

## Ruolo dell'Imaging per la Definizione Diagnostica di una Coxopatia

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Paziente di anni 75, giunto alla ns osservazione nel febbraio scorso

## Anamnesi patologica prossima:

Graduale insorgenza, circa 6 mesi prima, di algie a carattere infiammatorio e tumefazione polso dx e gomito dx, persistenti, con rigidità mattutina della durata di circa 1 h a livello delle mani

## Anamnesi patologica remota:

- da circa tre anni sintomatologia dolorosa arti inferiori, a carattere meccanico e di grado moderato, non ben definita topograficamente, tuttavia prevalente a liv. delle art. coxofemorali
- ipertensione arteriosa
- ipertrofia prostatica

## Esame obiettivo reumatologico:

Quadro di artrite polso e gomito dx, dolente nella mobilizzazione anche la radiocarpica sn; segni di radicolopatia L5 (>dx); Faber-test +, limitazione escursione articolare anca (> sn)

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Reumatologi e Radiologi a confronto:  
l'importanza del Decision Making  
dalla diagnosi al follow up

## Documentazione iconografica remota Rx/TC (2018)



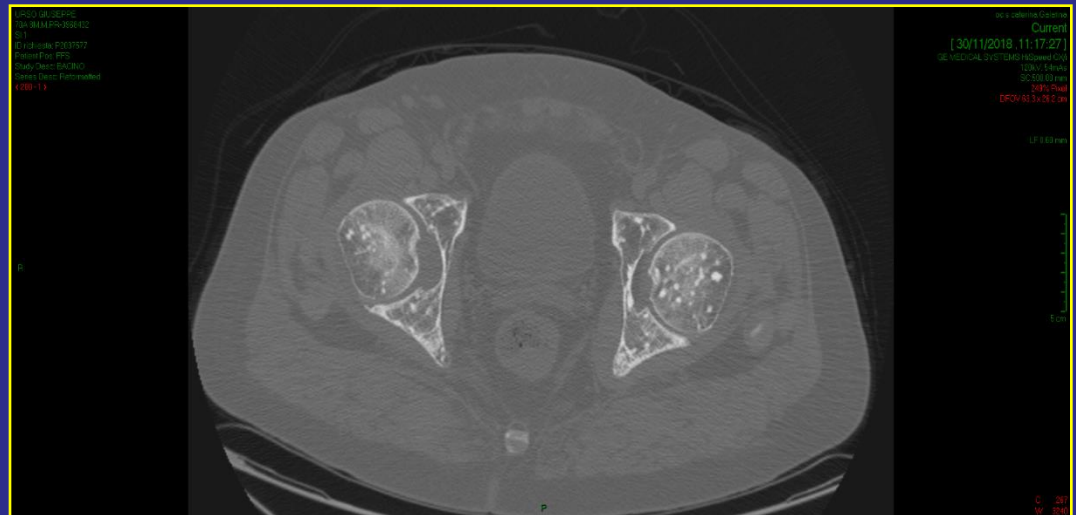
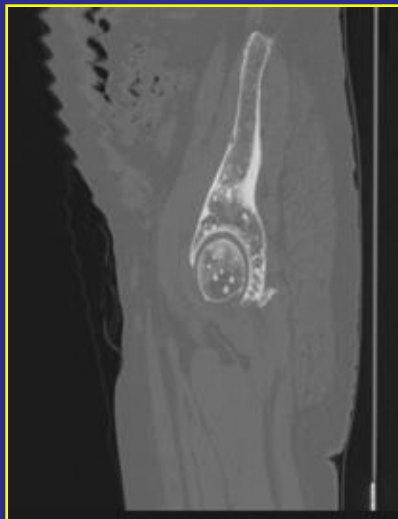
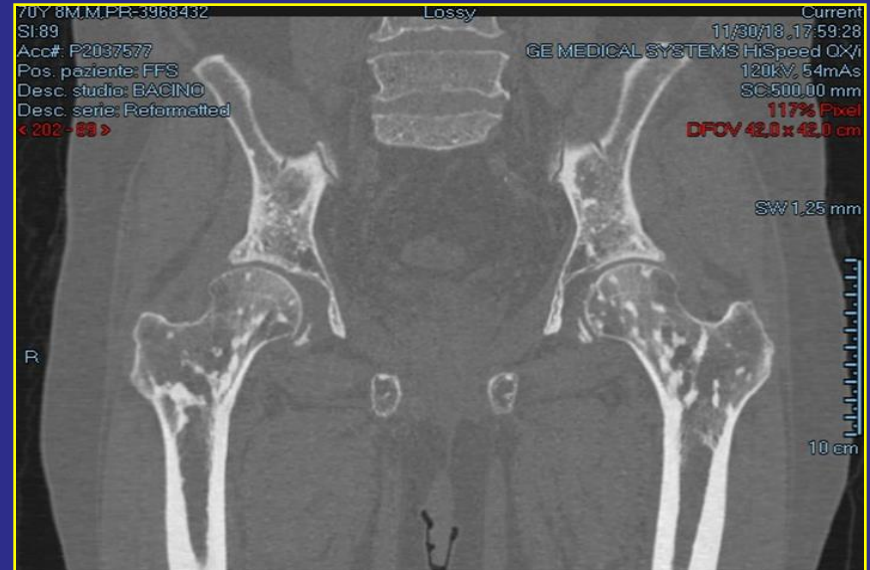
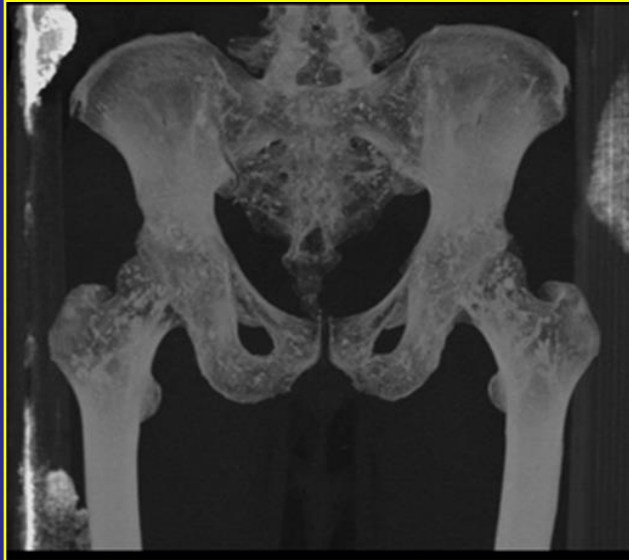
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...oltre ad elementi che hanno lasciato ipotizzare la presenza di un conflitto femoro-acetabolare di tipo misto, con calcinosi labrale a destra ed os acetabuli a sinistra

il reperto caratterizzante è rappresentato da  
**lesioni ossee di tipo sclerotico\***

## Workup of sclerotic lesions of bone

Sclerotic lesions of bone are areas of increased bone density seen on plain radiographs. When viewing an image showing spotted bones, it is necessary to consider the following three elements:

the appearance of the spots,  
their number,  
their location and distribution

Benign spotted bones: a diagnostic dilemma  
Gina Di Primio MD  
CMAJ, March 8, 2011, 183(4)

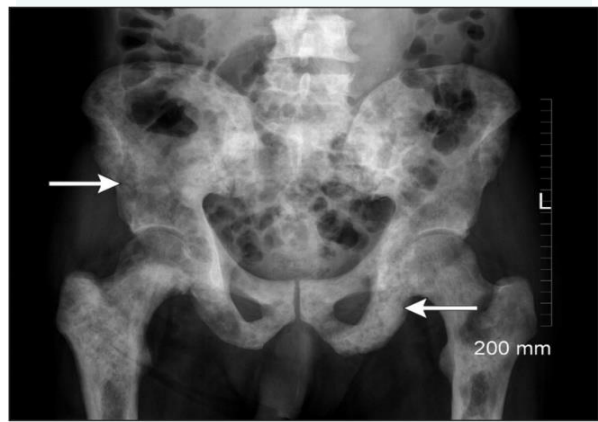
\* rotondeggianti/ovalari, ben circoscritte, di dimensioni dell'ordine di alcuni mm-fino a circa 1 cm, multiple, distribuite simmetricamente in sede periarticolare coxo-femorale (teste femorali ed acetaboli, estensione a colli femorali e reg. trocanteriche), a liv. sinfisi pubica, branche ischio-pubiche, art. sacro-iliache;  
aspetti simili erano evidenti in più sedi nella radiografia del piede e caviglia dx

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Quali le principali ipotesi diagnostiche suggerite dall'imaging?

Metastasi osteoaddensanti



Osteopoichilosi



Mastocitosi sistemica



Sclerosi tuberosa



Sono tutte condizioni accomunate dalla presenza di lesioni sclerotiche...

con alcuni aspetti che aiutano a definirne ulteriormente la  
caratterizzazione...

ma ciò che è virtualmente diagnostico è il pattern di distribuzione

La distribuzione simmetrica periarticolare e la predominante localizzazione meta-epifisaria sono caratteristiche dell'**osteopoichilosi**



Nel prosieguo dell'iter diagnostico, ai fini della principale diagnosi differenziale (metastasi osteoblastiche), erano state eseguiti:

- - markers neoplastici e del metabolismo fosfocalcico,
  - risultati nella norma
  
  - $^{99m}\text{Tc}$ -Scintigrafia ossea t.b.
  - FDG-PET/CT
- } non rilievo di aumentato uptake

## Osteopoikilosis - Pathology and clinical findings

Osteopoikilosis is a rare hereditary autosomal dominant bone dysplasia present in families, but can also present sporadically; there is an abnormality in endochondral bone maturation process and collagen regulation, which leads to the formation of trabeculae along the lines of stress. Heterozygous LEMD3 gene mutations have been shown to be the genetic cause of the disease

Histologically defined as bone islands that form dense core structure and are not associated with bone marrow in trabecular or cancellous bone

There are no specific clinical features; usually, the patients are asymptomatic, only  $\frac{1}{4}$  of them having mild joint pain and joint effusion; diagnosis is made by incidental typical radiological findings

There have been reported some disorders associated with OPK, like rheumatoid arthritis, synovial chondromatosis, ankylosing spondylitis.

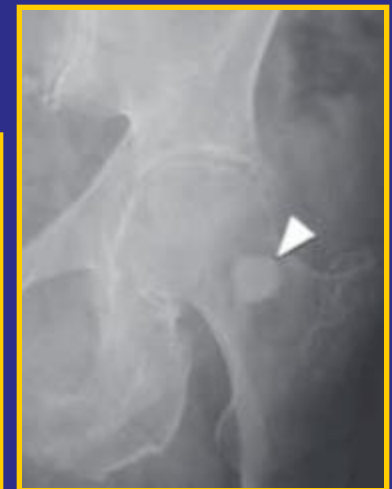
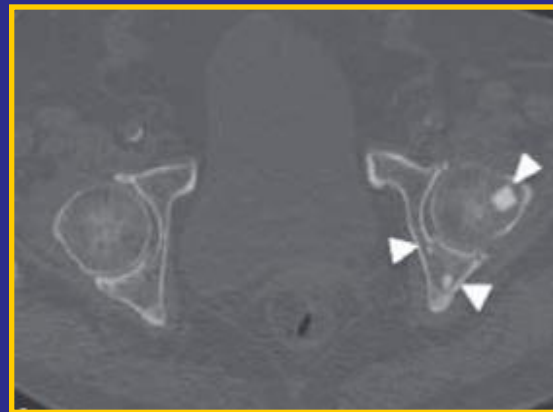
Frequently, the disease is associated with dermatological disorders like: disseminated lenticular dermatofibrosis (connective tissue disease associated with pigmented nevi, that, in combination with OPK, leads to Buschke-Ollendorff syndrome), keloid or discoid lupus and similar lesions to scleroderma

## Osteopoikilosis - Imaging

The characteristic radiologic feature is multiple sclerotic lesions of homogeneous structure, that are typically circular or ovoid, well marginated with spiculated margins in continuity with the nearby trabecular bone

The elementary lesion sharing the same appearance of a classical bone island

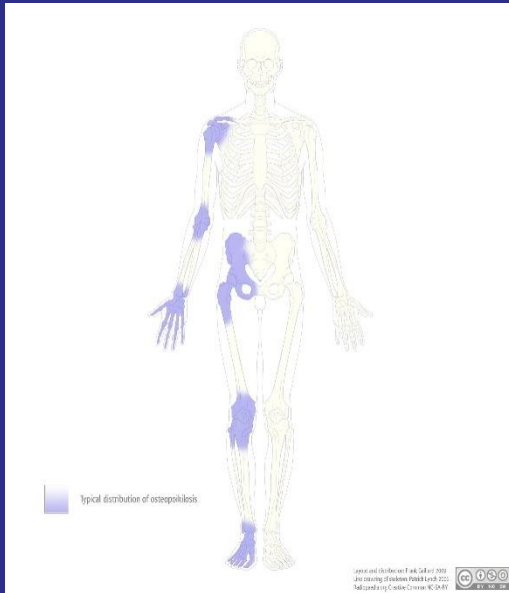
No transitional zone is observed around the osteoblastic spots



Each lesion has a size of a few millimeters usually 5-10 mm, but ranging from only 1-2 mm up to 1-2 cm

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They have a characteristically symmetrical periarticular distribution within the epiphyseal and metaphyseal regions of the appendicular skeleton (sites of endochondral bone formation), that is virtually diagnostic of the condition;

they are also common at the ends of short tubular bones (metacarpal/metatarsal bones and phalange),

in the spongy tissues of the tarsal and carpal bones;

the axial skeleton is largely spared;

osteopoikilosis has not been described in the skull

All bones were free of any cortical erosion or periosteal reaction

The bone islands of OPK align themselves parallel to surrounding trabeculae (thus predominantly longitudinal in the areas of well-defined linear trabeculae, while more-or-less spherical where the trabeculations are not as well organized linearly)

An example of former is any of the five major groups of trabeculae seen in femur head and neck



## Osteopoikilosis

[Dr Mostafa El-Feky](#) and [Assoc Prof Frank Gaillard](#) et al.

Radiopaedia

## Osteopoikilosis - Imaging

### Bone Scintigraphy Findings

- usually normal
- abnormal bone scan does not exclude osteopoikilosis and can reveal slightly increased activity similar to the bone island or enostosis that reflects an active osseous remodeling, especially in young patients
- an abnormal scintigraphy image found in older patients should be highly investigated because, sometimes, osteopoikilosis can mask a bone metastasis
- if the cellular activity in the lesions of osteopoikilosis continues, it should not be neglected that malignant transformation may develop in these areas; in literature, osteosarcoma and condrosarcoma have been reported

## Osteopoikilosis -Differential Diagnosis

### OPK

Main diagnostic features of

uniform size, symmetrical periarticular distribution

predilection for appendicular skeleton

never induce cortical erosion

no transitional zone around the osteoblastic spots (CT)

normal bone scan or slightly increased activity



### Osteoblastic Metastasis

varied in size, asymmetry

tends not to follow a periarticular distribution

predilection for axial skeleton

bony destruction, may cause subcortical destruction

periosteal reaction, transitional surrounding zone

positive bone scan

increased alkaline and acid phosphatase

In patients with a known or suspected primary malignancy, radionuclide bone scan has a critical role in distinguishing OPK from osteoblastic bone metastases.

## Osteopoikilosis -Differential Diagnosis

### Main diagnostic features of

#### Systemic Mastocytosis

Multiple osteoblastic lesions are found in late phase of the bone involvement, located at the metaphyses and diaphyses of the appendicular skeleton and at the axial skeleton

compared to  
osteopoikilosis

- asymmetric
- less well defined and uniform
- lesser periarticular preference

- an irregular thickening of the cortical is usually associated

- bone scintigrams may show more widespread involvement in patients with diffuse disease and detect a greater number of focal lesions

#### Tuberous Sclerosis (Bourneville's disease)

Sclerotic deposits are found focally or diffusely and can occur anywhere in bones including the axial skeleton and the extremities; calvarium is typically involved

- a periosteal reaction and different degrees of hyperostosis are associated with bone lesions

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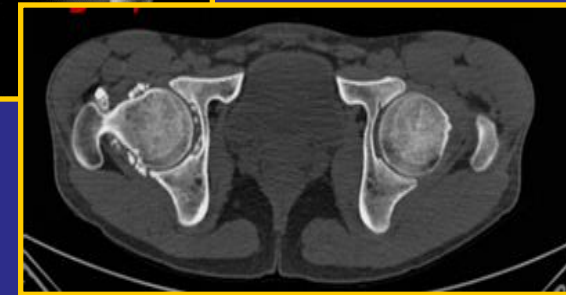
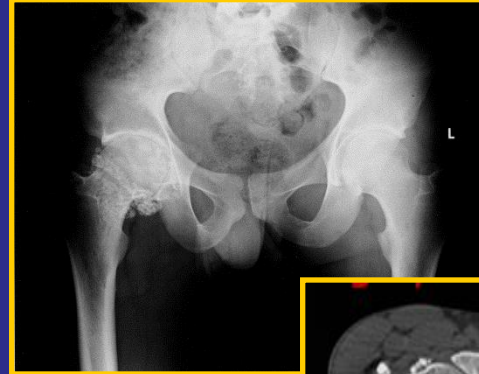
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## Osteopoikilosis -Differential Diagnosis

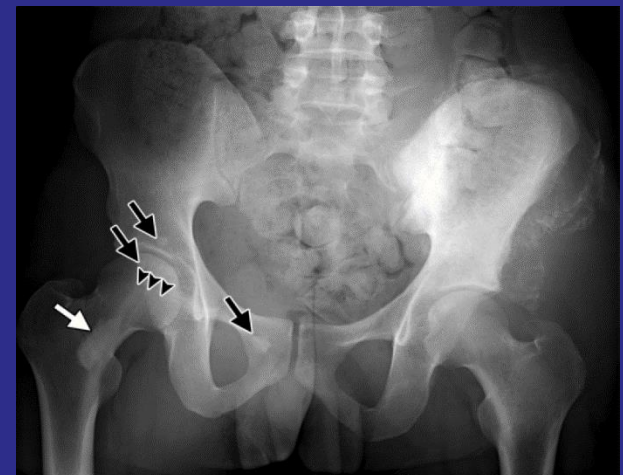
### Synovial Osteochondromatosis

Different localization compared to OPK:

- monoarticular involvement
- numerous calcified foci be found free within the joint cavity or they may be embedded within the proliferating synovium, which may extend into the surrounding tissues



**Osteopoikilosis** (black arrows) is often found concurrently with **osteopathia striata** (sclerotic areas linearly fine striated in the diaphyses and metaphyses of long and tubular bones, that run parallel to the long axis of the bone - arrowheads) and **melorheostosis** (regions of sclerosing bone with a characteristic dripping wax appearance - white arrow) and it is thought by some that they represent a spectrum of the same condition termed **mixed sclerosing bone dysplasia**





## Tornando al ns paziente....

relativamente alla sintomatologia dolorosa a carattere infiammatorio degli ultimi 6 mesi,

il quadro

- clinico

- laboratoristico (incremento indici di flogosi )

- ecografico (reperti da sinovite essudativa a liv. del polso e gomito dx, distensione capsula, come da versamento, art. radiocarpica sn e di alcune IFP delle mani)

ha orientato per la diagnosi di

**poliartrite cronica ad esordio senile in pz con osteoporosi**

A tutt'oggi non si dispone delle radiografie delle articolazioni interessate, richieste ma non effettuate per la sospensione delle prestazioni in relazione all'emergenza coronavirus

Sarà interessante verificare se le lesioni ossee sclerotiche rilevate a liv. degli arti inferiori siano anche presenti a liv. periarticolare degli arti superiori

## Take Home Message

- Tra le condizioni causa di lesioni ossee sclerotiche, è da tenere in considerazione l'OPK, sebbene sia di raro riscontro, specialmente in soggetti giovani
- La diagnosi è agevolata quando i caratteristici reperti di imaging siano riscontrati occasionalmente (ad es., per indagini effettuate a seguito di un trauma) in soggetti altrimenti sani, mentre maggiore difficoltà può occorrere in presenza di dolore, come nel ns caso clinico (anche se presenti altri fattori, di natura meccanica, che potevano determinarlo) o di sintomi di malattia sistemica
- In tale situazione, una rivisitazione di precedente imaging è importante per definire la stabilità delle lesioni nel tempo; laddove essa non sia disponibile, è necessario procedere ad indagini di II livello, tra cui, in particolare, la scintigrafia ossea
- Oltre all'importanza della diagnosi differenziale, è necessario prestare attenzione anche alla possibile associazione di tale condizione con altre patologie, ai fini di un corretto approccio diagnostico/terapeutico