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# Primary Intracranial Myxoid Chondrosarcoma: Case Report

Xu Su[1], Weifang Yang[2], Liguo Qi[3], Limei Lou[4], Lili Jiang[5], Bo Zhu[6], Huai Chen[7], Jian Yin[7], Qingping Lv[7], Fuchuang Qin[7], Xuhong Jin[7], Kang Zheng[8], Wei Zhu[8], Song Zhang[7]*

[1]Hangzhou Emergency Center, Zhejiang Province, Hangzhou 310003, China

[2]Departments of Cardiology, Hang zhou Red Cross Hospital/Zhe Jiang Chinese Medcine and Western Medicine Integrated Hospital of Zhejiang Province, Hangzhou 310003, China

[3]Departments of Neurosurgery, Taian City Central Hospital of Shandong Province, Taian 271000, China

[4]Departments of Pathology, Hang zhou Red Cross Hospital/Zhe Jiang Chinese Medcine and Western Medicine Integrated Hospital of Zhejiang Province, Hangzhou 310003, China

[5]Departments of Dermatology, Hangzhou Third Hospital of Zhejiang Province, Hangzhou 310009, China

[6]Departments of orthopedics, Hang zhou Red Cross Hospital/Zhe Jiang Chinese Medcine and Western Medicine Integrated Hospital of Zhejiang Province, Hangzhou 310003, China

[7]Departments of Neurosurgery, Hang zhou Red Cross Hospital/Zhe Jiang Chinese Medcine and Western Medicine Integrated Hospital of Zhejiang Province, Hangzhou 310003, China

[8]Departments of Neurosurgery, Huashan Hospital, Shanghai Fudan University, Shanghai 200040, China

***Abstract:*** **Primary intracranial myxoid chondrosarcoma (PIMCS) is extremely rare, with only seven patients previously have been reported. We present a case report of a 58-year-old woman admitted for numbness in her right face. However, there is no symptoms of increased intracranial pressure (ICP). Magnetic resonance imaging (MRI) revealed a well-enhanced large mass around her middle and posterior cranial fossa (Figure 1-4). A right pterional craniotomy was performed. Subtotal surgical resection was also performed, and pathology results confirmed an extraskeletal myxoid chondrosarcoma (Figure 5). Postoperative MRI showed some residual tumor, and the patient underwent radiotherapy. So far, the patient recovers well without discomfort complaints. This malignant tumor showed high rates of recurrence in previous reports. We here report another occurrence of this highly malignant and rare tumor in a patient treated using subtotal surgical excision and adjuvant radiotherapy.**

***Keywords:*** Brain neoplasms; Myxoid; Chondrosarcoma

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*Corresponding Author: Song Zhang, zhsg1049@163.com

## 1. Introduction

Intracranial primary chondrosarcoma is an extremely rare tumor of the central nervous system, its diagnosis and treatment are difficult[1-6]. Its clinical and pathological features are as follows: female is slightly more than male, the usual age of presentation varies between 20 and 40 years old, with higher incidence among males, and the lesions could be found in all skull or intracranial tissues, including muscles, skull, meninges, brain, cerebellum. The diagnosis of intracranial primary chondrosarcoma depends on histopathological examination. Mucinous chondrosarcoma is a variant of chondrosarcoma, accounting for about 5% of chondrosarcoma[7]. There are only a few cases concerning intracranial primary myxoid chondrosarcoman have been published in domestic and foreign literature, only 6 cases was reported by foreign literature[1-4]. Only one case has been reported in China[5], this article reports another case of PIMCS occurred in the right middle posterior fossa .

## 2. Case report

A 58 years old woman was suffered from right-sided facial numbness for half of one month. Neurological and physical examinations revealed no abnormalities, GCS score were 15, double pupils were equal roundness same size, diameter 3mm, reaction to light was normal, muscle tension and muscle strength of extermities has no change. Preoperative images demontrated a tumor located on the right posterior fossa, which uncertain to judge whether benign or not (Figure 1, 2, 3). The differential diagnosis frequently includes trigeminal schwannoma or meningioma. The patient underwent subtotal surgical resection to remove the mass, using a pterional approach. Cerebrospinal fluid was released after cutting the dura, via raising the frontotemporal lobe, removed the tumor by sub-block. The tumor, which appeared to originate from the brain tissue, was fish-shaped with colored gray, and the margin between the brain parenchyma and supporting tissue was unclearly distinguishable.

 

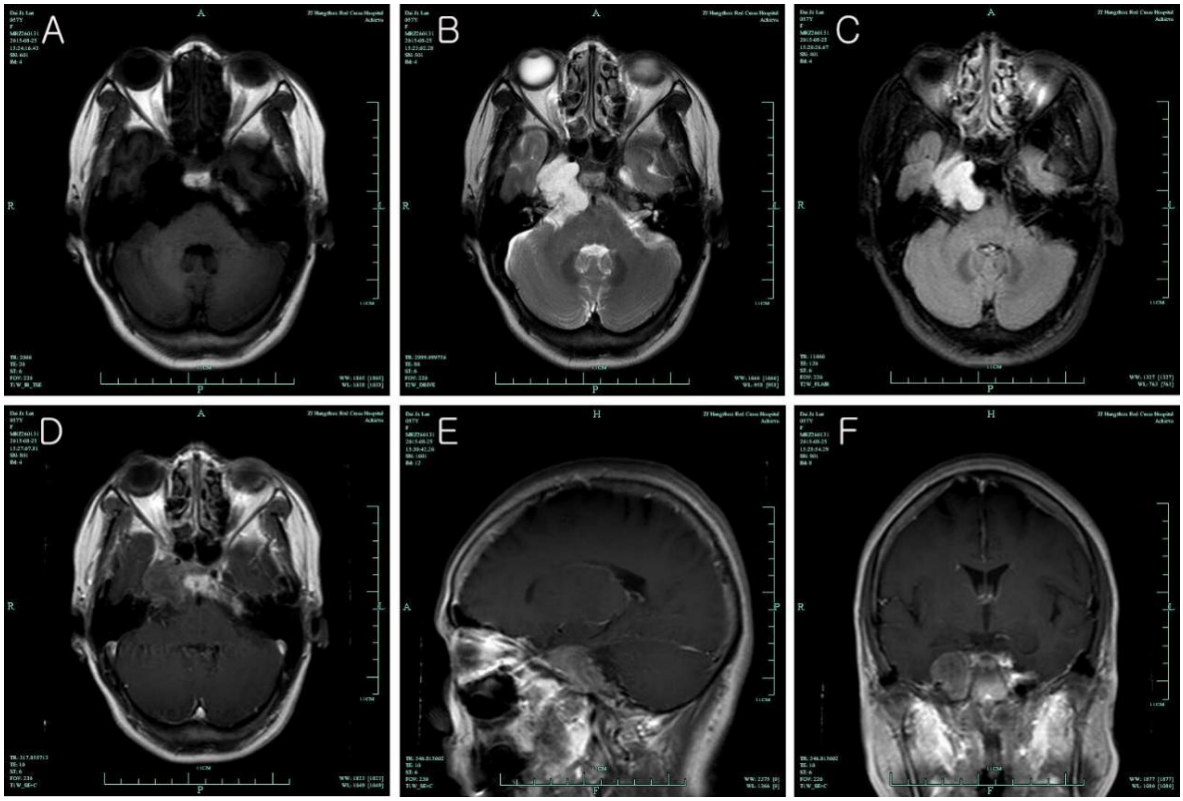


**Figure 1. Preoperative MRI: the right of petrous bone rock showed abnormal signal lesions, which presented as hypointense on T1WI(A), hyperintense on T2WI(B), and hyperointense on DWI(C), the lesion size was about 3.3*2.7cm. The lesion' border revealed irregular with lobuated appearance, seemed dumbbell-like growth, with variable low to morderate intensify after enhancement(D-F).**

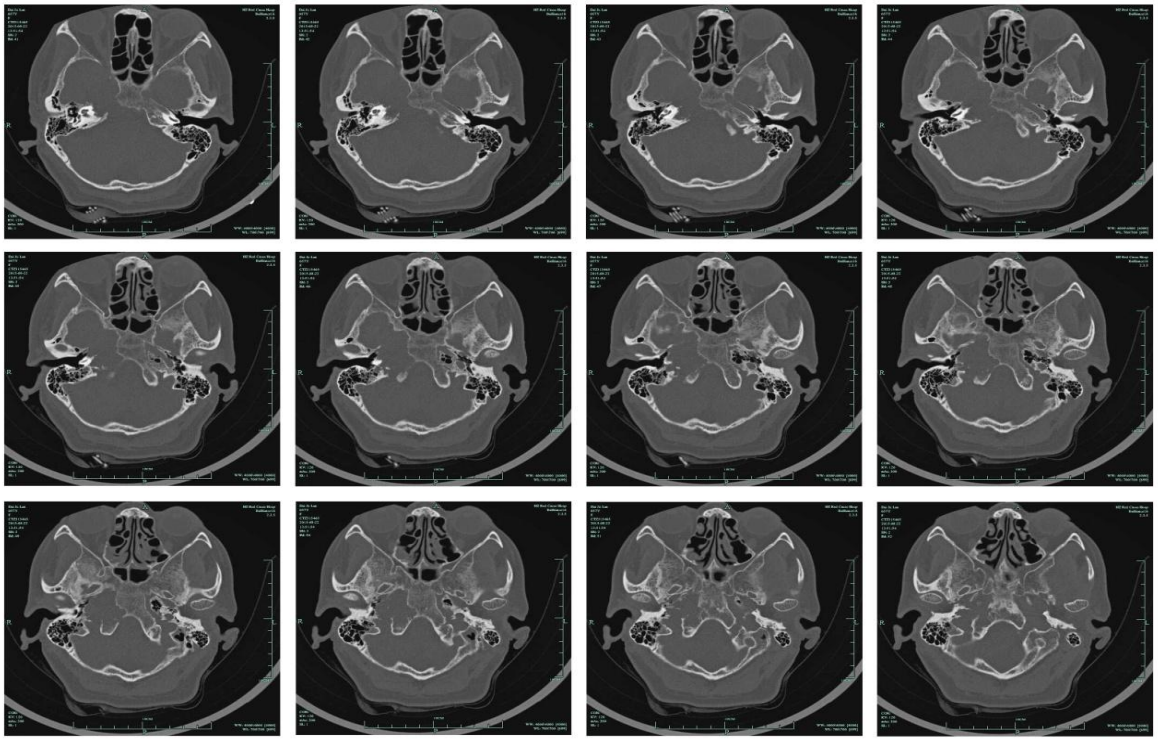

**Figure 2. Internal auditory canal CT: the right of petrous bone rock and the slope of the bone is destroyed. A little of residual bone spine are existed and internal of the lesion is full of cystic degeneration.**

 

The tumor was hypovascularized, soft. And definitive open repair with bone flap. No significant complications were found except the right frontal pattern had been disappeared (Figure 4), and the patient recovered well after surgery. Microscopically, the tumor cells revealed lobulated appearance with irregular, which extensively distributed in a myxoid background. Cartilage differentiation had been seen from the tumor. Immunohistochemistry findings supported thediagnosis of myxoma (positive staining withS-100, VIM, P53 and ß-Catenin) (Figure 5). The consultant note of pathologic from Affiliated Tumor Hospital of Fudan University was that (right middle posterior fossa, mid-skull base) Chondrosarcoma that one part of the area for the I-level, and the other for the II-level.

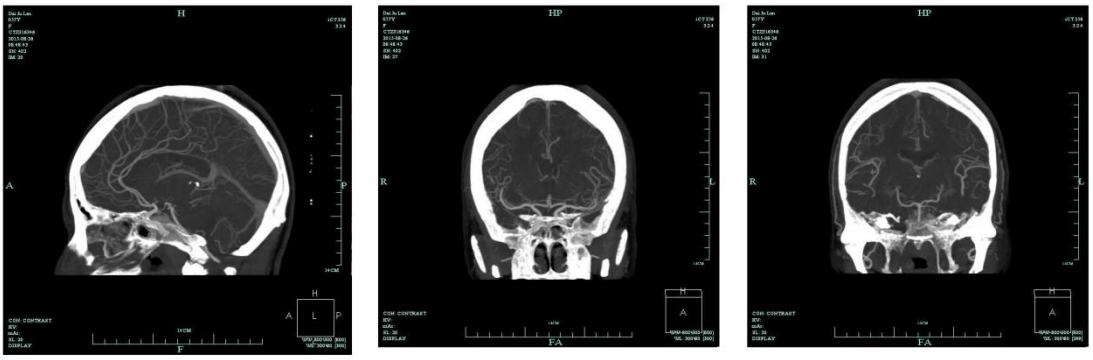

**Figure 3. Cerebral artery CTA : The right internal carotid artery is pushed forward.**

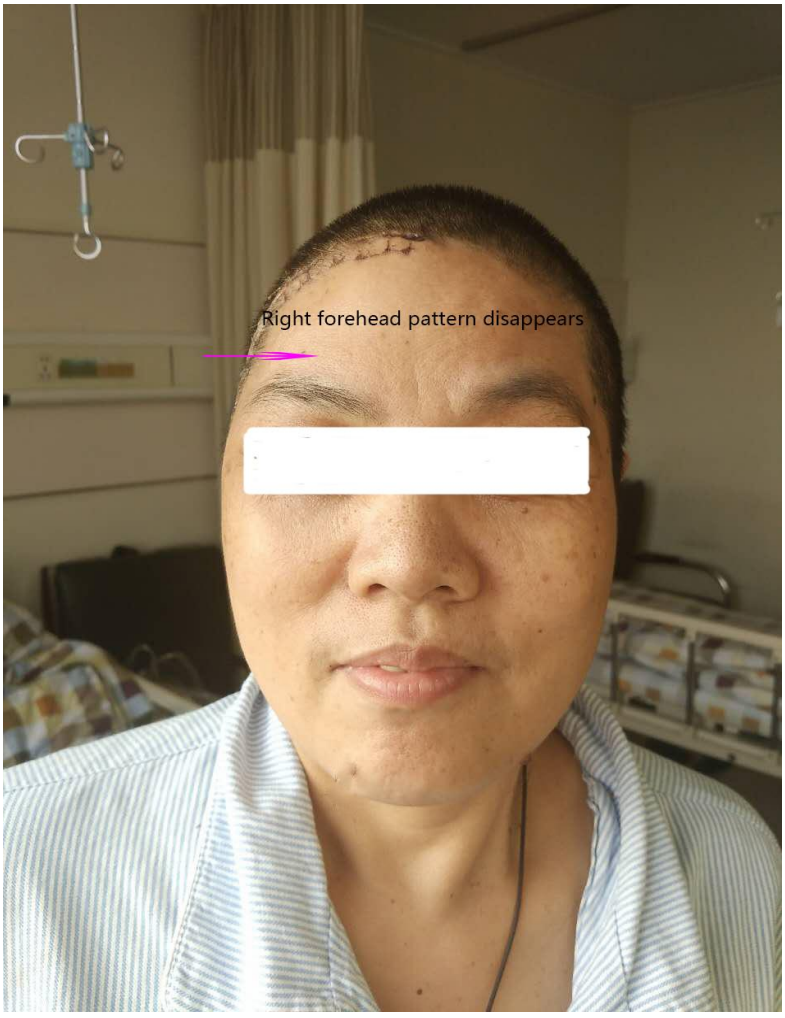


**Figure 4. Postoperative patients recovered well except for the only symptom that right frontal pattern disappeared.**

 

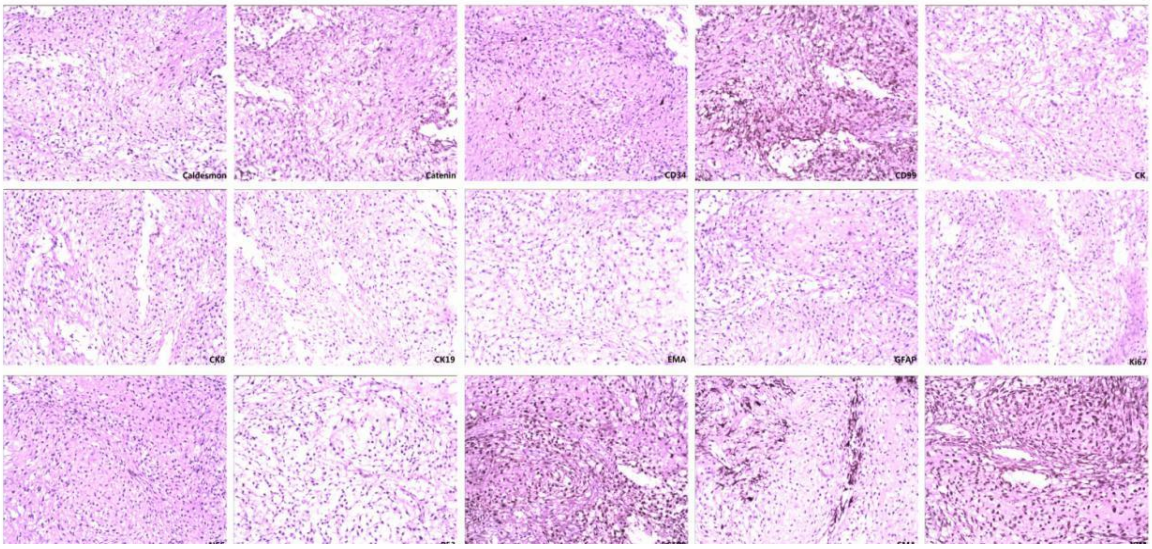


**Figure 5. The Report of Section immunohistochemistry and pathology: (Right middle posterior fossa, mid-skull base) the tumor cells revealed lobulated appearance with irregular, which extensively distributed in a myxoid background. Cartilage differentiation had been seen from the tumor. Immunohistochemistry findings supported thediagnosis ofmyxoma.**

## 3. Discussion

Intracranial chondrosarcoma has obvious mass effect on CT and MRI without clear edema zone. The features of MRI signal of it are as follows: hypointense on T1-weighted imaging (T1WI), hyperintense on T2-WI imaging (T2WI), and hypointense in diffusion weighted imaging (DWI), irregular borders with visible lobes, mild to moderate enhancements, absorption damage changes visible in peripheral bone around the tumor. Therefore it is difficult to pre-diagnosis in clinical practice. In this paper we report this case of primary chondrosarcoma on the right side of the middle cranial fossa. The case is easily to misdiagnosed due to there is no typical radiological findings. Preoperative diagnosis of the lesion located in the right side of the middle cranial fossa is trigeminal schwannoma or meningiomas.

The radical surgery is the best treatment for PIMCS. Postoperative radiotherapy and chemotherapy are recommended because of recurrence. However, radiotherapy and chemotherapy resistance exist. The combination of rapamycin and cyclophosphamide can be improved efficacy[5].

## 4. Conclusion

Intracranial primary myxoid chondrosarcoma is a very rare intracranial malignancy. Radical surgery is the best treatment for PIMCS. Postoperative radiotherapy and chemotherapy are recommended because of postoperative recurrence. Histopathology and immunohistochemistry is a reliable basis for the diagnosis of PIMCS.

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