ECRONICON OPEN ACCESS

EC ORTHOPAEDICS
Case Report

Diagnosis and Literature Review of Chondromyxoid Fibroma - A Pathological Puzzle

Hemantha Kumar[1], Sushma[2], M Sathish[3]* and Kavin Kumar[3]

[1]*Senior Assistant Professor, Institute of Orthopaedics and Traumatology, Rajiv Gandhi Government General Hospital, Chennai, India*

[2]*Senior Consultant Pathologist, Apollo Cancer Institute, Teynampet, Chennai, India*

[3]*Junior Resident in MS Ortho, Institute of Orthopaedics and Traumatology, Rajiv Gandhi Government General Hospital, Chennai, India*

***Corresponding Author:** M Sathish, Junior Resident in MS Ortho, Institute of Orthopaedics and Traumatology, Rajiv Gandhi Government General Hospital, Chennai, India.

Received: May 09, 2019; **Published:** June 26, 2019

Abstract

Aim: The aim of our study is to evaluate the histological and radiological findings of CMF and difficulties in diagnosis of CMF from potential differential diagnosis.

Case Report: 38 years old female patient presented with history of limping for 5 months and on evaluation revealed an expansile osteolytic lesion in fibular head with septations and soft tissue component. Excision biopsy was done.

Results: Histological examination revealed a cellular neoplasm arranged as vague nodules in chondroid background. Occasional mitosis with giant cells in periphery without any calcification. To rule out chondroblastoma, S-100 and epithelial markers were done which was negative establishing diagnosis of chondromyxoid fibroma by exclusion.

Conclusion: Chondromyxoid fibroma is often misdiagnosed being a radiological and pathological mimicker. Histology remains key to diagnosis. En bloc resection remains the mainstay of management in expendable bone like fibula.

Keywords: *Chondromyxoid Fibroma; Benign Bone tumour; En-block resection; Fibula*

Introduction

Chondromyxoid fibroma (CMF) is a benign rare bone tumor of slow-growing nature arising from chondroblastic derivation [1-4]. Jaffe and Lichtenstein described the condition at first in 1943 [1]. In their work they differentiated CMF from chondrosarcoma, which is a much more common, but with malignant nature. Before their description in 1943, CMF has been considered as a giant-cell variant or a benign cartilaginous tumour. A myxoid element in the tumor is the defining characteristic feature.

Around 500 CMF cases has been described in the entire literature [6]. The diagnostic frequency seems to be in decline. CMF mainly affects second and third decade of young adults. Around 80% of patients are less than 36 years. The tumor is not gender specific and both males and females are affected equally, however some series showed a slight male predominance [7].

Aim of the Study

The aim of our study is to evaluate the histological and radiological findings of CMF and difficulties in diagnosis from potential differential diagnosis of CMF.

Case Report

We present a case report of a 38-year-old female who presented with difficulty in walking for the past 5 months associated with pain in the knee for the past 3 months. The pain was dull aching in nature, mild intensity. There was no diurnal variation. Physical examination showed mild tenderness. Overlying skin was normal with restricted range of movement due to pain. Blood investigations were under normal limits.

On evaluation radiologically an expansile osteolytic lesion located in the head of the fibula with thinning and septations measuring 33 x 25 x 18 mm noted. Narrow zone of transition and no matrix mineralisation noted. Cortex discontinuity noted in the lateral aspect. Joint space was normal and proximal tibial shaft appeared normal. and soft tissue component was noted as shown in figure 1.

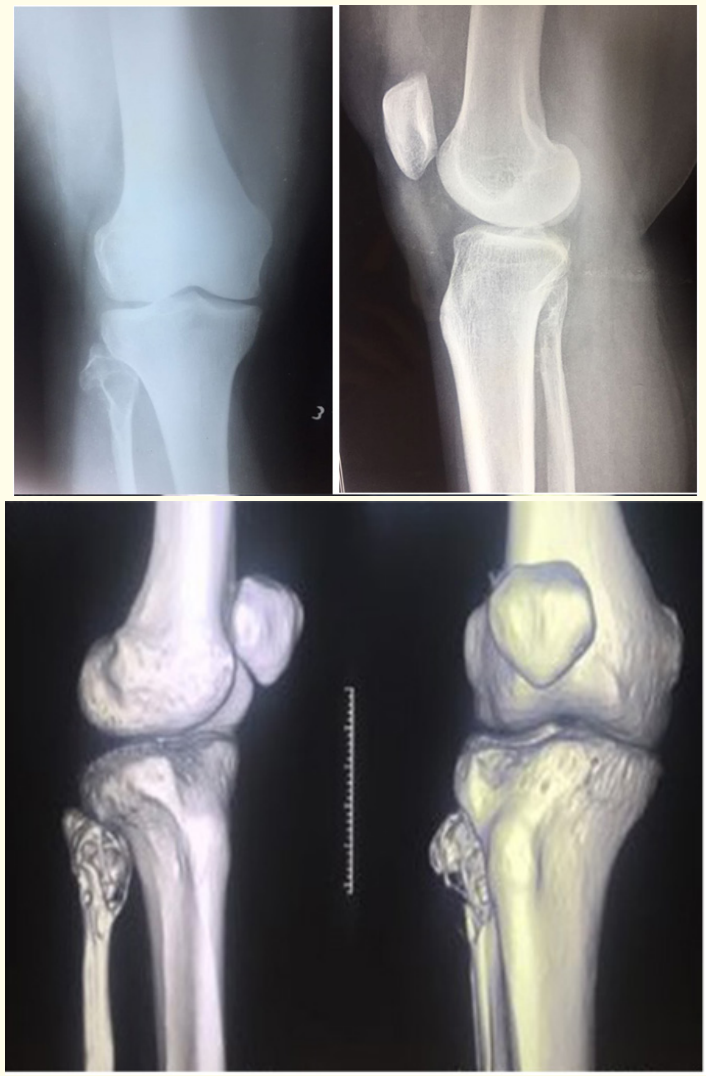

Figure 1: *Shows the X ray and CT localisation of the tumour which revealed multilobulated osteolytic lesion with scalping, thinning and expansion cortex seen in fibular head with narrow zone of transition without any matrix mineralisation.*

On MRI T1 T2 hypointense and STIR hyperintense lesion with thinning and endosteal scalloping of the cortex with focal break noted in the proximal fibula without any periosteal reaction as shown in figure 2.

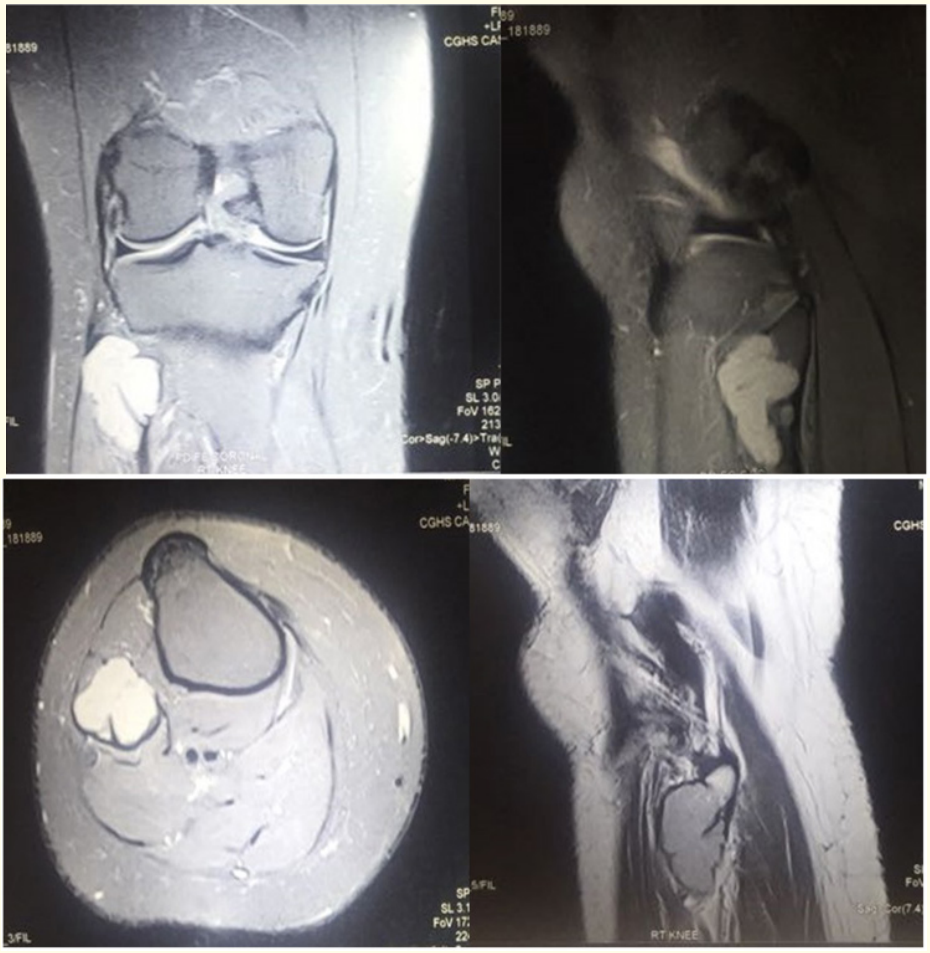

Figure 2: *Shows MRI study with T1 T2 hypointense and STIR hyperintense lesion with thinning and endosteal scalloping of the cortex with focal break noted in the proximal fibula without any periosteal reaction.*

Excision biopsy was planned and after obtaining consent from the patient an En-block excision was done. The resected specimen was sent to histopathological examination.

Results

Gross examination showed a greyish white ill-defined lesion at the epi-metaphyseal region measuring 2.5 x 1.5 x 1 cm which is focally extending into the adjacent soft tissue. On serial microscopic sectioning and examination as in figure 3 revealed cellular neoplasm which was well demarcated from the adjacent osseous tissue arranged as vague nodules in chondroid background. The cells were round to oval with vesicular nuclei and abundant eosinophilic cytoplasm and displayed well delineated cell borders. Mitosis are occasional 1 - 2 per 10 hpf.

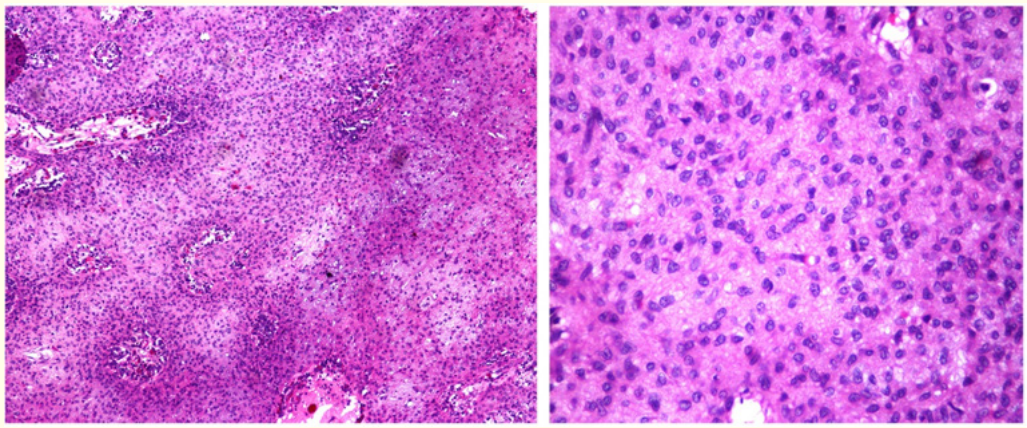

Figure 3: *Shows histology slide revealing cellular neoplasm which is well demarcated from the adjacent osseous tissue arranged as vague nodules in chondroid background. The cells were round to oval with vesicular nuclei and abundant eosinophilic cytoplasm and displayed well delineated cell borders. Mitosis are occasional 1 - 2 per 10 hpf. Giant cells are noted in the periphery of the lesion. There was no evidence of calcification.*

Giant cells are noted in the periphery of the lesion. There was no evidence of calcification. With a suspicion for chondroblastoma S-100 and epithelial markers were done which all came out to be negative establishing the diagnosis of chondromyxoid fibroma by exclusion. Although myxoid element was not much evident on histology considering the tumor to be in the earlier stages of evolution diagnosed early diagnosis of chondromyxoid fibroma was made.

Post operatively patient was put on partial weight bearing for initial 2 weeks and later resumed full weight bearing by 4 weeks. Case was followed up for 2 years without any recurrence till date (Figure 4).

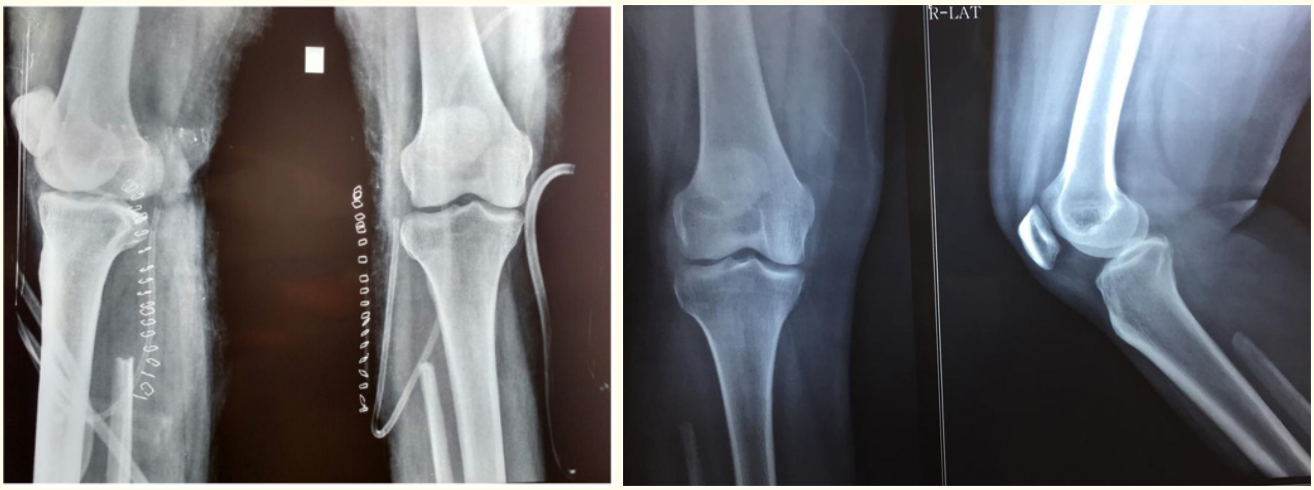

Figure 4: *Shows the post-operative X rays of the patient after en-block resection of the fibular head and 2 year follow up x ray.*

Discussion

The common location of CMF is metaphysis adjacent to the growth plate, which strengthens the theory that lesion is of cartilaginous remnant origin [4]. Similar reports of fibular CMF have been made by Karan Mane., *et al.* [8] from India and Hakan Atalar., *et al.* [9] from Turkey and Dimitri Merine., *et al.* [10] from New York who also corroborate the rarity of the tumor incidence and location in fibula.

To make a certain diagnosis, a thorough clinic radiological and pathological examination was important as CMF is often misdiagnosed as chondroblastoma or chondrosarcoma because of some pathological similarities.

A myxoid element is the defining characteristic of this tumour and hence histology remains the key in diagnosing this rare entity. However, the myxoid element was not so evident in our case which made us to diagnose this pathology by exclusion of the other possibilities like chondroblastoma by special stains and epithelial markers. Since the tumor is in initial stages of differentiation myxoid element may not be evident in our case.

Grossly, the tumor is firm, greyish-white in colour which may be lobulated or pseudo lobulated. Their structure resembles that of fibrous tissue or hyaline cartilage as in tumors like Chondroblastoma or Chondrosarcoma. The lesions often thin the cortex and rarely destroy trabecular bone. Rarely, the tumor has some areas of haemorrhagic degeneration and cystic degeneration and sometimes mimic aneurysmal bone cysts.

Many CMFs exhibit morphological features that show to be in different stages of chondrogenesis [11,12]. The most characteristic pathological fracture is the myxoid element. Giant cells are mostly seen towards the periphery of the tumor architecture.

Low grade Chondrosarcoma sometimes mimic CMF histologically, except for and lack of myxoid element and soft tissue involvement. Chondrosarcoma has peculiar demographic and radiographic features. Chondrosarcoma has peak incidence around sixth and seventh decades of life, whereas CMF occurs in the second and third decade of life.

Radiographically, location of the chondrosarcoma is central and have rich calcifications. Both Chondrosarcoma and CMF sometimes show cortical expansion. In chondrosarcomas with long standing 0r higher grade type, soft tissue involvement and cortical erosion is noted [13,14].

CMF histopathological picture shows a lobular pattern of growth with focal hyaline deposits and rare mitotic figures and even cellular pleomorphism sometimes which may be similar to that of chondrosarcoma. However, chondrosarcoma shows features like rich mucinous material, higher nuclear atypia and multiple pleomorphic or multinuclear cartilage cells. Moreover, it behaves in a more malignant fashion.

Patients with CMF may be misdiagnosed in the early stages as Giant cell tumour (GCT) of bone but they are much older than persons with CMF moreover cellular and radiographic features of GCT differ from that of CMF. GCTs are usually of metaphyseal origin with epiphyseal extension. Multinucleated Giant cells with nucleus identical to the background nucleus of the stromal cells is the major defining histologic characteristic of GCT [15]. CMFs show only a few giant cells which are also located near the periphery. GCTs show neither myxoid nor chondroid elements.

CMFs are managed surgically with intra-lesional curettage or En-bloc excision depending upon the location of the lesion [17]. Although Jaffe and Lichtenstein showed in their original description of CMF that incomplete removal did not lead to recurrence and the remnant tissue shows regression [5], some series noted recurrence rates of approximately 25% with curettage and bone grafting which may be higher in young children who are in the first or second decade of life and in myxoid predominant CMFs. Aggressive curettage of the tumor in the vicinity of physis resulted in physeal damage many cause growth arrest.

En-bloc excision is usually curative in expendable bones. Recurrence is noted in cases in which marginal excision was done. In a study by Bharma., *et al*. of 22 consecutive cases of CMF treated by intra-lesional curettage [16], 2 patients (9%) showed local recurrence within 2 years after curettage.

CMFs in axial skeleton may behave in an aggressive fashion after resection. Malignant conversion following En-block resection is extremely rare. Consequently, no case of mortality was reported due to true CMF.

Extended curettage with phenol, methyl methacrylate and liquid nitrogen have not decreased the recurrence. In occurrence of this tumour in expendable bones like fibula, En-block resection is the ideal treatment of choice to prevent future recurrence.

Radiotherapy may be considered an option for unresectable tumors [15]. Radiotherapy is generally avoided to prevent post radiation sarcoma. However malignant transformation in the absence of preceding radiotherapy has been noted which many authors believe could be a misdiagnosed chondrosarcoma.

Recurrence if at all happens mostly within 2 years, but sometimes recurrence after 19 years has also been reported [13,16-20]. Hence routine patients follow up with periodic physical and radiological examination for a minimum period of 2 years is recommended.

Conclusion

Chondromyxoid fibroma is a rare tumor of bone which is often confused with other radiological and pathological mimickers and misdiagnosed many times. Histological evaluation remains a key in diagnosing this rare pathology which is at times a diagnosis of exclusion as in our case. En bloc resection remains the mainstay of management of this tumour in expendable bone like fibula.

Ethical Approval

Consent was obtained for publication of the case details without revealing the identity of the patient.

Bibliography

1. McGrory BJ., *et al*. "Chondromyxoid fibroma". *Orthopedics* 18.3 (1995): 307-310.
2. Ralph LL. "Chondromyxoid fibroma of bone". *Journal of Bone and Joint Surgery* 44B.1 (1962): 7-24.
3. Schutt PG and Frost HM. "Chondromyxoid fibroma". *Clinical Orthopaedics and Related Research* 78 (1971): 323-329.
4. White PG., *et al*. "Chondromyxoid fibroma". *Skeletal Radiology* 25.1 (1996): 79-81.
5. Jaffe HL and Lichtenstein L. "Chondromyxoid fibroma of bone: a distinctive benign tumour likely to be mistaken especially for chondrosarcoma". *Archives of Pathology* 45.4 (1948): 541-551.
6. Blackwell JB., *et al*. "Benign bone tumours in Western Australia, 1972-1996". *Pathology* 39.6 (2007): 567-574.
7. Wu CT., *et al*. "Chondromyxoid fibroma of bone: a clinicopathologic review of 278 cases". *Human Pathology* 29.5 (1998): 438-446.
8. Mane K., *et al*. "Chondromyxoid fibroma of the distal one-third of the fibula: a rare tumour at rare site". *Clinical Trials in Orthopedic Disorders* 1.3 (2016): 126-127.
9. Atalar H., *et al*. "Chondromyxoid fibroma: an evaluation of 11 patients". *Acta Orthopaedica et Traumatologica Turcica* 41.1 (2007): 31-35.
10. Merine Elliot K., *et al*. "Chondromyxoid Fibroma of the Fibula". *Journal of Paediatric Orthopaedics* 9.4 (1989): 468-471.
11. Romeo S., *et al*. "Benign cartilaginous tumours of bone: from morphology to somatic and germ-line genetics". *Advances in Anatomic Pathology* 16.5 (2009): 307-315.

12. Bhamra JS., *et al*. "Chondromyxoid fibroma management: a single institution experience of 22 cases". *World Journal of Surgical Oncology* 12 (2014): 283.

13. Dahlin DC. "Chondromyxoid fibroma of bone, with emphasis on its morphological relationship to benign chondroblastoma". *Cancer* 9.1 (1956): 195-203.

14. Marin C., *et al*. "Juxtacortical chondromyxoid fibroma: imaging findings in three cases and a review of the literature". *Skeletal Radiology* 26.11 (1997): 642-649.

15. Durr HR., *et al*. "Chondromyxoid fibroma of bone". *Archives of Orthopaedic and Trauma Surgery* 120.1-2 (2000): 42-47.

16. Heydemann J., *et al*. "Soft tissue recurrence of chondromyxoid fibroma". *Journal of Pediatric Orthopaedics* 5.6 (1985): 725-727.

17. Hristov B., *et al*. "The role of radiation treatment in the contemporary management of bone tumours". *Journal of the National Comprehensive Cancer Network* 5.4 (2007): 456-466.

18. Gherlinzoni F., *et al*. "Chondromyxoid fibroma. The experience at the Istituto Ortopedico Rizzoli". *Journal of Bone and Joint Surgery-American Volume* 65.2 (1983): 198-204.

19. Mikulowski P and Ostberg G. "Recurrent chondromyxoid fibroma". *Acta Orthopaedica Scandinavica* 42.5 (1971): 385-390.

20. Zillmer DA and Dorfman HD. "Chondromyxoid fibroma of bone: thirty-six cases with clinicopathologic correlation". *Human Pathology* 20.10 (1989): 952-964.

Volume 10 Issue 7 July 2019