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Case Report

Intracranial Chondrosarcoma in a 22-Years Old Woman: Report of A Case

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Abstract

Background: Intracranial chondroid tumors are consist of less than 0.16% of all primary brain neoplasms. They are classified as mesenchymal, classic and myxoid according to their cytoarchitecture. They usually arise at the skull base from the cartilaginous synchondroses and the pluripotent mesenchymal cells of the overlying dura mater.

Case: We present a 22-year old female patient, who had history of seizures for 10 years and recent increase in the frequency of the seizures. Cranial magnetic resonance imaging (MRI) demonstrated a well-demarcated, parasagittal left frontal mass, which compressed to the lateral ventricle. It was hypointense on T1-weighted and hyperintense on T2-weighted images and making an edema on the surrounding tissue. Gross total resection of the firm mass was performed with clear margins. The tumor was composed of lobules of mature-appearing hyaline cartilage. The tumor cellularity was increased, each lacuna contained one or two atypical chondrocytes which had two or more nuclei per cell. Neoplastic chondrocytes had plump and hyperchromatic nuclei. Mitotic rate was low and no atypical mitotic figures were noted. Neoplastic chondrocytes showed nuclear staining for S–100 protein, but not for pancytokeratin. Histopathologic examination revealed a classic low grade chondrosarcoma.

Conclusion: Intracranial chondrosarcomas are rare malign tumors. We report a patient with a grade I intracranial chondrosarcoma of the classical subtype without any neurologic problem after complete surgical excision. She did not received any adjuvant therapy and 28 months follow-up showed no recurrence. So it seems that, especially in the low grade chondrosarcoma, radical neurosurgical resection is the first line of treatment, and if necessary adjuvant therapy can be planned.

Keywords: Chondrosarcoma; cranium

Olgu Sunumu: 22 Yaşında Kadında İntrakranial Kondrosarkom

Özet

İntrakranial kondroid tümörler tüm primer beyin tümörlerinin %0.16'sından azını oluşturur. Sitoarşitektürlerine göre mezenkimal, klasik ve miksoid olarak sınıflandırılırlar. Sıklıkla kafa tabanınıdaki kıkırdak sinkonrozlardan ve dura materi örten pluripotansiyel mezenkimal hücrelerden gelişir. Olgu: 10 yıldır nöbetleri olan ve son zamanlarda nöbet sıklığında artış olan 22 yaşında bayan bir hasta sunduk. Kranial magnetik rezonanas görüntüleme lateral ventriküle bası yapan, iyi sınırlı, parasagital sol frontal kitle gösterdi. Kitle T1 ağırlıklı kesitlerde hipointens, T2 ağırlıklı kesitlerde hiperintens olup çevre dokuda ödem etkisi oluşturuyordu. Temiz cerrahi sınırlar ile birlikte kitlenin total rezeksiyonu yapıldı. Tümör matür görünümlü hyalen kıkırdak lobüllerinden oluşuyordu. Tümör sellülaritesi artmış olup, her bir lakün bir veya iki atipik kondrosit içermekteydi. Neoplastik kondrositler S-100 ile nükleer boyanma gösterirken pansitokeratin ile boyanmadı. Histopatolojik değerlendirmede

klasik bir düşük dereceli kondrosarkom izlendi. Sonuç: İntrakranial kondrosarkomlar nadir tümörlerdir. Tam cerrahi çıkarım sonrası herhangi bir nörolojik problemi olmayan klasik subtipte derece 1 bir intrakranial kondrosarkom sunduk. Hasta herhangi bir adjuvan tedavi almadı ve 28 aylık takipte rekürrens görülmedi. Böylece görünmektedir ki; özellikle düşük dereceli kondrosarkomlarda radikal nörocerrahi rezeksiyon ilk tedavi seçeneğidir ve eğer gerekli olursa adjuvan tedavi planlanabilir.

Anahtar Kelimeler: Kondrosarkom; kranium

INTRODUCTION

Intracranial chondroid tumors are consist of less than 0.16% of all primary brain neoplasms[3,5,6]. They are classified as mesenchymal, classic and myxoid according to their cytoarchitecture. They usually arise at the skull base from the cartilaginous synchondroses and the pluripotent mesenchymal cells of the overlying dura mater but occasionally from the meninges along the falx cerebri, tentorium and cerebral convexity[2,5,6,7]. Mesenchymal chondrosarcomas are the most frequent histologic type at cranial location.

METHODS

The surgical specimen was fixed in 10% buffered-formalin, embedded in paraffin, and stained with hematoxylin&eosin, and reticulin. Immunohistochemistry was performed using avidin-biotin peroxidase complex method with the antibodies Ki–67 (Labvision Neomarker, 1/200, clone=SP6), S–100 (Biogenex, 1/400, clone 15E2E2) and pancytokeratin (Biogenex, 1/100, clone AE1 and AE3).

RESULTS

Macroscopically, the mass was a firm, well defined, bluish white, a lobulated nodular mass, which was 4.5x4x2 cm in diameter. It was grey-white homogeneous and chondroid in appearance on the cut surface. Histopathologically, the tumor was composed of lobules of mature-appearing hyaline cartilage. The tumor cellularity was increased, each lacuna contained one or two atypical chondrocytes which had two or more nuclei per cell. Neoplastic chondrocytes had plump and hyperchromatic nuclei (Figure 2). Mitotic rate was low and no atypical mitotic figures were noted. Neoplastic chondrocytes showed nuclear staining for S–100 protein, but not for pancytokeratin. A final diagnosis of intracranial chondrosarcoma was rendered based on histologic features including increased cellularity, low mitotic rate, focal cytological atypia and nuclear S–100 reactivity.

CASE PRESENTATION

We present a 22-year old female patient, who had history of seizures for 10 years and recent increase in the frequency of the seizures. She had no known medical problems. On her initial evaluation, she had normal physical findings. Neurologic examination revealed a well-oriented patient with intact cranial and motor nerves and reflexes. Cranial magnetic resonance imaging (MRI) demonstrated a well-demarcated, parasagittal left frontal mass, which compressed to the lateral ventricle. It was hypointense on T1-weighted and hyperintense on T2-weighted images and making an edema on the surrounding tissue (Figure 1). Preoperative diagnosis was a meningioma radiologically.

Surgery was performed with a left interhemispheric approach and gross total resection of the firm mass which was attached to the dura with a small vascular peduncle was performed with clear margins. The patient did well with an uncomplicated recovery. Although histopathologic examination revealed a classic low grade chondrosarcoma, she did not receive any additional adjuvant treatment.

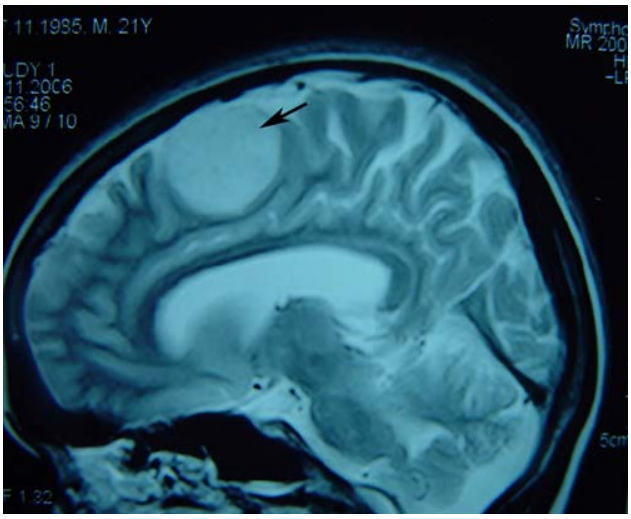

Figure 1: *MRI demonstrated a well-demarcated, parasagittal left frontal mass*

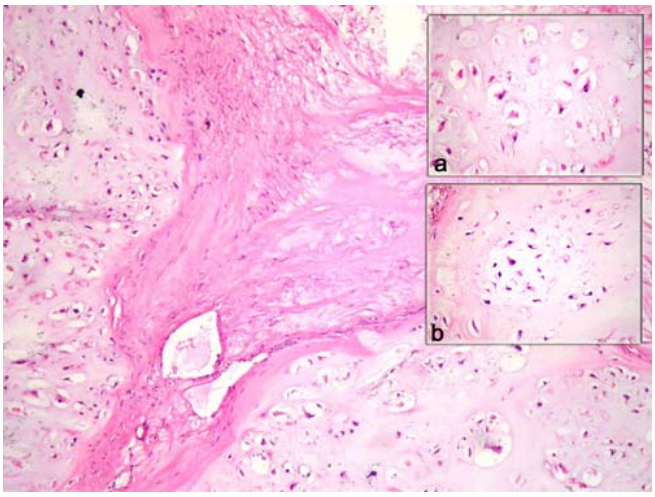

Figure 2: *The tumor was composed of lobules of mature-appearing cartilage, a-b: Neoplastic chondrocytes had plump and hyperchromatic nuclei.*

DISCUSSION

Chondroid tumors are predominant lesions of the axial skeleton but may infrequently have an extraskeletal origin. Intracranial chondroid tumors are consist of less than 0.16% of all primary brain neoplasms[3,5,6]. They may be associated with Maffucci's syndrome[2]. There is no sex predilection according to Korten et al[4].

First clinical signs are caused by oculomotor dysfunction[4]. But patients can also present with a long-standing history of headaches, signs related to increased intracranial pressure, cranial nerve palsies, hemiparesis, dizziness, tinnitus and sensory disturbances of the face[2]. 75% of intracranial chondrosarcomas originate at the skull base[5]. Our patient was presented with a longstanding history of seizures.

They are classified as mesenchymal, classic and myxoid according to their cytoarchitecture. Classic chondrosarcomas had predominance in the 6th to 7th decades[3,5]. Chondrosarcomas are subdivided into three histological grades

according to cellular density, nuclear size, mitotic rate and multiple nuclei of in each lacuna[3,4]. Mesenchymal chondrosarcomas are most malignant as illustrated by a strong tendency to intradural and cerebral growth[1] and microscopically show a distinctive appearance with a densely cellular stroma of anaplastic mesenchymal cells and hyaline cartilage. They predominate in the younger age group[3,4,5]. Myxoid chondrosarcomas, although common in soft tissues, are extremely rare within the cranium[5].

The chondrosarcoma described in this report is of the classical type. There was a lobulated architecture and the cellularity was increased. Each lacuna contained one or two atypical chondrocytes which had two or more nuclei per cell. Neoplastic chondrocytes had plump and hyperchromatic nuclei with a few mitotic figures and showed nuclear positive staining for S–100 protein, and negative for pancytokeratin, differentiating this tumor from meningioma. Imaging findings of a classical chondrosarcoma may resemble that of other neoplasms, and the differential diagnosis includes meningioma, hemangiopericytoma, metastases and vascular malformations[5]. In our case, preoperative radiologic diagnosis is also a meningioma. The certain features that differentiate a classical chondrosarcoma from a meningioma are; higher signal intensity on the T2-weightened images than that of a meningioma, islands of low signal intensity despite lack of gross calcification on all sequences due to well-differentiated cartilage, frequent preservation of the pial barrier, ‘honeycomb' enhancement and most importantly, lack of perfusion[5].

Radical excision is the mainstay of treatment. Radiotherapy is mostly used as adjuvant therapy[1]. 57% of the patients, who had treated by the excision of the mass, had recurrence by the 32 months average time[4].

We report a grade I intracranial chondrosarcoma of the classical subtype without any neurologic problem after complete surgical excision. She did not received any adjuvant therapy and 28 months follow-up showed no recurrence. So it seems that, especially in the low grade chondrosarcoma, radical neurosurgical resection is the first line of treatment, and if necessary adjuvant therapy can be planned.

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