Monostotic humeral Paget's disease

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DESCRIPTION

A woman in her late 50s with no significant comorbidities presented with constant dull aching pain in her right arm for over a few years without any shoulder pain or restrictions in arm movement. She had mild symptomatic improvement in pain with non-steroidal anti-inflammatory medications. Clinically, she was found to have tenderness along the right deltoid region. Plain radiograph of the right shoulder and humerus (figure 1) showed cortical thickening, and trabecular coarsening throughout the right humerus and proximal shaft, suspicious of metastatic/metabolic disease. Blood biochemistry showed elevated alkaline phosphatase 280 U/L (45–117 U/L) with bone-specific alkaline phosphatase being 76% (14%–68%). Rest of the laboratory work-up was unremarkable with complete blood count, liver function tests and renal function tests within normal range. A radionuclide whole-body scan with 21.8 mCi technetium 99m hydroxyapatite phosphate (figure 2) showed increased radiotracer activity in the right humerus consistent with Paget’s disease. Patient was treated with a single dose of 5 mg intravenous zoledronic acid. At the 6-month follow-up, she continued to have dull ache in her arm. However, bone-specific alkaline phosphatase levels and whole-body scan remained stable.

Paget’s disease of the bone results in abnormal bone remodelling (poorly woven mix of highly vascularised loose and compact bone tissue) due to the disproportionate activity between osteoclasts and osteoblasts.1 The geographical prevalence of Paget’s disease is variable with predominance in people of British ancestry and affecting 1%–2% of white adults older than 55 years.2 It can be monostotic (single bone) or polyostotic (multiple bones) predominantly involving the skull, spine, pelvis and lower long bones. Monostotic Paget’s disease can be asymptomatic with elevated total alkaline phosphatase (ALP) or upper normal total ALP levels and significant radiographic findings, depending on the disease activity.3 However, pain can be the most common presenting symptom with varying severity owing to bony lesion or its complications.4 5 6 Plain radiographs are often diagnostic and help identify complications associated with Paget’s disease, including fractures, osteoarthritis or other bone abnormalities.6 Nonetheless, a baseline radionuclide bone scan should be conducted to rule out the involvement of other sites.7 Biopsy is rarely required to establish the diagnosis unless the monostotic lesion lacks typical radiological findings of Paget’s disease.8

A case such as this one with dull chronic pain with no complications can delay the diagnosis, given the low prevalence and less frequent exposure to monostotic Paget’s disease. Early initiation of treatment has proved good functional outcomes.

Learning points

- Monostotic Paget’s disease is rare and needs a high index of suspicion in any patient presenting with bone pain at any region with/without elevated total alkaline phosphatase.
- Elevated bone–specific alkaline phosphatase and plain radiograph can help reveal the diagnosis. Bone scan is sensitive for an early disease but not specific for diagnosis.
- Bisphosphates are recommended for the treatment and prevention of disease progression.

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Images in...

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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