

# Intraosseus and extraosseus juxtaarticular calcification: Osteopoikilosis with synovial osteochondromatosis - an association

Parangama Chatterjee<sup>1</sup>, Jyoti Sureka<sup>1</sup>, Elizabeth Joseph<sup>1</sup>, Sniya Sudhakar<sup>1</sup>, Samuel Chittaranjan<sup>2</sup>

1. Department of Radiodiagnosis and Imaging, Christian Medical College, Vellore, Tamil Nadu, India

2. Department of Orthopaedics, Christian Medical College, Vellore, Tamil Nadu, India

\* **Correspondence:** Dr. Parangama Chatterjee, Department of Radiodiagnosis and Imaging, Christian Medical College, Vellore-632004, Tamil Nadu, India (✉ [parangamadr@gmail.com](mailto:parangamadr@gmail.com))

Radiology Case. 2009 Mar; 3(3):1-5 :: DOI: 10.3941/jrcr.v3i3.138

## ABSTRACT

Osteopoikilosis presents as round or ovoid sclerotic lesions with an appearance like enostosis on pathology. Synovial osteochondromatosis occurs due to cartilaginous metaplasia with synovial villous proliferation with calcified nodules in proximity to joints. A case of osteopoikilosis associated with synovial osteochondromatosis is described. Intraosseus and juxta osseus sclerotic bone lesions were identified on radiographs and computed tomography in a patient with knee pain. The association of osteopoikilosis with synovial osteochondromatosis is rare and to our knowledge has received little attention in the literature.

## CASE REPORT

### INTRODUCTION

Osteopoikilosis is a hereditary condition. The hallmark of this condition is numerous discrete or confluent round or ovoid calcific or ossific densities often with minimally spiculated margins, in bones, often in proximity to joints (1). These are often incidentally discovered and pathologically represent bone islands. They are occasionally associated with cutaneous lesions as well as other osteosclerotic disorders (2).

Synovial osteochondromatosis is a rare benign monoarticular arthropathy. The inciting stimulus which results in the rapid development of this synovial metaplastic process is unknown (3). An embryonic rest has been hypothesised to be the causative factor for the disease (4, 5). Trauma has also been identified as a precipitating factor (4, 6). A neoplastic basis for the cartilaginous metaplasia within the synovial villi is considered by some authors (3, 4, and 5). Although malignant transformation has been reported occasionally (4, 7, and 8) it

is controversial as to whether or not chondrosarcomas actually originate in this entity. Synovial chondromatosis most commonly involves the knee, elbow, and hip in young adults (6, 9). The association of osteopoikilosis with synovial osteochondromatosis is rare and to our knowledge has received little attention in the literature.

### CASE REPORT

A 25 year old man presented to the orthopaedics department in our institution with right knee pain and difficulty in walking, progressively increasing for 5 years. He also gave a history of fall, which happened 1 month before the onset of pain. There was no history of tingling or numbness or significant family history. Outside MRI showed tears of both menisci on the right, longitudinal tear of the lateral meniscus and radial tear of the medial meniscus, along with a tear of the lateral collateral

ligament of the right knee. The MR also showed nodular lesions in relation to the knee which were hypointense on all sequences, few of which were intra osseous and few were extra osseous. (Figures 1a-c) The patient did not have any other complaints. Examination of the right lower limb revealed wasting of the quadriceps. Rest of the clinical examination was normal. His laboratory values including erythrocyte sedimentation rate, rheumatoid factor, C-reactive protein, uric acid, serum calcium and parathyroid hormone levels were normal.

He was referred to the radiology department for a radiograph of the knee joint, which showed mild juxta-articular osteopenia and multiple small discrete, round, oval, dense calcific lesions in a symmetric distribution (Fig. 2). These lesions were distributed more in the epiphysis and metaphysis of the femur distally (Fig. 3). An AP radiograph of the pelvis did not show any abnormality. All findings were suggestive of osteopoikilosis. There were also few intra articular soft tissue density lesions with peripheral calcifications. A CT scan done thereafter confirmed the findings. (Fig. 4a and 4c). After the diagnosis, the patient was re-evaluated, and any overlooked pathology was searched. There was no pain or swelling of the joints, no muscle contractures, no soft tissue or skin changes.

He underwent an arthroscopic proceed open biopsy of the lateral condyle of the right femur to exclude osteoblastic metastases. Arthroscopy revealed typical features of proliferative frond like synovial disease with associated loose bodies as seen in synovial chondromatosis. A synovial debridement with removal of the loose bodies was done. These were however not sent for histopathological analysis by the operating surgeon in order to reduce patient expenses and because the arthroscopic appearances were typical. Microscopic histopathology revealed few thickened trabeculae merging with normal surrounding bone with no evidence of metastases, which can be seen with osteopoikilosis. We do not have archived pathological images as the femoral condyle biopsy was done to exclude osteoblastic metastases. Images were not archived as it was a negative study for the diagnostic question i.e. metastases.

## DISCUSSION

Osteopoikilosis, also called osteopathia condensans disseminata, or "spotted bone disease" is an incidentally detected disorder characterized by an abnormality in enchondral bone maturation. There is no gender predilection, men and women are equally affected (1, 9). It is usually inherited as an autosomal dominant condition. Osteopoikilosis results in multiple small densities of oval or rounded shape that are symmetrically distributed within the metaphyses and epiphyses of the long bones, in periarticular osseous regions. On histology these foci are formed by dense trabeculae of spongy bone, occasionally forming a nidus without communication with bone marrow. These appear in childhood and persist thereafter (2, 5, 10). Sites of predilection include phalanges (100%), carpal bones (97.4%), metacarpals (92.5%), foot phalanges (87.2%), metatarsals (84.4%), tarsal bones (84.6%), pelvis (74.4%), femur (74.4%), radius (66.7%), ulna (66.7%), sacrum (58.9%), humerus (28.2%),

tibia (20.5%), and fibula (2.8%) in one study of 4 families. The ribs, skull and vertebrae are spared (2).

Some patients have associated connective tissue nevi called dermatofibrosis lenticularis disseminate, this is called Buschke-Ollendorff syndrome (11). Osteopoikilosis is also associated with keloid formation, dwarfism, spinal stenosis, dystocia, melorheostosis, tuberous sclerosis and scleroderma (6, 7, and 13). Mixed sclerosing bone dystrophy comprises osteopathia striata and melorheostosis along with osteopoikilosis (2).

Differential diagnoses include osteoblastic metastasis, tuberous sclerosis and mastocytosis (1, 9). Blood investigations like alkaline and acid phosphatase and bone scan are typically normal (1, 2, 14).

The diagnosis of primary synovial osteochondromatosis is based on the rapid onset of monoarticular pathology in a young adult and on the gross histopathological appearance (3). The typical features are ectopic chondroid matrix formation under the synovium, synovial hyperplasia, multiple small loose bodies and the absence of articular cartilage destruction. However secondary synovial metaplasia occurs in the more common traumatic and degenerative joint disorders (3, 5, and 6). Radiographs may or may not reveal calcific densities.

Synovial chondromatosis progresses through various stages of activity (4, 13). In the acute stage, the entire joint synovium is hypertrophied and hyperemic with numerous foci of cartilage formation. Small cartilaginous fragments are extruded into the joint and dystrophic calcification may occur. (3,6) During the intermediate stage, the acute synovial reaction gradually subsides (7,10,13, 15)The free cartilaginous loose bodies undergo degenerative, proliferative (enlargement on multiplication), or resorptive changes, depending on stresses peculiar to the joint involved and location within the joint (4,7). Endochondral bone formation may occur but requires a blood supply and is confined to loose bodies with a pedicle or to free loose bodies that have regained a synovial attachment (4,9).

In the late stages of the disease, the generalized synovial reaction reverts to normal. Secondary osteoarthritis results from the presence of loose bodies within the joint in chronic cases (6, 8, 9). This may cause localized secondary synovial chondromatosis, as well as osteochondritic loose bodies formed by detached osteophytes. Therefore, in older cases of long duration only a presumptive diagnosis of primary synovial chondromatosis can be made based on the history and the number and character of the loose bodies.

Synovial chondrosarcoma has to be considered in the differential diagnosis of synovial chondromatosis since both lesions may exhibit synovial calcification and atypical multinucleated cartilage cells (9, 13, and 16). The diagnosis of chondrosarcoma is established on the basis of a large, lobulated synovial lesion with extra-articular invasion and histological sections which exhibit numerous multinucleated cartilage cells having bizarre hyperchromatic nuclei. Secondary synovial chondromas, attached or free, which occur in association with traumatic internal derangement and osteoarthritis, are distinguished by the existence of primary joint disease and localized synovial reaction. Traumatic joint surface separations and detached chondro-osteophytes represent true osteochondritic loose bodies and are readily differentiated. Loose bodies resulting from traumatic

disruption of epiphyses, joint fibrocartilage or articular cartilage exhibit the gross and histological features of the normal joint structures involved. Chondro-osteophytes, usually no more than two or three in number, are often large and are found in a diseased or arthritic joint.

Microscopically they exhibit a periphery of fibrocartilage with underlying cancellous dead bone and a zone of calcification. The classic loose body (osteochondritis dissecans) is most often single and involves an otherwise normal joint. It is distinguished microscopically by a periphery of articular cartilage, fibrotic reparative reaction, and underlying necrotic bone.

A close differential of this condition would be melorrrheostosis with soft tissue involvement. This condition demonstrates an inner undulating margin encroaching into the medullary cavity with a linear track pattern. Typically there is a wavy cortical hyperostosis in the bone which simulates molten wax flowing down the side of a candle (17, 18). This may have an intra articular extension with or without joint effusion. Heterotopic bone formation and soft tissue calcification can rarely occur. Pathological examination shows variable degree of marrow fibrosis with irregular mixed areas of lamellar and woven bone. This was not seen in our patient.

Osteopoikilosis is differentiated from melorrrheostosis by symmetrical involvement, a normal scintigraphy, absence of soft tissue involvement, and no related symptomatology. Other differential diagnoses to consider would include myositis ossificans and calcified hematoma. Other sclerosing bone dysplasias encompass Jaffe-Lichtenstein disease which is a polyostotic fibrous dysplasia, which shows more frequent osteolytic areas and related histopathological changes.

The only other case report describing this association depicted multiple loose bodies in the hip joint and small spherical areas of increased density in keeping with generalized osteopoikilosis. (15). The authors speculated that chondromatosis is the synovial manifestation of osteopoikilosis (synosteoikilosis). To our knowledge the combination of the two entities has received little attention in literature, although several case reports have described osteopoikilosis and its associations, synovial osteochondromatosis is also an association to bear in mind when evaluating intra articular and juxta articular calcifications. As reported by Havitcioglu, (15) when osteopoikilosis is detected or suspected, lesions of fibroproliferative origin, should also be sought.

#### TEACHING POINT

When evaluating intraosseous and extraosseous calcifications, synovial osteochondromatosis with osteopoikilosis is an association to bear in mind. When osteopoikilosis is detected or suspected, lesions of fibroproliferative origin should also be sought.

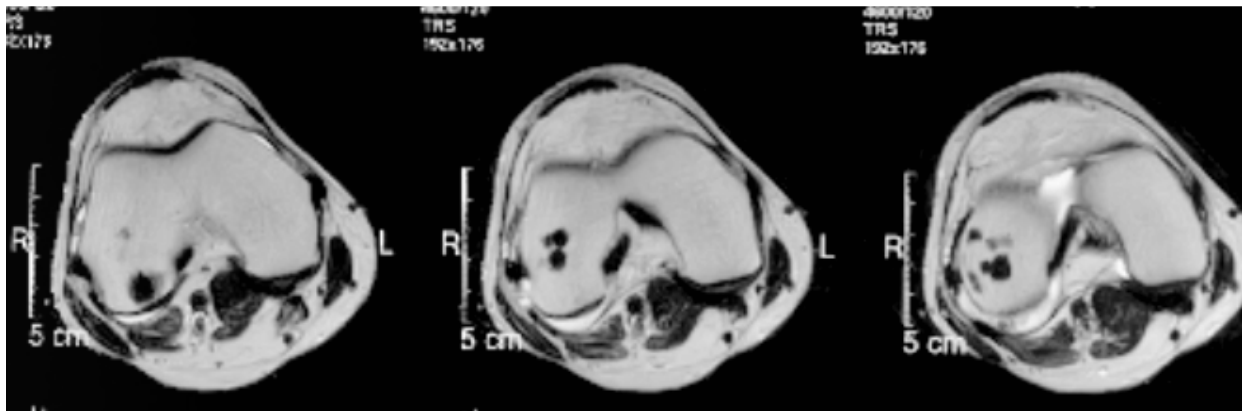
#### ABBREVIATIONS

CT - Computed Tomography  
MR - Magnetic Resonance Imaging  
AP - anteroposterior

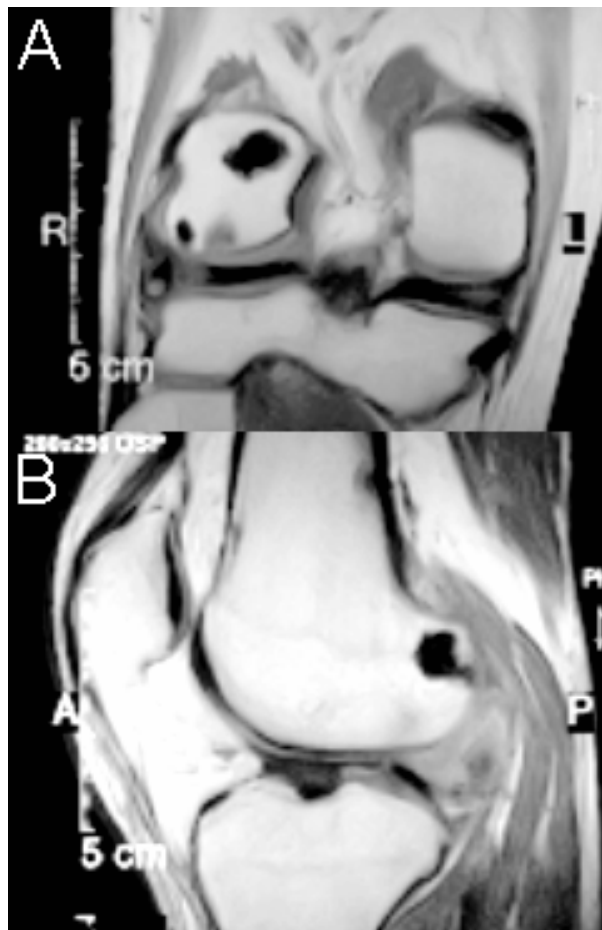
#### REFERENCES

1. Resnick D, Niwayama G. Enostosis, hyperostosis and periostitis. *Diagnosis of Bone and Joint Disorders*. W.B Saunders Company Philadelphia 1988, 4084-4088.
2. Lagier R, Mbakop A, Bigler A. Osteopoikilosis: a radiological and pathological study. *Skeletal Radiol* 1984.11(3):161-8.
3. Bloom, Ross, Pattinson, JN. Osteochondromatosis of the Hip Joint. *J. Bone and Joint Surg.*, Feb. 1951. 33-B: 80-84.
4. Fisher AGT. A Study of Loose Bodies Composed of Cartilage or of Cartilage and Bone Occurring in Joints. With Special Reference to Their Pathology and Etiology. *British J. Surg*, 1921, 8: 493-523.
5. Young LW, Gersman I, Simon PR. Radiological case of the month. Osteopoikilosis: familial documentation. *Am J Dis Child* 1980, 134(4):415-6.
6. Mussey RD, Henderson M S. Osteochondromatosis. *J. Bone and Joint Surg*, July 1949. 31-A: 619-627.
7. Murphy FP, Dahlin DC, Sullivan CR. Articular Synovial Chondromatosis. *J. Bone and Joint Surg*. Jan. 1962, 44-A: 77-86.
8. Nixon JE, Frank GR, Chambers G. Synovial Osteochondromatosis. With Report of Four Cases, One Showing Malignant Change. *U.S. Armed Forces Med. J.* 1960, 11: 1434-1445.
9. Bennett OA. Reactive and Neoplastic Changes in Synovial Tissues. *Proc. Inst. Med. Chicago*, 1950, 18: 26-37.
10. Benli IT, Akalyn S, Baysan E et al. Epidemiological and radiological aspects of osteopoikilosis. *J Bone Joint Surg*, 1992. 74(4):504-6.
11. Roberts NM, Langtry JA, Branfoot AC. Case report: Osteopoikilosis and the Buschke-Ollendorff syndrome. *Br J Radiol* 1993, 66(785):468-70.
12. Weisz GM. Lumbar spinal canal stenosis in osteopoikilosis. *Clin Orthop* 1982, 166:89-92.
13. Ackerman LV, Rosai J. *Surgical Pathology*. Ed. 5. St. Louis, The C. V. Mosby Co. 1974, 1086-1088.
14. Mungavan JA, Tung GA, Lambiase RE et al. Tc-99m MDP uptake in osteopoikilosis. *Clin Nucl Med* 1994. 19(1): 6-8.
15. Havitcioglu H, Gunal I, Gocen S. Synovial chondromatosis associated with osteopoikilosis: a case report. *Acta Orthop Scand*, 1998; 69:649- 650.
16. CL Holm. Primary synovial chondromatosis of the ankle. A case report, *J Bone Joint Surg Am* 1976; 58:878-880.
17. Khot R, Sikarwar JS, Gupta RP, Sharma GL. Osteopoikilosis: A Case Report. *Ind J Radiol Imag* 2005 15:4:453-454.
18. Patel AM, Vaghela DU, Kumar S, Shah UA, Shah AK, Shah HR. A rare case of melorrrheostosis with articular involvement: MR appearance. *Ind J Radiol Imag* 2006 16:4:453-454619-621.

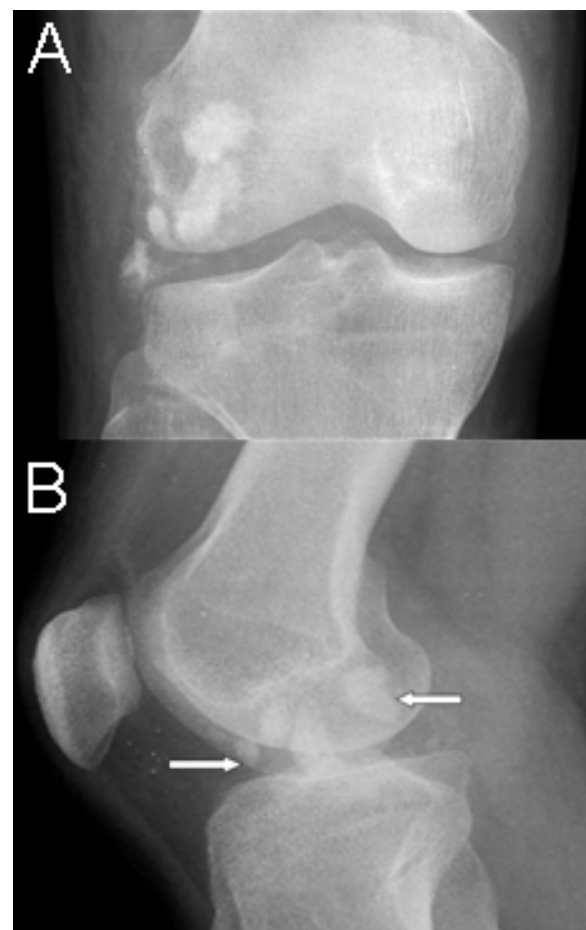
FIGURES



**Figure 1:** 25 year old man with osteopoikilosis and synovial osteochondromatosis of the knee. Axial proton density MR done elsewhere, images of the right knee, showing juxtaarticular rounded hypointense ossific foci, in the lateral femoral condyle, and posterior intrarticular soft tissues.



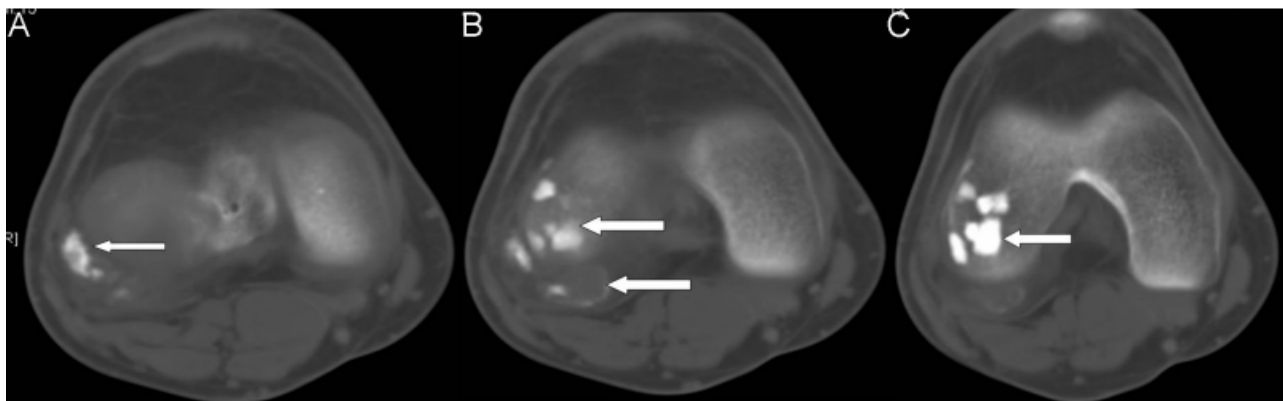
**Figure 2:** 25 year old man with osteopoikilosis and synovial osteochondromatosis of the knee. A: Coronal proton density MR done elsewhere, images of the right knee, showing juxtaarticular rounded hypointense ossific foci, in the lateral femoral condyle, and posterior intrarticular soft tissues. B: Sagittal proton density MR done elsewhere, images of the right knee, showing juxtaarticular rounded hypointense ossific foci, in the lateral femoral condyle, and posterior intrarticular soft tissues.



**Figure 3:** 25 year old man with osteopoikilosis and synovial osteochondromatosis of the knee. A: AP radiograph of the right knee showing intra osseus and extraosseus juxta articular ossific densities, in the lateral femoral condyle, and posterior intrarticular soft tissues. B: Lateral radiograph of the right knee, lateral view, showing intra osseous and extraosseous juxta articular ossific densities, in the lateral femoral condyle, and posterior intrarticular soft tissues.



**Figure 4 (left):** 25 year old man with osteopoikilosis and synovial osteochondromatosis of the knee. AP radiograph of the pelvis showing no significant abnormality.



**Figure 5:** 25 year old man with osteopoikilosis and synovial osteochondromatosis of the knee. Axial CT images without contrast of the right knee in bone window showing A: intra osseous juxtaarticular ossific densities, in the lateral femoral condyle, B: intraosseous and extraosseous juxtaarticular ossific densities, in the lateral femoral condyle, and posterior intrarticular soft tissues and C: extraosseous juxtaarticular ossific densities in the posterior intrarticular soft tissues.

KEYWORDS

Osteopoikilosis, synovial osteochondromatosis

**Online access**

This publication is online available at:  
[www.radiologycases.com/index.php/radiologycases/article/view/138](http://www.radiologycases.com/index.php/radiologycases/article/view/138)

Published by EduRad



[www.EduRad.org](http://www.EduRad.org)