



Case Report

Intracranial Falcine Chondroma: A Case Report and Review of The Literature

Merih Güray DURAK¹, Gülen Gül NİFLİOĞLU¹, Mehmet Mustafa AKIN², Emel ADA³,
Ümit ACAR⁴, Tülay CANDA¹

¹Dokuz Eylül University Faculty of Medicine, Department of Pathology, Izmir, Türkiye

²Mustafa Kemal University Faculty of Medicine, Department of Pathology, Hatay, Türkiye

³Dokuz Eylül University Faculty of Medicine, Department of Radiology, Izmir, Türkiye

⁴Dokuz Eylül University Faculty of Medicine, Department of Neurosurgery, Izmir, Türkiye

Summary

Intracranial chondromas are rare neoplasms that are most commonly located at the skull base. Localization of this tumor in other areas, such as the convexity, falcine area, brain parenchyma or ventricles is less common. These slow-growing benign tumors have non-specific symptoms, and variable radiological findings. Complete surgical removal is the treatment of choice, and long-term prognosis is favorable. Herein, we describe a rare case of intracranial falcine chondroma in a 29 year-old female, and review the pertinent literature.

Key words: Intracranial tumor, Chondroma, Falx cerebri

Intrakranial Falsin Kondrom: Olgu Sunumu ve Literatür Derlemesi

Özet

İntrakranial kondromlar sıklıkla kafatası tabanından gelişen nadir görülen neoplazmlardır. Bu tümörlerin konveksite, falks serebri, beyin parenkimi ya da ventrikül gibi bölgelerde görülmesi çok daha seyrek. Yavaş büyüyen bu benign tümörlerin semptomları non-spesifik olup radyolojik bulguları değişkendir. Komplet cerrahi eksizyon sonrası uzun süreli prognozları oldukça iyidir. Burada, 29 yaşında kadın hastada saptanan nadir görülen bir intrakranial falsin kondrom olgusu sunularak, konuyla ilgili literatür bilgileri derlenmiştir.

Anahtar Kelimeler: İntrakranial tümör, Kondrom, Falks serebri

INTRODUCTION

Chondroma is a benign cartilaginous tumor that may arise in any part of the body⁽⁵⁾. Intracranial chondroma, which was first described by Hirschfield in 1851, is rather rare^(3,5,6). The incidence of intracranial chondroma has been reported to be approximately 0.2-0.3% of all intracranial neoplasms^(3,5,8). They can present as solitary masses, or in association with systemic chondromatoses, such as Ollier disease and Maffucci's syndrome⁽¹⁻¹⁰⁾. Most of these tumors are located at the skull base, especially the sellar and

parasellar regions or paranasal sinuses with extension into the cranial cavity. Tumors arising from the falcine area, convexity, leptomeninges, parenchyma, ventricles, cavernous sinus or choroid plexus are less common^(1,2,4,5-8). Herein, we present a rare case of solitary intracranial chondroma of the falx cerebri in a 29 year-old female, and review the pertinent literature.

CASE PRESENTATION

A 29 year-old female presented with a 3 month history of dizziness and persistent headache that was resistant to analgesics. She also had a sudden decrease in her

vision initiated about 3 months ago. Her medical history was otherwise unremarkable on admission. Neurological examination was within normal limits except for bilateral papilledema. Cranial computerized tomography revealed a 7x5x4 cm well-defined extraaxial mass lesion in the left fronto-parietal region (Figure 1). The mass had isodense to hypodense central core and a thin peripheral rim of contrast enhancement. There was a shift to the right side and no edematous changes. Because the patient had initially applied to a state hospital in another city, and was referred to our hospital after her radiologic imaging tests were performed, we could not obtain the images of her magnetic resonance (MR) scan. However, we have learned from the radiologist that her MR scan demonstrated the lesion to have a heterogenous signal with intermediate to low intensity, and no peritumoral edema.

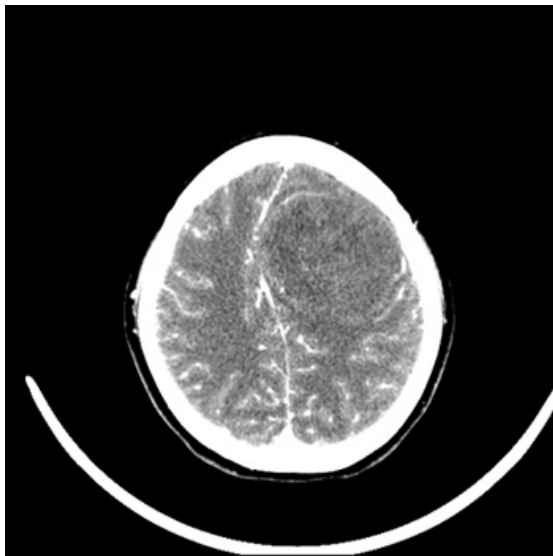


Figure 1: Contrast-enhanced axial CT scan shows an oval-shaped well-defined left fronto-parietal extraaxial mass lesion

The tumor was completely excised with its dural attachment via left craniotomy in our hospital. Macroscopically, the tumor was a slightly lobulated, gray-white, firm mass lesion, 7x6x4 cm in size and 88 gr in weight. The dura attached to the tumor was approximately 7x3.5 cm in size. Histopathologic examination of the tumor showed mature hyaline cartilage consisting of scattered lacunas containing single chondrocytes within a dense chondroid extracellular matrix (Figure 2). There was no evidence of pleomorphism, multinucleation or mitotic activity. Immunohistochemically, tumor cells were positive for S-100 protein (Figure 3) and vimentin (Figure 4). Morphologic and immunohistochemical findings were consistent with chondroma. The patient remains free of disease after 13 months of clinical follow-up.

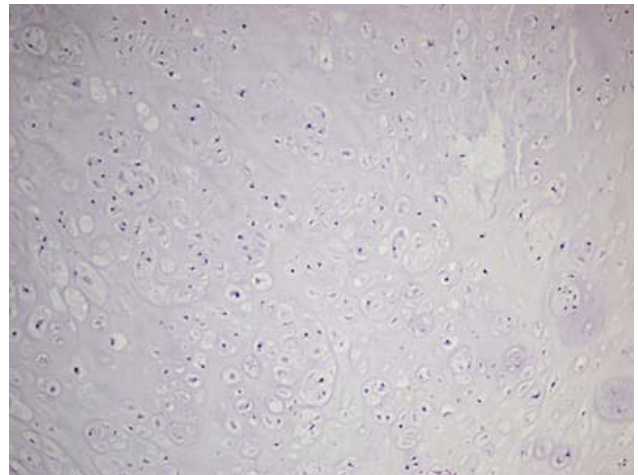


Figure 2: Microscopic appearance of the well-differentiated chondroid tumor showing mature hyaline cartilage with no pleomorphism or mitotic figures (H&E, x100).

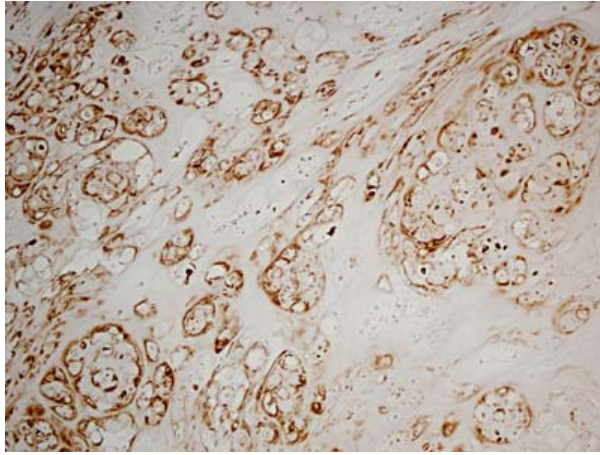


Figure 3: Immunohistochemically, tumor cells were positive with S-100 protein (x100).

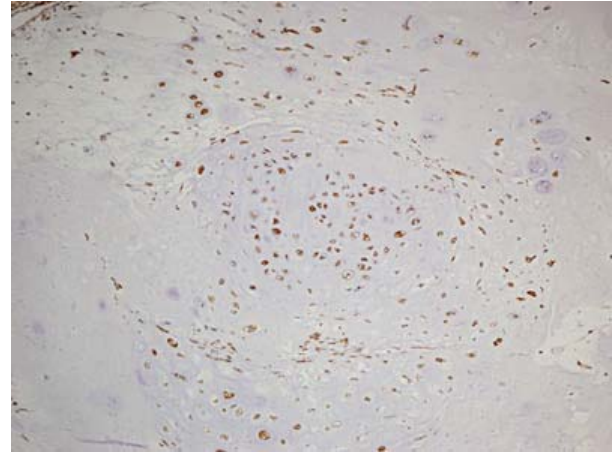


Figure 4: Immunohistochemically, tumor cells were positive with vimentin (x100).

DISCUSSION

Intracranial chondromas are rare tumors that account for approximately 0.2-0.3% of all intracranial neoplasms. They usually occur in young adults with peak prevalence in the third decade. However, patients ranging from 15 months to 60 years of age have been reported^(1-6,8,9). There is no sex predilection^(1,8). They can grow as solitary tumors, or as components of Ollier disease and Maffucci's syndrome⁽¹⁻¹⁰⁾. Some cases have been reported to arise at sites of previous trauma⁽¹⁾. The disease-related chondromas have a tendency to be hypercellular with cytologic atypia. It is also well known that patients with Maffucci's syndrome have increased risk of malignant change^(8,9). Very rarely, chondromas presenting concomitantly with other glial tumors have been reported in the literature⁽⁶⁾.

The majority of intracranial chondromas develop from the cartilage found in the basilar synchondroses at the skull base. Tumors arising in the dural portions, leptomeninges, brain parenchyma, ventricles or choroid plexus are less common^(1,2,4,5-8). Several theories have been proposed in the literature explaining the pathogenetic mechanism of development of convexity or falxine chondromas. These include heterotopic

chondrocytes, cartilaginous metaplasia of meningeal fibroblasts, abnormal multipotential mesenchymal dural cells or their differentiated cellular descendants, and traumatic displacement of cartilage^(1,3,5,6). However, the exact pathogenetic mechanism is still uncertain.

The clinical presentation of chondromas is non-specific. The symptoms are usually associated with the localization of the tumor. The most common symptoms are due to either increased intracranial pressure such as headache, papilledema, or to focal neurologic deficits such as new onset seizures^(1,2,5,6,9). Because chondromas are slow-growing tumors, they may stay silent with no clinical symptoms for many years, and may reach very large sizes at presentation^(5,6). It has been reported that the mean diameter of these tumors is 6 cm, and their mean weight is 170 gr⁽⁶⁾. Recently, Patel et al. have described a giant cystic intracranial chondroma of the falx measuring 12x10x7 cm and weighing 480 gr⁽⁸⁾.

Neuroimaging of these tumors are non-pathognomonic. The most frequently encountered imaging features include a well-circumscribed mass with calcifications in 60-90% of cases, associated hyperostosis, erosion and destruction of surrounding bone in 50-60%

of cases, and usually mild to moderate, patchy contrast enhancement^(1,4,6,8,9). Falcine or convexity chondromas typically show no or minimal peritumoral edema, which demonstrates the slow-growing, benign nature of these tumors⁽⁸⁾. Cerebral angiography has revealed avascularity in nearly all published cases in the literature⁽⁶⁾.

The differential diagnosis includes craniopharyngioma or meningioma when the tumor is predominantly destructive and only partially calcified; chordoma when the calcified tumor extends near the clivus and cerebellopontine angle; and acoustic neuroma, meningioma, epidermoid or metastasis when the tumor is noncalcified and causes destruction of the petrous apex⁽⁹⁾.

Complete surgical resection together with the dural attachment is the treatment of choice in these tumors^(1-3,5-7). Because chondromas are radioresistant tumors and radiotherapy may increase the risk of malignant transformation, post-operative radiation therapy is not advised in cases of subtotal resection^(2,3,5-8). After total resection, long term prognosis is excellent with no recurrences reported so far. However, malignant degeneration may occur after subtotal resection, and should always be a matter for consideration especially in recurrent cases^(2-5,6,8).

Correspondence to:

Merih Güray Durak

E-mail: merihguray@yahoo.com

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