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Case Report Open Access

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.

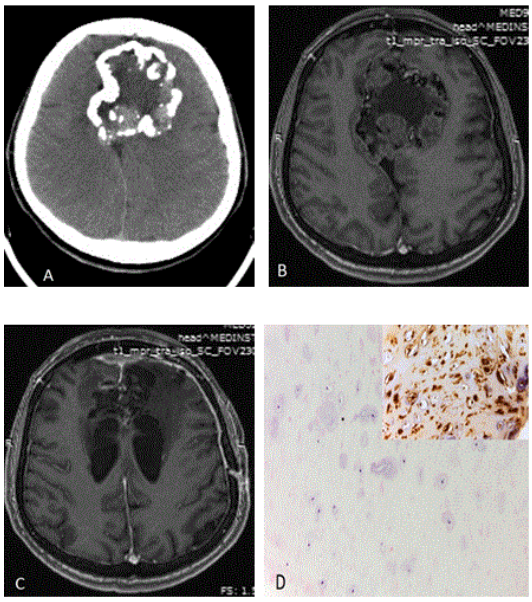

Figure 1: (A) The axial non-contrast CT scan showed a giant, calcified, well-circumscribed tumor, with low density center and hyperdense, calcified wall, located at the anterior falx; (B) Axial gadolinium-enhanced MRI showed a 8 × 8.5 × 4.6 cm, multilobular, well-circumscribed tumor that passed the falx anteriorly and expanded in both frontal lobes, displaying a patchy enhancement and without surrounding cerebral edema; (C) The six months follow-up postoperative axial T1-weighted enhanced image showed total removal of the tumor and no recurrence; (D) Hystopathology of the tumor. Microscopic aspects show chondromas lobules with well-differentiated chondrocytes and absence of nuclear atypism. Inset: chondrocytes showing positive reaction to anti S-100 protein (Hematoxylin-Eosin, original magnification 200x; inset – 400x).

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 contour and random cystic spaces on cut-surface. Microscopic examination disclosed well-differentiated cartilaginous lobules formed of midly 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 cerebri chondroma [17].

Most authors agree that most of chondromas developed from cartilaginous rests along the basilar synchondroses [18,19]. The etiology 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 remanants 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 cerebri. 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 and 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 chordoma 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 radioresistent 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

1. Berkmen YM, Blatt ES (1968) Cranial and intracranial cartilaginous tumors. Clin Radiol 19: 327-333.
2. Kretzschmar HA, Eggert HR, Beck U, Furmaier R (1989) Intracranial chondroma: case report. Surg Neurol 32: 121-125.
3. Mapstone TB, Wongmorengkolrit T, Roessman U (1983) Intradural chondroma: A case report and review of the literature. Neurosurgery 12: 111-114.
4. Ghogawala Z, Moore M, Strand R, Kupsky WJ, Scott RM (1991) Clival chondroma in a child with Ollier's disease: Case report. Pediatr Neurosurg 17: 53-56.
5. Kretzschmar HA, Eggert HR, Beck U, Fürmaier R (1989) Intracranial chondroma: Case report. Surg Neurol 32: 121-125.
6. Traflet RF, Babaria AR, Barolat G (1989) Intracranial chondroma in a patient with Ollier's disease: case report. J Neurosurg 70: 274-276.
7. Chakrabortty S, Tamaki N, Kondoh T (1991) Maffucci's syndrome associated with intracranial enchondroma and aneurysm: Case report. Surg Neurol 36: 216-220.
8. Colpan E, Attar A, Erekul S, Arasil E (2003) Convexity dural chondroma: A case report and review of the literature. J Clin Neurosci 10: 106-108.
9. Kurt E, Beute GN, Sluzewski M, van Rooji WJ, Teepen JL (1996) Giant chondroma of the falx: Case report and review of the literature. J Neurosurg 85: 1161-1164.
10. Ahyai A, Spoerri O (1979) Intracerebral chondroma. Surg Neurol 11: 431-433.
11. Padhya TA, Athavale SM, Kathju S, Sarkar S, Mehta AR (2007) Osteochondroma of the skull base. Otolaryngol Head Neck Surg 137: 166-168.
12. Yang PJ, Seeger JF, Carmody RF, Fleischer AS (1986) Chondroma of falx: CT findings. J Comput Assist Tomogr 10: 1075-1076.
13. DeCoene B, Gilliard C, Grandin C, Nisolle JF, Trigaux JP, et al. (1997) Unusual location of an intracranial chondroma. Am J Neuroradiol 18: 573-575.
14. Nakazawa T, Inoue T, Suzuki F, Nakasu S, Handa J (1998) Solitary intracranial chondroma of the convexity dura: Case report. Surg Neurol 40: 495-498.
15. Erdogan S, Zorludemir S, Erman T, Akgulergin M, Idan F, et al. (2006) Chondromas of the falx cerebri and dural convexity: Report of two cases and review of the literature. J Neurooncol 80: 21-25.
16. Fountas KN, Stamatiou S, Barbanis S, Kourtopoulus H (2008) Intracranial falx chondroma. Literature review and case report. Clin Neurol Neurosurg 110: 8-13.
17. Zivkovic N, Berisavac I, Markovic M, Milenkovic S (2014) Falx chondroma with hyperostosis of the skull: A case report. Srp Arh Celok Lek 142: 464-467.
18. Dutton J (1978) Intracranial solitary chondroma, case report. J Neurosurg 49: 460-463.
19. Matz S, Israeli Y, Shalit MN (1981) Computed tomography in intracranial supratentorial osteochondroma. J Comput Asist Tomogr 5: 109-115.
20. Acampora S, Troisi F, Fusco G (1982) Voluminous intracranial chondroma. Surg Neurol 18: 254-257.
21. Berkmen YM, Blatt ES (1968) Cranial and intracranial cartilaginous tumors. Clin Radiol 19: 327-333.
22. Sharafeddine H, Elias E, Jabbour M, Najjar MW (2014) Giant convexity chondroma of the dura matter presenting with epilepsy. Austin J Neurol Disord Epilepsy 12: 311-313.
23. Laghmari M, Metellus P, Fuentes S, Adetchessi T, Dufour H, et al. (2007) Cranial vault chondroma: A case report and literature review. Neurochirurgie 53: 491-494.
24. Patel A, Munthali L, Bodi I (2009) Giant cystic intracranial chondroma of the falx with review of literature. Neuropathology 29: 315-317.
25. Awan LM, Niaz A, Vohra AH (1998) Chondroma of cerebral falx: A rare intracranial diagnosis. J Coll Physicians Surg Pak 25: 771-773.
26. Ozgen T, Pamir MN, Akalan N, Bertan V, Onol B (1984) Intracranial solitary chondroma, case Report. J Neurosurg 61: 399-401.
27. Yang PJ, Seeger JF, Carmody RF (1986) Chondroma of falx: CT findings. J Comput Assist Tomogr 10: 1075-1076.
28. Tanohata K, Maehara T, Aida N (1987) Computed tomography of intracranial chondroma with emphasis on delayed contrast enhancement. J comput Assist Tomogr 11: 820-823.
29. Nakayama M, Nagayama T, Hirano H, Oyoshi T, Kuratsu J (2001) Giant chondroma arising from the dura mater of the convexity. J Neurosurg 94: 331-334.
30. Abdelhamid K, Camras LR, Nijenshom EM, Rosseau GL, Cerullo LJ (1996) Intracranial chondroma arising from the cranial vault: CT and MR appearance. J Comput Assist Tomogr 20: 556-558.
31. Hardy RW Jr, Benjamin SP, Gardner WJ (1978) Prolonged survival following excision of dural chondroma: Case report. J Neurosurg 48: 125-127.
32. Tin-Chou T, Chen Y, Swei-Ming L, Sheng-Hong T (2005) Chondroma of the falx: Case report. Tzu Chi Med J 17: 269-272.
33. Valdueza JM, Freckmann N, Herrmann HD (1990) Chondromatosis of the choroid plexus: case report. Neurosurgery 27: 291-294.