the most common symptoms observed are headache and eighth cranial nerve deficits[13, 14, 16, 18]. In the present study, our patient showed no cranial nerves deficit, but headaches and left leg weakness probably resulting from motor long tract compressing.

Hydrocephalus is a common and life-threatening presentation of CPP of the cerebellopontine angle[13]. More than 50% of patients present some degree of ventricular dilation and this event may result from two reasons: I) obstruction of the cerebrospinal fluid (CSF) pathways and II) CSF overproduction[13, 14]. Cerebellopontine angle's CPP are usually unable to produce high amounts of CSF due to decreased blood supply compared with intraventricular tumors [13, 14]. Hydrocephalus resolves after complete tumor removal and in most cases does not require permanent shunt procedures [14]. In our case, the patient presented fourth ventricle displacement and mild temporal horn dilation, which resolved completely after surgery.

Surgery remains the most important and effective treatment for patients with CPPs with a survival rate of nearly 100% in more recent series[15, 16, 19]. Total resection was associated with a significant increase in both progression-free survival and overall survival compared with subtotal resection[14, 15]. Luo et al.[14] performed suboccipital retrosigmoid approach or far-lateral suboccipital approach to resect CPP of the cerebellopontine angle, and reported that using those surgical routes, there would be a wider field of vision to dissect the adhesion to the brainstem and the cranial nerves and to control bleeding. Additionally, when tumors were more inclined to protrude inferiorly into the foramen magnum region and to be adhesive to lower cranial nerves, the far lateral suboccipital approach seems to be the preferred access for preservation of neurologic function and total resection [14]. In the present case, as most of the tumor was directed inferiorly and compressing the brainstem and lower cranial nerves, we decided to perform a right far-lateral approach in order to expose as much tumor as possible and prevent excessive cerebellar retraction.

In conclusion, CPP of the cerebellopontine angle is an extremely rare neoplasm that challenges the neurosurgeon ability to deal with tumors in close relation to lower cranial nerves. Appropriate neurosurgical route, surgeons experience and adequate anatomical knowledge of important neural and vascular structures are fundamental to safely remove the CPP of the posterior fossa. Suboccipital far-lateral craniotomy gives enough view with minimal retraction to manage the tumor of this patient here reported.

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