CASE REPORT

Monostotic Paget's disease of the second metacarpal

Selami Cakmak, Levent Umur, Kenan Keklikci, Osman Rodop

Gata Haydarpasa Hospital,
Department of Orthopaedics
and Traumatology, Istanbul,
Turkey

Correspondence to
Dr Selami Cakmak,
selamicakmak@gmail.com

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SUMMARY

Paget's disease of the bone is a focal chronic disorder proceeding with elevated osteoblastic and osteoclastic activity in the affected area. The most common sites are pelvis, femur, lower lumbar vertebrae and skull. Monostotic disease is reported in 15–30% of all cases. We report a case of monostotic Paget's disease of the second metacarpal, which is a rare location even for polyostotic disease.

BACKGROUND

Paget's disease of the bone is a common skeletal disorder characterised by osteoclastic resorption following bone formation and is usually seen as polyostotic (affecting the pelvis, femur, spine, tibia and skull). Polyostotic Paget's disease of the hand has been reported in the literature, however, monostotic Paget's disease of the hand (confirmed with histology) is rarely reported.^{1–10} Included bones of the hand are the second metatarsal,^{3 5} third metacarpal,^{3 5 7 9} fourth metacarpal,^{2 5} fifth metacarpal,^{5 10} proximal phalanx,^{1 4–6} middle phalanx,^{1 4 5} distal phalanx,⁵ os hamatum,⁵ scaphoid and capitate.⁵ Although Grundy and Patton did not clearly mention whether the patterns in their 11 cases were monostotic or polyostotic, we considered them to

be monostotic lesions.⁵ We report another case of hand localised Paget's disease incidentally discovered at the second metacarpal.

CASE PRESENTATION

A 63-year-old woman presented with mild persisting pain in the radial aspect of the right wrist for the past 3 months after a trauma. Physical examination and range of motion were normal. Initial X-rays targeting the wrist incidentally revealed radiological changes of the second metacarpal (figure 1). They showed structural deformity with a mosaic pattern in the second metacarpal (figure 2). MRI showed diffuse cortical growth and mass lesion indicating fibrous dysplasia (figure 3). A bone scan revealed no other focal activities other than in the second metacarpal, indicating monostotic disease (figure 4). Laboratory findings confirmed inactive disease (serum (ALP)-alkaline phosphatase levels 34 IU/L, tartrate resistant acid phosphatase 17.08 IU/L, hydroxyproline: 8.07 mg-hydroxyproline/g-creatinine, bone specific ALP: 62.23 IU/L).

Treatment requirements were considered, but as the patient was asymptomatic and the disease biochemically inactive, no treatment was considered necessary; however, the patient was called for follow-up examinations.

Figure 1 Initial radiographs of the patient focused on the wrist joint and diffuse increased density of the second metacarpal was found incidentally.

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Figure 2 Radiograph of the hand shows increased density, homogenous sclerosis with rare lucent areas localised at the second metacarpal.

DISCUSSION

Paget's disease of the bone is a focal chronic disorder proceeding with elevated osteoblastic and osteoclastic activity in the affected area. Men, especially over 55 years of age, are more commonly affected. Approximately 80% of patients are asymptomatic. Symptoms vary depending on the affected body part and activation levels of the lesions. Pain and deformity are the

first symptoms. Pain can be caused by periosteal irritation, nerve pressure and microfractures. Headache, hearing and visual loss can be seen in cases involving the skull. Diagnosis depends on X-rays (characteristic appearances), bone scans (radiolabelled bisphosphonate),¹¹ elevated levels of serum ALP, serum and urinary hydroxyproline, bone turnover markers such as urinary and serum deoxypyridinoline, N-telopeptide and C-telopeptide

Figure 3 MRI shows diffuse cortical growth of the whole second metacarpal (which can be easily confused with fibrous dysplasia).

Figure 4 No other focal activities other than increased uptake at the second metacarpal indicates monostotic Paget's disease.

and serum TRAP.^{12 13} Serum ALP, serum TRAP, hydroxyproline and bone specific ALP levels were at normal ranges in our patient, indicating an inactive phase of disease.

Most commonly affected sites are the pelvis, femur, lower lumbar vertebrae and skull. Monostotic disease is present in 15–30% of all cases. Monostotic Paget's disease of the hand is usually asymptomatic and patients can be diagnosed incidentally on radiographs taken for other reasons.⁵ However, it is reported that some patients with monostotic disease of the hand experienced pain and/or swelling.^{4 8} Our patient also had mild pain but it was localised at the radial side of the wrist, not specifically at the region of the second metacarpal.

The radiographic appearance of monostotic Paget's disease is the same as in the polyostotic form, and is characterised by three different radiographic patterns: osteolytic, sclerotic and combined osteolytic-sclerotic.¹⁰ Our patient had increased bone density, homogenous sclerosis and rare lucent areas, showing it to be a sclerotic pattern.

Since Paget's disease of the bone is a chronic disorder with remission and activation periods, asymptomatic and low risk patients in remission may not need treatment. Symptomatic patients or those with active disease (elevated ALP levels) or

disease at high-risk sites (skull, spine and weight-bearing bones), should undergo an appropriate treatment regimen prior to elective surgery of the pagetic site (arthroplasty). Monostotic Paget's disease of the hand usually does not require treatment, as in our case, but does call for follow-up.

Contributors All authors made valuable contributions to the diagnosis, investigation and follow-up of this patient and are still on follow-up duty. They also wrote the manuscript for this case presentation.

Competing interests None.

Patient consent Obtained.

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Learning points

- Paget's disease is a chronic disorder characterised by elevated osteoclastic activity followed by excessive osteoblastic activity and is usually seen as polyostotic disease.
- Monostotic Paget's disease of the hand is rarely seen, and is usually asymptomatic and diagnosed incidentally.
- Monostotic Paget's disease of the hand is usually inactive biochemically and rarely needs treatment.

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