# CHONDROMYXOID FIBROMA, A RARE TUMOR OF THE CRANIAL VAULT: ABOUT A CASE WITH REVIEW OF THE LITERATURE

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**Résumé** : Le fibrome chondromyxoïde est une tumeur osseuse bénigne d'origine cartilagineuse, site de prédilection au niveau de la métaphyse des os longs des membres inférieurs. La localisation de cette tumeur dans le crâne est exceptionnelle, seuls 16 cas ont été rapportés dans la littérature à ce jour. Le diagnostic est difficile et repose essentiellement sur l'étude histologique. La symptomatologie est polymorphe variant en fonction de la localisation de la tumeur, de la taille et de l'extension mais reste dominée par les céphalées. Le meilleur traitement est la chirurgie, seule une excision large de la tumeur assure la guérison et la prévention des récidives. Nous rapportons notre expérience sur le cas d'une femme de 40 ans atteinte d'un fibrome chondromyxoïde frontal découvert après un traumatisme crânien.

***Mots clés :*** *Fibrome chondromyxoïde, Tumeur osseuse bénigne, Crâne, Chirurgie.*

**Abstract** : Chondromyxoid fibroma is a benign bone tumor of cartilaginous origin, site of predilection at the level of the metaphysis of the long bones of the lower limbs. The location of this tumor in the skull is exceptional, only 16 cases have been reported in the literature to date. Diagnosis is difficult and is essentially based on histological study. The symptomatology is polymorphic varying according to the location of the tumor, the size and the extension but remains dominated by headaches. The best treatment is surgery, only wide excision of the tumor ensures healing and prevention of recurrence. We report our experience about a case of a 40-year-old woman with a frontal chondromyxoid fibroma discovered after a head trauma.

***Keywords:*** *Chondromyxoid fibroma, Benign bone tumor, Skull, Surgery.*

## INTRODUCTION

Chondromyxoid fibroma (CMF) is a rare benign bone tumor of cartilaginous origin representing less than 1% of primary bone tumors and less than 0.5% of all benign bone tumors (Baujat and Attal, 2001). It was first described in 1948 by Jaffe and Lichtnestein as a polylobed tumor made up of spindle cells arranged on a chondromyxoid framework (Baujat and Attal, 2001; Bucci and Dell'Aversana Orabona, 2006).

Its preferred site is the metaphysis of the long bones, particularly at the level of the femur and the tibia. However, any bone in the body can be affected.

The aim of this article is to report a case localized at the level of the median frontal part of the cranial vault and to review the literature in order to specify the radio clinical presentation and the treatment of this tumor.

Indeed, the cranial location is extremely rare, representing 2 to 5.4% of the cases described (Wu et al., 1998) and of the 35 cases of FCM of the skull published, only 16 cases affect the cranial vault since its first description. (Morimura and Nakano, 1992; O. Barbier et al., 2011).

## CASE REPORT

A 40 years old patient, with no particular medical history, suffering for five years from frontal headaches, had a minimal head trauma wich hence the realization of radiological explorations. The patient presented with a small median frontal swelling, of hard consistency, painless on palpation. The rest of the clinical examination was normal. The biological assessment found no abnormality.

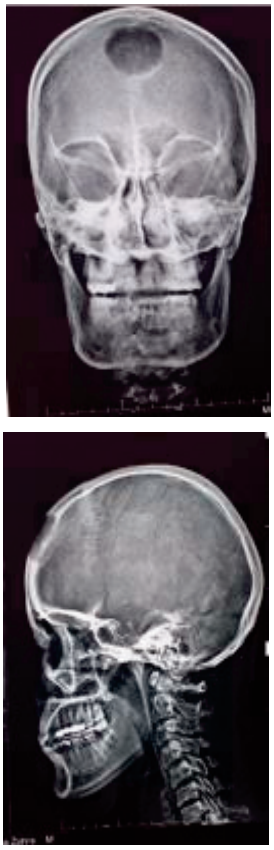

**Figure 1: standard X-ray revealed a frontal bone gap**

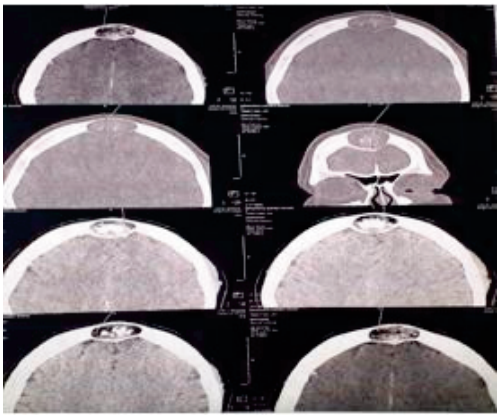

**Figure 2: cerebral CT-scanner showed an osteolytic lesion blowing the internal and external tables.**

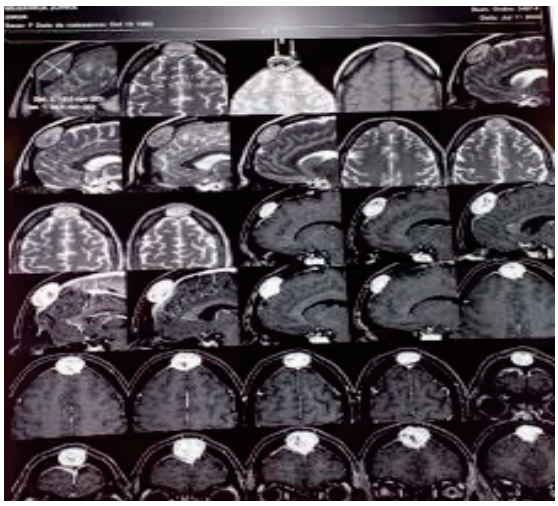

**Figure 3: cerebral MRI found a tumor tissue mass of 32x25mm developed within the median diploe of the frontal bone, it appears in hypo signal T1, moderate hypersignal T2, with intense enhancement, moderately heterogeneous by the presence of central micro calcifications.**

## DISCUSSION

FCM is one of the rarest bone tumors, accounting for less than 1% of all bone tumors and less than 0.5% of benign bone tumors (LeMay and Sun 1997; Baujat and Attal, 2001). We conducted a review of the English literature on Pubmed using the keywords: chondromyxoid fibroma, surgery, skull neoplasms and radiology. We found 16 published cases of FCM located in the cranial vault.

FCM is observed at any age, most often in patients under 30 years of age, particularly in the second decade, with a slight female predominance. Nevertheless, congenital cases have been described as well as a case reported in a 79-year-old patient. The long bones are the sites usually affected, particularly the tibia. The craniofacial location represents 2% of cases. The first symptoms are pain or a slowly growing lump. Under the microscope, as defined by the World Health Organization, chondromyxoid fibroma is a well-circumscribed tumor consisting of “lobules of spindle-shaped or stellate cells with abundant myxoid or chondroid intercellular material”. The lobules can be variable in size and subtly defined.

The center of the lobules is paucicellular, with hypercellular peripheries which may contain osteoclast-like giant cells. The bottom of the lobules can vary from fibrous to myxomatous to chondroid, although hyaline cartilage is uncommon.

Due to its variable histology, occasional soft tissue extension, and recurrence, chondromyxoid fibroma can be mistaken for a malignancy, such as high-grade myxoid chondrosarcoma or a chondromyxoma-like variant of low-grade osteosarcoma. The recommended therapeutic approach is surgical excision due to the high risk of malignancy. Although benign, local recurrence is frequent.

## CONCLUSION

FCM is a benign bone tumor that is rare in the cranial vault. Its diagnosis is based on imaging and histological examination, although cytogenetics and immuno-histo-chemistry can be helpful. It must be differentiated from a malignant tumor of the chondrosarcoma type. Therapeutic management involves extensive excision of the lesion in a healthy area, the only way to avoid local recurrences.

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