Falx Cerebri Giant Chondroma – Case Report

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Abstract

Intracranial chondromas are rare skull base tumors, but those arising from the falx cerebri are extremely rare. Of our knowledge, there have been reported no more than seventeen cases. Surgical gross removal of falx chondroma is associated with good outcome. We bring into focus the case of a large falcine chondroma and discuss its diagnosis, surgical management and prognosis.

Keywords: Brain tumor; Giant chondroma; Falx cerebri; Surgery

Introduction

Chondromas are infrequent brain tumors, with a very low incidence, estimated at 0.2-0.5% of all intracerebral tumors [1,2]. The skull base synchondrosis represents the usual origin of chondromas [3], but, exceptionally, they develop from the convexital dura mater or the falx [4,5]. We present the case of a large intracranial chondroma, which arises from the falx cerebri.

Case Report

A 46 years old female presented with a one-year history of headache, lack of concentration and motor weakness in her right lower limbs, in the last three months. On physical examination, except for a mild right hemiparesis, no other positive neurological signs were found.

Axial brain computed tomography (CT) scan showed a giant well-defined, calcified tumor, with poor contrast enhancement, developed over the corpus callosum, between frontal lobes, mainly in the left hemisphere (Figure 1A). Magnetic resonance (MR) imaging showed a 8 × 8.5 × 4.6 cm, multilobular, well-circumscribed tumor that crossed the anterior falx and extended in both frontal lobes, suggesting that its origin was the falx (Figure 1B). The tumor appeared hypointense on T1-weighted images and mixed iso- to hyper-intense on T2-weighted images. It showed no surrounding cerebral edema and minimum gadolinium enhancement. Angiogram confirmed the presence of an avascular mass that dislocated the anterior cerebral artery on both sides and lack of opacification of the anterior third of the superior sagittal sinus.

A left frontal parasagittal craniotomy was performed disclosing a large tumor that was firmly attached to the falx and superior sagittal sinus. The tumor was easily separated from the surrounding brain tissue and was sharply resected, piece by piece, with micro scissors. It was entirely removed, along with its falx attachment.

The patient’s operative course was initially uneventful and CT scans obtained postoperatively showed total excision of the tumor. The patient recovered well without any new neurological deficits. A postoperative cerebrospinal fluid (CSF) leak with rhinorrhea due to the...
opening of the left frontal aeric sinus needed reoperation and re-
sealing of the aeric sinus. The patient was discharged without any
complaint and pathological finding. Contrast CT scans obtained
postoperatively showed total excision of the tumor. Six months after
surgery the patient remained neurologically normal and has resumed
her job. A follow-up MRI obtained 6 months post-surgery revealed left
frontal atrophy and no evidence of recurrence (Figure 1C).

Gross examination revealed a firm mass, with a plain convex
countour and random cystic spaces on cut-surface. Microscopic
examination disclosed well-differentiated cartilaginous lobules formed
of mildly pleomorphic chondrocytes and localized ossification. Mitotic
figures, atypical cells, multinucleation were not identified. Immunohistochemical examination revealed positive reactions to
S-100 protein (Figure 1D).

Discussion

Intracranial chondromas are rare benign tumors, which, usually,
grow as solitary lesions, although an association with Ollier disease [6]
and Maffucci’s syndrome [7] has been reported. The majority of
patients are between 20 and 60 years of age with a high frequency
around the third decade [1,8]. Although a slight female preference has
been reported, there is no gender predominance [9]. Our patient is a
48 years old female.

The first case of intracranial chondroma was reported in 1851, but
only in 1982 the first surgical resection was reported. Intracranial
chondromas mostly arise from the skull base, from ectopic hyaline
cartilaginous rests trapped within suture lines [10,11]. Exceptionally
rare, chondromas developed from the falx cerebri [12,13], with an
estimated incidence of 5.6-6.3% of all intracranial chondromas [14].
Depending on various authors, between 14-15 cases were reported in
the literature [1,8,12-16]. In a very recent study, Zivkovic et al.
summarized 16 cases, including their, of falx cerebi chondroma [17].

Most authors agree that most of chondromas developed from
cartilaginous rests along the basilar synchondroses [18,19]. The
etiopathology of intracranial chondromas without attachment of the basal
bones is not clearly established. They develop due to heterotrophic
chondrocytes, metaplasia of perivascular mezenchymal cells or
meningeal fibroblasts or migration caused by trauma or inflammatory
process [18,20,21]. Heterotypically located embryonal cartilaginous
remnants are most likely the origin of falx chondromas [8]. Without a
head trauma history, we can support the idea that the chondroma in
our report had its origin in intradurally located embryonal
cartilaginous rests in the falx cerebi. However, we cannot prove this
theory.

The clinical features of chondromas are different from those of
meningiomas. The patients usually present with long time history of
symptoms and signs because of the slow growing nature of these
tumors [22]. At time of surgery, most of the tumors are usually large.
The clinical presentation of the tumor is mild and non-specific and
mostly depends of anatomic location. Manifestations of the tumor are
related to dysfunctions that are secondary to either, local parenchymal
compression, epileptic seizures or increased intracranial pressure
[23,24]. Our patient presented intermittent headaches, lack of
concentration, and three months history of difficulty in naming objects
and mild right hemiparesis. It has been reported that the mean
diameter of convexity chondromas is 6 cm [16]. In our case, the size of
the tumor was 8.5 × 8 × 5 cm.

Brain chondroma is almost typical on the neuroimaging studies. According to Lacerte et al. intradural chondromas have two distinct
CT-scan presentation [8,25]. Type 1 (classical) is more frequent, and
reveals mixed density with minimal or moderate gadolinium
enhancement, whereas type 2 is less common, usually presenting a
central hypodense area due to cystic degeneration. The tumor’s appearance on CT scan is variable, probably reflecting differences in
the degree of calcification [18,26,27]. The most frequently encountered
imaging features of falx chondromas on CT scan and MRI include a
well circumscribed and demarcated mass, with mild to moderate
patchy gadolinium enhancement, and a minimal peritumoral edema
[7,21]. Calcifications, with hyperostosis and erosion of the surrounding
bone, are encountered in 60-90% of cases [21]. In our case, the tumor
has a central cystic degeneration and peripheral thick calcification
both demonstrated on CT or MRI studies. The signal intensities where
mixed of non-specific on both T1 and T2 weighted images.

A falx chondroma should be differentiated from a falx meningioma
using contrast-enhanced imaging studies and angiography [9,13,14].
Usually, chondromas exhibit no enhancement or present late and
patchy contrast enhancement, in contrast to meningiomas, which show
an early, intense and homogenous enhancement [28]. Cerebral
angiography is probably the best diagnostic method to differentiate
these two tumors. Chordomas are usually avascular, whereas
meningiomas exhibit the late capillary tumor blush, due to the feeders
from meningeal arteries [13,14,28]. Differential diagnosis of falcine
lesions mostly includes falx meningiomas but is not limited to
oligodendroglioma, glioblastoma multiforme, teratoma or chondrosarcoma.

Treatment of these tumors is entire tumor removal and resection of
the attached falx, since they are well demarcated and there is little
adherence to surrounding brain structures [29,30]. We performed a
complete resection of a large falcine chondroma attached to the dura
mater and anterior third of the superior sagittal sinus.

The long-term prognostic is good when complete resection of the
falx chondroma is achieved, and no recurrence should be expected
[14,26,31]. Hardy et al. [31] reported a patient with a survival period of
44 years after complete removal of a convexity chondroma. After
subtotal resection, malignant degeneration of the tumor rest into
chondrosarcoma has been reported [3,9]. Therefore, in cases with
subtotal resection of the chondroma, long-term imaging follow-up
with cerebral CT-scan or MRI may be necessary to early detect local
invasion or recurrences [32]. Radiation therapy is not advice, since
chondromal tumors are radioresistant and can undergo a malignant
transformation [8,33].

Conclusion

Intracranial chondromas are rare, slow growing tumors that arise
from the skull base and tend to recur after treatment. Very few cases of
falcine chondromas have been reported. They are well-circumscribed
tumors, with calcification, intratumoral cystic degeneration, lack of
enhancement and no or minimal perifocal edema. Surgical gross
removal of falx chondroma is associated with good outcome and any
recurrence should raise suspicions for malignant degeneration into
chondrosarcoma. If total surgical removal is achieved, the long-term
prognosis for falx chondroma could be excellent.
References