

## CASE REPORT

# **Osteopoikilosis in a patient with frozen shoulder – a case review of a rare incidental radiographic finding.**

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## **Abstract**

Osteopoikilosis, known as 'spotted bone disease', is a rare and usually asymptomatic bone disorder but can be associated with malignant bone tumours. We present a patient with type I diabetes and Frozen Shoulder that was diagnosed with osteopoikilosis as an incidental finding when assessed radiographically. The radiographic features of this pathology are highlighted and the investigations described to exclude more sinister pathology. One of the usual diagnostic criteria of Osteopoikilosis is normal alkaline phosphatase blood levels however this is the first report of a patient whose alkaline phosphatase levels were persistently raised. This was attributed to diabetes. Raised alkaline phosphatase levels of an unknown cause have been previously reported to be a common finding in this patient group.

*Keywords:* Osteopoikilosis, frozen shoulder, diabetes, alkaline phosphatase

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## **BACKGROUND**

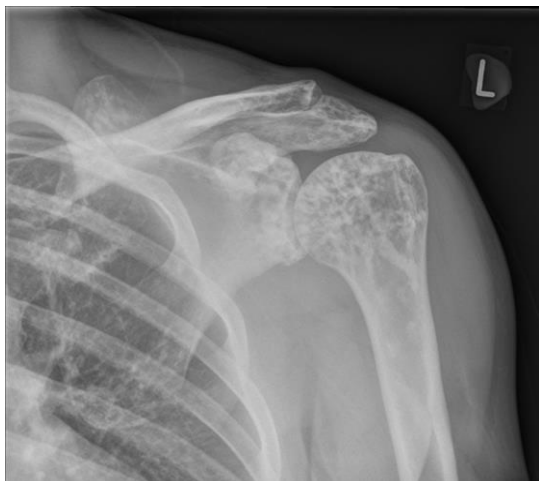
Osteopoikilosis, or osteopathia condensans disseminata, is a rare and usually asymptomatic bone disorder first described by Albers-Schönberg in 1915 [1-3]. The incidence is reported to be 1 in 50,000 [1-6]. It is an autosomal dominant inherited disorder linked to the LEMD-3 gene [1, 7]. It is characterised by the radiographic appearance of multiple small round or oval sclerotic spots at the epiphyses and metaphyses of long bones that give rise to its lay description of 'spotted bone disease' [2,5]. It is mostly reported to be asymptomatic but some authors have described 15-20% of patients having mild joint pains or effusion [6,8]. It is important to exclude malignancy in

these patients as osteosarcoma, chondrosarcoma and giant-cell tumour have been described in patients with osteopoikilosis [7, 9]. Blood tests can be used to exclude sinister pathology as blood count, liver and kidney function tests, inflammatory markers and measures of bone metabolism such as alkaline phosphatase should all be normal [2, 6, 10].

## **CASE STUDY**

A 41 year old male patient was referred to the orthopaedic clinic by their general practitioner with a six month history of insidious onset left shoulder pain and stiffness. They were assessed by an extended scope physiotherapist specialising in upper limb pathology. The

patient was a type 1 diabetic but was otherwise medically well. Clinical examination revealed a reduced passive range of shoulder movement in all planes with a 50% loss of external rotation. Rotator cuff tests were normal. The patient's subjective history of pain in the region of the deltoid insertion, disturbed sleep and jerk pain alongside the objective clinical findings suggested a diagnosis of frozen shoulder, but shoulder radiographs were requested as per usual practice to exclude arthropathy or sinister pathology. Patients with frozen shoulder should have normal radiographic appearances; however the images obtained were far from the normal (see Figures 1a and 1b) with the trabecular pattern of the humerus and scapula appearing significantly mottled.



*Figure 1a & b - AP and Axial radiographs of the left symptomatic shoulder.*

Advice was sought from a consultant radiologist and orthopaedic consultants before further tests were arranged. Initially full blood count, blood chemistry, ESR and CRP were requested along with a radiographic skeletal survey. The blood tests identified mild lymphocytopenia (1.29 /L) and raised alkaline phosphatase (205 iu/L) but were otherwise normal including ESR and CRP. The radiographic survey identified that the unusual trabecular pattern seen at the left shoulder was widespread throughout the skeleton (see figures 2-5)



*Figure 2a & b - AP and Axial radiographs of the right asymptomatic shoulder.*



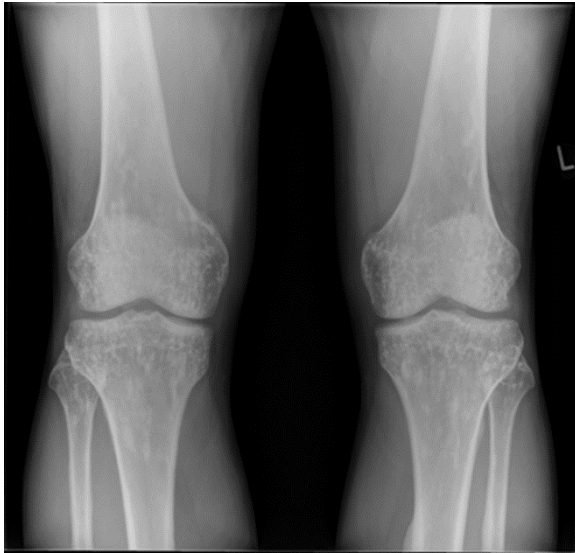


Figure 3: AP radiographs of both knees.



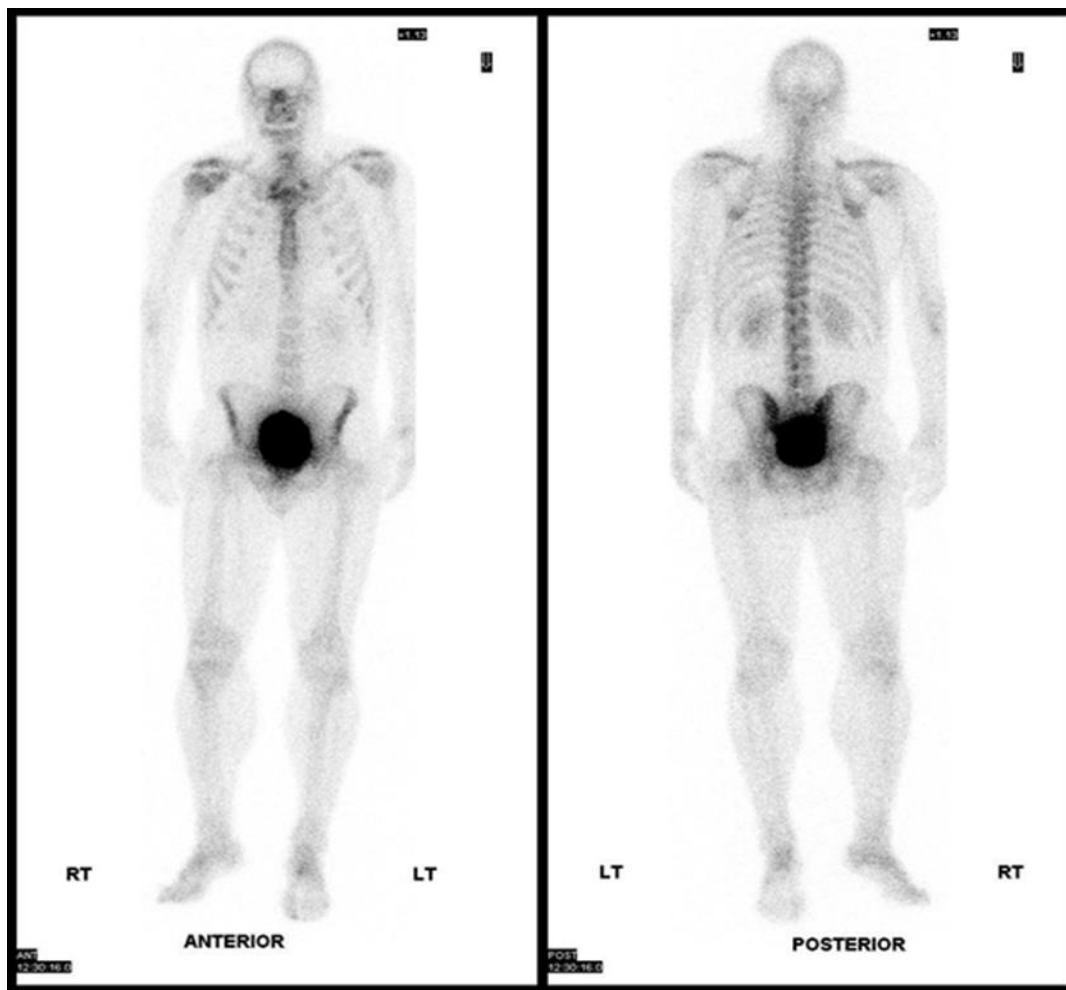
Figure 4: AP Radiograph of the pelvis.

Subsequently a radio-isotope bone scan was arranged (see Figure 5) which did not show any areas of increased uptake. Repeat blood tests later showed that the white cell count had normalised but alkaline phosphatase remained high. Gamma-glutamyltransferase (GGT) levels were normal. Therefore based upon the radiographic appearance and normal scintigraphy, a diagnosis of osteopoikilosis was made and the patient underwent usual treatment for the presenting frozen shoulder. The shoulder symptoms resolved following a combination of a single glenohumeral joint corticosteroid and local anaesthetic injection plus gentle home stretching exercises. The finding of osteopoikilosis was deemed incidental.

## DISCUSSION

Osteopoikilosis has been described in association with numerous other disorders including Buschke-Ollendorff syndrome, synovial osteochondromatosis, rheumatoid arthritis, familial Mediterranean fever, rheumatoid arthritis, ankylosing spondylitis and melorheostosis [1, 2, 7]. The limited literature available suggests that patients with

osteopoikilosis usually have normal blood count and blood chemistry [2, 6, 10] however our patient had persistently raised alkaline phosphatase. Similar findings have only been described in one previous paper written in Spanish describing the features of a parent with osteopoikilosis and her two young children with osteopoikilosis and Buschke-Ollendorff syndrome [11]. One child initially had raised alkaline phosphatase levels that subsequently normalised. Patients with raised alkaline phosphatase levels should be screened for liver pathology by assessing GGT levels [12]. These were found to be normal in our patient. It was thought that the raised alkaline phosphatase level must be related to diabetes as raised alkaline phosphatase of unknown cause has previously been reported to be common in this patient group, related to poorly controlled blood sugar levels [13]. Scintigraphy findings of zero or very low level uptake of the radio-isotope, as found in our patient, are considered to be the crucial factor in differentiating between osteopoikilosis and bone tumours [3, 10, 14]. A case of osteopoikilosis has been previously reported in a patient with diabetes and frozen shoulder



*Figure 5: Radio-isotope bone scan (scintigraphy) findings.*

[15] however the paper only described the imaging findings with no reference to the blood chemistry.

## **CONCLUSION**

Osteopoikilosis is a rare pathology that may be identified in the clinic during radiographic assessment. It is well recognised by radiologists but may lead to concern regarding neoplastic disease for clinicians with less expert knowledge of radiology. If the initial radiograph is not conclusive, a limited skeletal survey will usually confirm the diagnosis and eliminate the need for other tests. If bone pain is a symptom scintigraphy or MR

imaging is used to exclude dual pathology. Blood biochemistry is usually normal but, as discussed, in patients with diabetes alkaline phosphatase levels may be raised.

## **DECLARATION**

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

The Authors declare that there is no conflict of interest.

Ethical approval was not required and all patient identifiable information has been removed.

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