Primary Synovial Chondromatosis of the Wrist

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INTRODUCTION

- Primary synovial chondromatosis = an unfrequent pathology
- The wrist (intra-carpal site) = an extremely uncommon affected site
- Only case seen in our Department in Department of Radiology
CASE REPORT

- CLINICAL CHARACTERISTIC: joints symptoms
  - 44 y-old, Taxi-man driver
  - > 1 y swelling and left wrist pain
  - Progressive \ ROM (Flex. 75°, Ext. 35°, Pro-sup. 70°)
  - Locking sensation
  - Palpable mass (ulnar side first)

- Laboratory tests:
  - no inflammatory syndrome
  - rheumatoid test -
  - no hyperuricemia

- Joint puncture under US:
  - culture -
  - no crystals

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RADIOLOGIC FINDINGS: pathognomonic

- Standard X-Ray

- Unilateral affection
- Multiple intraarticular calcifications
  (1/3 chondromas = no radio-opacity)
- Extrinsic erosion of bone
  (< mechanical P° in a very less capacious joint)
- Joint space typically maintained

RADIOLOGIC FINDINGS: pathognomonic

- Standard X-Ray

Oblique incidence

- Target appearance
  (central focus + single peripheral rim of calcification)

- Similar size and shape

- Typical "ring and arc" chondroïd mineralisation
  (longer standing disease)
**RADIOLOGIC FINDINGS**: pathognomonic

- **CT**: detect and characterize calcification

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- **Extrinsic bone erosion**
- **IA soft tissue mass**
- **No marrow invasion**
- **Calcified IA fragments (innumerable, typical pattern)**
RADIOLOGIC FINDINGS: pathognomonic

CT: detect and characterize calcification

3D reconstruction

Anterior view

Posterior view
RADIOLOGIC FINDINGS: pathognomonic

- MRI: marrow invasion?

**T1 WI**: signal intensity = muscle

Lobulated IA mass
RADIOLOGIC FINDINGS: pathognomonic

MRI: marrow invasion?

T2 WI: signal intensity (high water content) with central areas of signal int.
RADIOLOGIC FINDINGS: pathognomonic

MRI: marrow invasion?

T1 gado: enhancement of the vascularised synovium

Avascular cartilaginous nodules
SURGICAL TREATMENT

- Intra-operative views: Synovectomy with Removal of chondral bodies

Posterior approach
SURGICAL TREATMENT

- Intra-operative views: Synovectomy with Removal of chondral bodies

Posterior approach
Surgical Treatment

- Intra-operative views: Synovectomy with Removal of chondral bodies

Anterior approach
PATHOLOGIC FEATURES

- Gross pathologic appearance / macroscopic aspect

Nodular projections of hyaline cartilage

Hyperplastic synovium

Posterior synovectomy

Anterior synovectomy


PATHOLOGIC FEATURES

- Gross pathologic appearance / macroscopic aspect

Loose IA chondral bodies
Usually similar in size and shape
Multilobulated typical lesion
PATHOLOGIC FEATURES

Microscopic analysis: photograph of the whole-mounted specimen

- Some degree of calcification without bone formation (ossification)
- Nodules localised into the synovial membrane
- Typical lobulated growth
- Area of normal synovial
PATHOLOGIC FEATURES

Microscopic analysis: high-power photomicrograph

- Lobules of hyaline cartilage
- Abundant matrix without mixoid changes
- Variation in size and shape of nuclei
- Numerous chondrocytes

suggest a grade 1 or 2 Chondrosarcoma
Hypercellularity and Nuclear Atypia are TYPICAL of this benign disease
Histological analysis CONFIRMED DIAGNOSIS of PSC
DISCUSSION

DEFINITION

- uncommon benign process
- etiology: *in the past* = originally considered a chondroïd metaplasia in the synovium $\Rightarrow$ IA chondral bodies
  - actually = cytogenetic evaluation $\Rightarrow$ molecular abnormalities
  (expression of FGF 2-3, proto-oncogene C-ERBB2, chrom 6 abnormalities, ....)


DISCUSSION

DEFINITION


⇒ BENIGN NEOPLASTIC process more than METAPLASTIC process
DISCUSSION

● DEFINITION

● affects predominantly ♂ (2-4x more than ♀)

● 3rd to 5th decades of life

● commonly occurs within joints, rare cases in EA sites (tendon sheath and bursa)

● invariably a mono-articular disease (in large series)

● knee = the most commonly affected site
  Follow by : hip, elbow, shoulder, ankle
  Less commonly : MP, IP, distal RU, AC, TM, proximal T-F joints

Intra-carpal site NOT DESCRIBED in the literature.
DISCUSSION

Definition

- Symptoms: joint symptoms usually (pain, swelling, locking, instability, ...)
  - insidious and gradually progressive
  - duration before diagnosis +/- 5y
- Peripheral enchondral ossification in long-standing disease
  BUT histologically absent in 45% of cases (Davis’ study 1998)

- Synovial Osteochondromatosis is NOT A GOOD TERM
DISCUSSION

Differential Diagnosis: similar complaints

- Secondary synovial chondromatosis:
  - Very common abnormality
  - Older patients
  - Joint abnormalities (mechanical or arthritic conditions)
  - Most commonly affect: knee, hip, shoulder

→ Radiologically:
  - Underlying articular disease
    - Smaller number and variable size of chondral fragments

→ Pathologically:
  - Concentric ring of growth (several ring of calcification)
DISCUSSION

Differential Diagnosis: similar complaints

- Chondrosarcoma:
  - arise in bone and extend into the joint
  - Radiologically: can be distinguished
  - Pathologically: similar histologic atypia

- Soft-tissue chondroma:
  - location: hand and feet
  - no IA
DISCUSSION

TREATMENT

Optimal R/ = Synovectomy and removal of the chondral bodies

... BUT CONTROVERSY EXISTS....

SHPITZER 1990 and MAURICE 1988 studies: no difference in prognosis

OGILVIE - HARRIS study 1994: 60% recurrence rate removal chondral bodies alone
DISCUSSION

- **EVOLUTION-PROGNOSIS**
  - recurrence rate 3-23% (related to incomplete resection in many cases)
  - chronic disease $\Rightarrow$ secondary OA
  - malignant transformation to chondrosarcoma:
    - extremely rare event (5% of cases)
    - signs of malignancy: - multiple local recurrences
      - rapid $\Rightarrow$ of the lesion
      - rapid clinical deterioration
      - marrow invasion

$\Rightarrow$ R/ Amputation
CONCLUSION

- Benign self-limited process, may recur locally
- Histologic appearance suggest a chondrosarcoma

Histologic CORRELATION with the Radiologic appearance = ESSENTIAL for a correct diagnosis and treatment.
Thank you