Cavernous Sinus Chondromas: Report of two Cases and Literature Review

Condromas do Seio Cavernoso: Relato de dois Casos e Revisão de Literatura

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ABSTRACT

Background: Chondromas are benign tumors with slow growth pattern that may develop along the body. Intracranial chondromas are rare, 0.1 to 0.3% of all intracranial tumors, usually in parasellar region. Few papers reported cavernous sinus chondromas.

Case Description: We reported two cases of cavernous sinus chondromas. Two patients presented diplopia, one facial hypoesthesia and one presented extension to the petroclival region. Surgical approaches were frontotemporal combining presigmoid/middle fossa. Literature was reviewed concerning incidence, clinical symptoms, treatment and outcome. Both cases achieved complete tumor resection without postoperative complication. Literature review confirms their rarity, clinical symptoms are related to deficits of cranial nerves passing through the cavernous sinus and cure can be achieved through radical surgical removal.

Conclusion: Cavernous sinus chondromas are rare benign lesions with insidious growth. Adequate knowledge of cavernous sinus anatomy, microsurgical experience, intraoperative monitoring of cranial nerves and neuronavigation allows radical tumor removal with no additional neurological deficits.

Key words: Chordomas; Cavernous Sinus; Surgery; Treatment; Literature Review

RESUMO

Introdução: Condromas caracterizam-se como tumores benignos com padrão de crescimento lento. Condromas intracranianos são raros correspondendo a cerca de 0,1 a 0,3% de todos os tumores intracranianos. Ocorrem com maior frequência na região paraselar. Na literatura encontram-se poucos relatos de condromas no seio cavernoso. Relato de Casos: Dois casos de condromas no seio cavernoso são relatados. Ambos apresentavam diplopia, um deles com hipoestesia facial e o outro com extensão à região petroclival. As abordagens cirúrgicas utilizadas foram: acesso frontotemporal combinado à abordagem pré-sigmoide e acesso de fossa média. Uma revisão da literatura considerando a incidência, os sintomas clínicos, tratamentos utilizados e os resultados é apresentada. Nos dois casos foi realizada uma ressecção total da lesão sem complicações pós-operatórias. Obteve-se uma melhora dos déficits pré-operatórios em ambos pacientes. Conclusão: Condromas no seio cavernoso são lesões benignas raras com crescimento insidioso. O conhecimento adequado da anatomia do seio cavernoso, somado a experiência microcirúrgica, monitorização intra-operatória de nervos cranianos e neuronavegação possibilitam a remoção radical do tumor sem déficits neurologísticos adicionais. Nossos casos confirmam os achados da literatura em relação a raridade destes tumores no seio cavernoso, aos sintomas clínicos relacionados a déficits de nervos cranianos e a possibilidade de cura através de remoção cirúrgica radical.

Palavras-chave: Cordomas; Seio Cavernoso; Cirurgia; Tratamento; Revisão de Literatura

INTRODUCTION

Surgical removal of cavernous sinus (CS) lesions is a neurosurgical challenge. Postoperative complications, surgical strategy and grade of tumor resection are directly related to the histology and extension of the tumor.

CS is a complex location of venous channels located laterally to the sella turcica. It has 5 walls: lateral, medial, superior, posterior and anterior. The lateral wall is composed by two layers: an outer and thicker (dura propria) and a thinner inner membranous layer. They are slightly attached to each other and are easily dissected. The inner layer separates the lateral wall and its contents from the venous channels where passes the internal carotid artery (ICA) and the abducens nerve. Oculomotor, trochlear and ophthalmic (V1) nerves, and occasionally the maxillary nerve (V2), cross between both layers. The inner layer continues to cover the content of the superior orbital fissure in periorbit, while the outer layer continues as temporal fossa dural covering.
Meningiomas are the most frequent tumors in this region and most papers in the literature focus the treatment of these lesions. Few articles discuss the management of non-meningeal lesions of CS. Chondromas are tumors that can develop in different regions of the body, but are rare in the cranium corresponding to 0.1 to 0.3% of all intracranial tumors. They occur mainly in the skull base, especially in the parasellar region involving the CS in different manners. CS chondromas are rare and few reports are found in the literature. In this article two cases of CS chondromas are presented and the literature concerning incidence, clinical symptoms, radiologic findings, treatment and outcome of these tumors are reviewed.

**METHODS**

Two cases with histological diagnosis of chondromas involving the CS are presented. Clinical aspects, radiologic findings, surgical approaches and outcome are discussed. Preoperatively the patients were evaluated with magnetic resonance imaging (MRI) to define location and extension of the tumor. Involvement of neurovascular structures within the CS was defined with digital subtraction angiography (DSA) in one patient and computed tomographic angiography (CTA) in the other. Postoperative Control MRI and CT examinations were performed in both cases. The frontotemporal approach with “peeling” and opening of the lateral wall of CS was done in one case. A combined presigmoid/extradural middle fossa approach was used in the other patient. Intraoperative monitoring of cranial nerves III, V, VI, VII & VIII and neuronavigation (Brainlab Vector Vision®) were performed.

A literature review through PubMed with the terms: ‘cavernous sinus chondroma’, ‘cavernous sinus tumors’, ‘skull base chondromas’ and ‘cranial chondromas’ was carried out.

**CASE REPORTS**

**Case 1:** This 41-year-old woman complained of progressive diplopia starting five months before admission to our Institute. Clinical examination revealed a left VI cranial nerve palsy. MRI showed a lesion occupying the left CS with hypointense signal in T1, hyperintense with hypointense areas in T2 and irregular contrast enhancement (Figure 1). The CTA showed anterior displacement of left ICA.

The tumor was approached through a frontotemporal craniotomy. After opening of the sylvian fissure tumor a protrusion was observed in the lateral wall of cavernous sinus. Neuronavigation (Brainlab Vector Vision®) was used to identify the internal carotid artery (ICA) within the CS (Figure 2).

The lateral CS wall was opened at the supratrochlear and Parkinson’s triangles exposing the whole lesion (Figure 3 a-c). The neurovascular structures within the CS were not infiltrated and total removal of the tumor was possible (Figure 3 d).

**Case 2:** This 25-year-old woman complained of diplopia and left facial hypoesthesia. Clinical examination revealed III and VI cranial nerve palsy and hypoesthesia in ophthalmic (V1) and maxillary (V2) nerves dermatomes. MRI showed a large lesion in left posterior CS with extension to the ipsilateral petroclival region with hyperintense signal in T2W and hypointense with homogeneous contrast enhancement in T1W (Figures 4 a,b). Digital subtraction angiography (DSA) showed ICA displacement and absence of pathologic vascularization (Figure 4 c).

The tumor was approached through a presigmoid retro labyrinthine craniotomy. The CS component was removed in two ways: one following the trigeminal nerve and its branches extending the dural opening until the lateral wall of CS; other via subttemporal extradural with resection of the residual component (Figure 5). To avoid CSF leakage the skull base was reconstructed using vascularized muscle and fascial flaps.

**RESULTS**

Complete tumor removal was achieved in both cases. The lesions were solid, greyish and slightly adherent, allowing dissection from the neurovascular structures. The first patient presented normal abducens nerve function the day after surgery and the second patient recovered trigeminal and abducens nerves function six months postoperatively. Searching the literature through PubMed only few reports on cavernous sinus chordomas could be found.
and 60 years of age with a peak in the third decade with no gender predominance\(^4,13,22\). Histologically, they have the same characteristic regardless of their location, being characterized by lobules of hyalinized cartilage that usually contain one neoplastic chondrocyte per lacuna. At the periphery of the tumor, the cartilage undergoes enchondral ossification and the centre frequently calcifies\(^11,13,22\).

There are many theories concerning the genesis of chondromas. The most accepted is that they arise at the base of the skull from residual rests of primordial cartilage in basilar synchondroses entrapped during endochondral ossification\(^11,18,22,31\). Due to this origin they are usually located at the site of sphenopetrosal, petro-occipital and spheno-occipital synchondrosis\(^19,22,31,38\). Other theory purposes that chondromas originate from metaplasia of meningeal fibroblasts or perivascular mesenchymal tissue, explaining their presence in other locations such as cerebral parenchyma, dura and different bone sites\(^11,13\).

Yasargil\(^40\) classified intracranial chondromas into three groups: a) chondromas from paranasal sinuses; b) chondromas from convexity and intracerebral structures and c) basal chondromas (in synchondrosis regions).

Sarcomatous degeneration usually occurs in 16 to 23% of chondroma in patients with Mafucci’s syndrome\(^21,24,27,31,38\). CS chondromas occupy the postero-superior venous space, causing displacement of ICA. Tumors originated in posterior clinoid process can displace the carotid antero-inferiorly\(^37\). One of our cases caused an anteromedial displacement of the ICA suggesting a sphenopetrosal origin, around the foramen lacerum (Figure 6). The other patient presented an antero-superior and lateral ICA displacement suggesting an origin in the petro occipital synchondrosis immediately inferior to the dorsum sellae.

### Clinical features

Chondromas are slow growing tumors usually presenting as large lesions at the time of diagnosis. They can develop from the childhood and present clinical manifestations only after the third decade of life\(^11,31\). The most frequent clinical symptoms of CS tumors are headache 50-88%, visual impairment 39-55%, diplopia 24-60%, facial numbness 22%, facial pain 20%, exophthalmos 15% and amaurosis 15%. With tumors located in
the lateral CS wall visual impairment is less frequent\textsuperscript{9,10,25,32,35}. The most affected cranial nerves are the optic 46%, the maxillary 37% and the abducens 35%\textsuperscript{10}.

**Image findings**

The image findings of chondromas are variable. CT usually shows a circumscribed lesion with regular or irregular limits and heterogeneous density\textsuperscript{4,11,13}. Typically, they have a slight and delayed contrast enhancement\textsuperscript{1,4,6,11,13,36}. Bone destruction is found in 50% of the cases and calcifications in over 60\%\textsuperscript{4,11,13,22,30,31}. These calcification areas may increase with time regardless of tumor growth\textsuperscript{12}.

In MRI they are hypointense in T1W and heterogeneous and hyperintense in T2W. Calcifications have curvilinear shape and may appear with hyperintense signal in T1W depending of the mineralization grade\textsuperscript{3}. In T2W calcification areas usually have low signal while solid components have an intermediate signal. These lesions may present variable contrast enhancement patterns\textsuperscript{1,11,30}. The neurovascular structures are usually displaced but not invaded\textsuperscript{30,31}. DSA shows an avascular tumor (Figure 4 c)\textsuperscript{6,29}.

The radiologic findings of chondromas are not typical and may suggest the presence of a meningioma\textsuperscript{11}. Tumors arising in the lateral wall of CS have an oval shape displacing the ICA without stenosis, while purely intracavernous and invasive tumors tend to involve or infiltrate this vessel causing stenosis\textsuperscript{10,35}. Schwannomas are iso- or hyperintense in T1W, hyperintense in T2W with different contrast enhancement depending on the size of the lesion (smaller tumors enhance homogeneously)\textsuperscript{10,29,35}. Meningiomas usually are hyperintense in T2W presenting intense and early heterogeneous enhancement in T1W may have adjacent hyperostosis, dural tail (in 60\% of the cases) and can cause stenosis of the ICA\textsuperscript{29,35}.

Chondromas and chondrosarcomas have similar image features. Chondromas usually originate in the clivus, near the sphenoid-occipital synchondrosis. The findings in chondromas includes: bone erosion and visible calcifications in CT. In MRI they are hypointense in T1W with intense and heterogeneous contrast enhancement and hyperintense in T2W\textsuperscript{1,35}. Definite diagnosis of chondromas is based on the anatomopathological findings\textsuperscript{13}.

![Figure 1. A. MRI (T2 sequences) showing a left CS tumour with hyperintense signal. B and C. MRI (T1 with contrast) showing enhancement with hypointense round areas which can correspond to calcification (chordomas are usually hypointense in T1).](image1)

![Figure 2. Intraoperative neuronavigation: images of the approach and location of ICA.](image2)
When possible complete surgical removal is the treatment of choice and may be curative in cases of intracranial chondromas\textsuperscript{6,11}. Sepehrnia et al.\textsuperscript{32} described different approaches to remove CS tumors: intradural anteromedial (with subfrontal craniotomy), anterolateral (with anterior clinoidectomy), lateral (through Parkinson triangle) and posterolateral (combined retrosigmoid and subtemporal). The most used surgical approach to treat these lesions is the pterional craniotomy with or without orbitozygomatic osteotomy\textsuperscript{7,9,10,16,20,25}. Higashida et al. partially removed a CS chondroma using a subtemporal orbitozygomatic approach\textsuperscript{17}. Tereasaka et al. completely resected a CS chordoma through an intradural frontotemporal approach with orbitozygomatic osteotomy and anterior clinoidectomy\textsuperscript{37}. Fratzoglou et al. reported subtotal removal using an intradural pterional approach\textsuperscript{14}. One of our cases underwent complete tumor removal through the frontotemporal approach without anterior clinoidectomy through a posterosuperior intradural window as purposed by Parkinson\textsuperscript{26}. Total resection was achieved in the other case by a combined presigmoid/ extradural subtemporal approach. There was no postoperative deficit. More than one procedure may be needed to obtain complete resection of theses tumors\textsuperscript{39}. Chondromas may have a calcified adherent component to neurovascular structures making total resection difficult\textsuperscript{20}. Chondromas do not respond satisfactory to irradiation\textsuperscript{4,6,11,12,30}. It has been suggested that radiotherapy may even increase the risk of malignization of these tumors\textsuperscript{46}.

**Figure 4.** A, B. MRI (T1W with contrast) showing a large lesion in the left CS. C. DSA showing lateral and superior displacement of ICA without contrast enhancement.

**Figure 5.** A. Intraoperative imaging of case 2 after sub temporal extradural resection of component related to Kawase and posterolateral triangles. B. Post-operative contrast enhanced CT showing gross total resection.

**Figure 6.** CTA images showing anterior displacement of the ICA (A) without lateral dislocation (B).
Preoperative cranial nerve deficits usually improves, and even faster, if complete resection is achieved\textsuperscript{9,32,37}. Oculomotor palsy often improves in 6 months of follow-up\textsuperscript{9,10,37}. In this series both patients recovered completely from their preoperative deficits.

Complications related to surgery for non-meningiomas CS tumors include CSF leakage, hematomas, ischemic complications, hydrocephalus, diabetes insipidus and pulmonary thromboembolism\textsuperscript{9,25}. Recurrence is usually not observed after complete resection of intracranial chordomas\textsuperscript{11,12,17,22}.

**CONCLUSION**

Intracranial chordomas are rare benign tumors presenting insidious growth. They may invade the CS. Radical tumor removal with preservation of cranial nerves and the ICA is possible. Very few cases of CS chordomas have been described in the literature. In this paper two additional cases of CS chordomas are presented. Total tumor removal with no postoperative complication and complete improvement of preoperative symptoms were achieved. The authors consider that the surgical approach should be tailored to each case. The use of intraoperative cranial nerve monitoring and neuronavigation are helpful to preserve the involved neurovascular structures. Complete surgical resection is the treatment of choice.

**REFERENCES**


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