

CASE REPORT

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Paget's disease of bone: the first case reported in Iraqi Kurdish

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Abstract

Background Paget's disease of bone (PDB) is a chronic bone disorder which insidiously evolves parallel with the aging process. The affected bone has a distinct imaging appearance, and the clinical manifestations are variable. This disease is ubiquitous among people of Anglo-Saxonian origin. However, it is rarely reported among Asian descents.

Case presentation Here, the first diagnosed case of Paget's disease of bone in an Iraqi Kurdish patient, a gentleman in his seventh decade of life underwent sectional imaging for evaluation of spine hopefully to find a potential musculoskeletal or neurological culprit behind his back discomfort. Biochemical confirmation tests were demanded based on the state of the art of the typical radiological judgment, perhaps negating pathological tissue affirmation. Marked geographical variation of the disease occurrence made it seldom to encounter such entity in the Middle East, particularly in Iraq. To the author's knowledge, scarce cases have been described in Iraq, none in the Kurdish population.

Conclusion This case report elicits an extremely rare metabolic osseous disease that was radiologically diagnosed at venture when bone window computed tomography (CT) is performed for an unrelated indication.

Keywords Paget's disease of bone, Computed tomography, Metabolic disease, Low back pain

Background

Osseous Paget's disease is an enigmatic disease, notorious for gradually progressive bone remodelling that affects the axial and appendicular bony skeleton. It is primarily the disease of elderly with male predominance [1]. However, sporadic cases have been recorded under 40 years of age [2]. The exact underlying etiology remains unclear; factors postulated to contribute to the pathogenesis include genetic mutation, environment, and even vitamin D deficiency [3].

The basic pathology of PDB is dysregulation of osteoblast and osteoclast function, which leads to altered bone texture, and it is a mechanical property accounting

for the pain and deformity that patients experience [2], although in 70% of cases the disease is asymptomatic and discovered incidentally [4]. Expected complications of PDB are either local such as fracture, malignant changes, osteoarthritis, and neurovascular compressions or systemic-like cardiac insufficiency [5]. PDB can affect any bone in the body, and the skull, lumbar vertebra, pelvis, and femur are among the common predilection sites for the disease [6].

The diagnosis of PDB is primarily by characteristic radiological findings [7]; biopsy is seldom required to establish the diagnosis [8]. Serum alkaline phosphatase, a bone formation marker, is used for the diagnosis and monitoring of the disease. Serum calcium and phosphorus levels are normal in most patients; when it is abnormal, secondary disorders such as hyperparathyroidism need exclusion [7]. Bone scintigraphy helps determine the extent of involvement of the bone [9]. MRI and CT are used in cases of complications such as fractures, staging of sarcomatous degeneration, and evaluation of

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neurological complications like compression of spinal and cranial nerves [10].

Pathology and radiology wise, PDB has three phases; these include the following: initial phase of bone resorption, intermediate, and late phase of sclerosis and remodeling with increased thickness of the bone trabecula and cortex [5].

Various derivatives and generations of bisphosphonates are used for the treatment of PDB owing to their potent inhibitory effects on bone turnover [11] by which the normalization of alkaline phosphatase can be achieved.

Case presentation

Our patient was a 69-year-old Iraqi man from the South Kurdistan region who visited the pain clinic outpatient of our hospital in July 2022, complaining of nonspecific long-standing enduring back pain. After meticulous onsite general and regional assessment alongside revision of the detailed clinical background history, the patient was referred to the radiology department for bone window multidetector computed tomography (CT) for dorsolumbosacral spine evaluation, in addition to basic blood screening. Clinically, the patient described long-duration, nonremitting, and nonradiating moderate severity mid-lower backache, which eased with rest. Nevertheless, no definite aggravating factor has been alluded from a diagnostic standpoint. Otherwise, he was healthy with ordinary spine contour, no posture deformity or abnormal gait, and neither locomotion limitation nor joint dysfunction. Upon physical examination, there were no back skin changes on inspection or point tenderness on local palpation. Straight leg and other provocative tests were negative. Neurological dysfunction was excluded by virtue of unremarkable neurological assessment including each of motor, sensory, and reflex tests. No sign and symptom concerning for cardiac insufficiency, infection, inflammation, or other pertinent medical or surgical history were confirmed apart from being hypertensive controlled with beta-blocker agents.

Preliminary laboratory test records were unremarkable in terms of complete blood count, C-reactive protein, and erythrocyte sedimentation rate. Further bone profile assessment revealed high alkaline phosphatase at 77 U/L, while serum calcium, phosphate, and prostate-specific antigen were within the normal range (Table 1).

Imaging findings

Multiplanar reconstructed bone and soft tissue window CT images of the spine were conducted in sagittal (Fig. 1), coronal (Fig. 2), and axial (Fig. 3) planes with a thin section (1 mm), in addition to volumetric rendering (Fig. 4), using the Siemens SOMATON Definition AS 2012B. Reviewing the obtained images on a multitask

Table 1 Patient's laboratory findings

Lab parameter	Patient measure
Alkaline phosphatase	77 IU/l
Serum calcium	8.2 mg/dl
Serum phosphate	3.5 mg/dl
Prostate-specific antigen	0.8 ng/ml
OH vit. D	24 ng/dl
Serum cholesterol	205 mg/dl
Serum triglyceride	210 mg/dl
LDL	100 mg/dl
HDL	40 mg/dl
VLDL	30 mg/dl
ESR	6

scrollable workstation revealed the following: a remarkably abnormal left hemipelvis bony skeleton including ileum, through the ischium and pubic bone, and to a lesser extent the upper sacral segments. The cortexes are thick, dense, irregular, and shaggy. Prominent, coarsened, and thickened trabecula seen as well as intermixed with islands of bone rarefaction and hypodensities encompassing the entirely involved medullary cavity of the bones, on the top of noticeable osseous contour expansion and hypertrophy. Such cardinal features were quite impressive for the intermediate-to-late phases of PDB. The remaining bones under vision within the scan (dorsolumbar vertebra, right hemipelvis, proximal femora, and lower most visible ribs) were unremarkable as such presumed both hip joints symphysis pubis.

Degenerative spondylosis spinal changes seen within lumbar and lower dorsal vertebral levels presented with lipping osteophytes, narrow disc spaces, endplate disc herniation (Schmorl's nodule), and facet joint arthrosis, which potentially explain the patients' presenting backache.

The diagnosis of PDB is suggested from the explicit imaging signs, which subsequently supported by the finding of raised serum alkaline phosphatase.

As the patient refused to undergo bone biopsy, after negotiation, the provider clinician placed him on bisphosphonate therapy, as the standard regime for PDB.

Discussion

Once the PDB was first depicted in 1876 by Sir James Paget, a British surgeon, and described as osteitis deformans owing to the clear hypertrophy and augmentation of the diseased bone [12], long-term studies in the literature have constantly approved the highest prevalence of the disease in England, the USA, Australia, New Zealand, and Europe, mainly among individuals older than 55

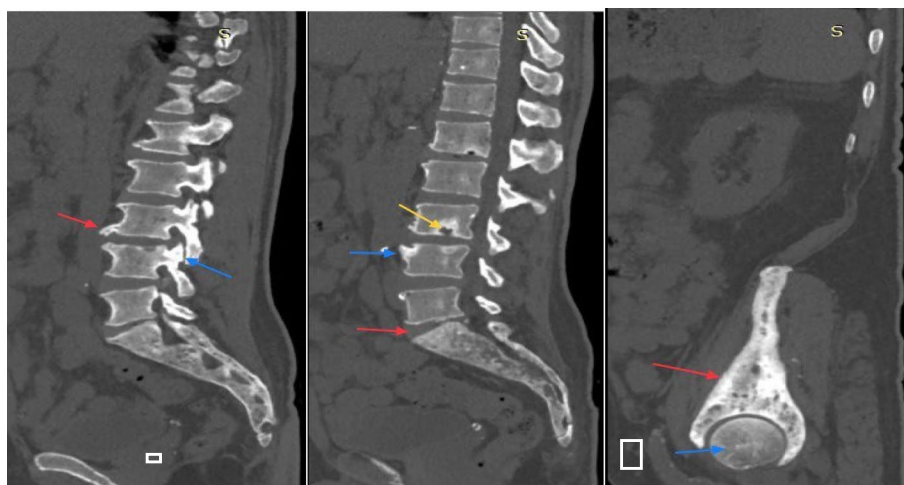


Fig. 1 Sagittal plane bone window MDCT shows spondylotic changes presented by vertebral osteophyte and facet joint sclerosis with narrowing (A red and blue arrows), Schmorl's nodes (B yellow arrow) and narrow L5-S1 disc space (B red arrow), and typical changes of PDB in left pelvic bone with normal left femoral head (C red & blue arrows)

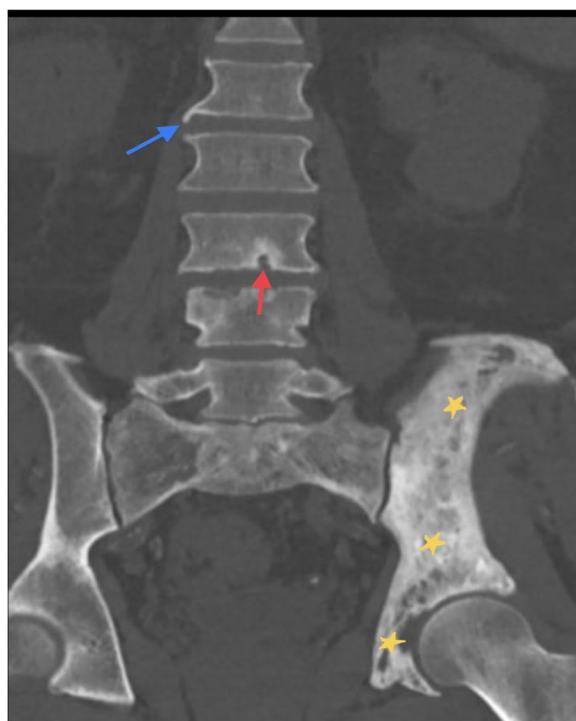


Fig. 2 Coronal plane bone window MDCT shows classical features of PDB in left-side pelvic bones (yellow stars) sparing the proximal femora and hip joints. Spinal degenerative changes in terms of osteophytes (blue arrow) and Schmorl's nodule (red arrow)

years [13]. The estimated prevalence in the USA is 1–2% [14], and that in Great Britain is 3–4% [15].

PDB is extremely rare in the eastern world especially in Korea [16], China, and Japan [17]. Few resources are available regarding the prevalence of PDB in the Middle

East and among the Arabic population. Alshaikh has reported four interesting cases of Saudi Arabian patients with PDB in 2011, which were preceded by one case report in 1998 and the first reported Saudi case of PDB in 1988 [18]. Few other individual cases have been reported in Iran and Turkey [19].

Paget's disease of mandible is first reported in Iraq by Uthman and Al-Shawaff (1969) from Baghdad [20]. Abd Al sahib (2007), an orthopedic surgeon, reported a PDB case presenting with pathological fracture in Karbala [21].

To the best of the knowledge of the author, no case of PDB has been reported from the Iraqi Kurdish population inhabiting South Kurdistan region, to the time being.

In our case, the diagnosis of PDB has sought out solely based on classical imaging findings in characteristic age and gender favorite of the disease entity. Fibrous dysplasia is a differential diagnosis for younger age patients. Metastatic bone deposit especially from prostatic primary in elderly male and breast in female patients is another consideration. Nevertheless, it is conclusively eliminated in this case report by the fact that prostate-specific antigen was normal, and imaging wise, the sclerosis has involved the cortex rather than medullary bone, the predicted site for metastasis.

Chronic osteomyelitis is another radiological mimicker for PDB, which is ruled out in this case by negative prior history and normal inflammatory markers.

Therefore, recognizing the potential radiological differential diagnosis of PDB is crucial [22].

The objective of this case report is to incite the related specialities for the consideration of PDB any time clinical, laboratory, and radiological suspicion encountered.

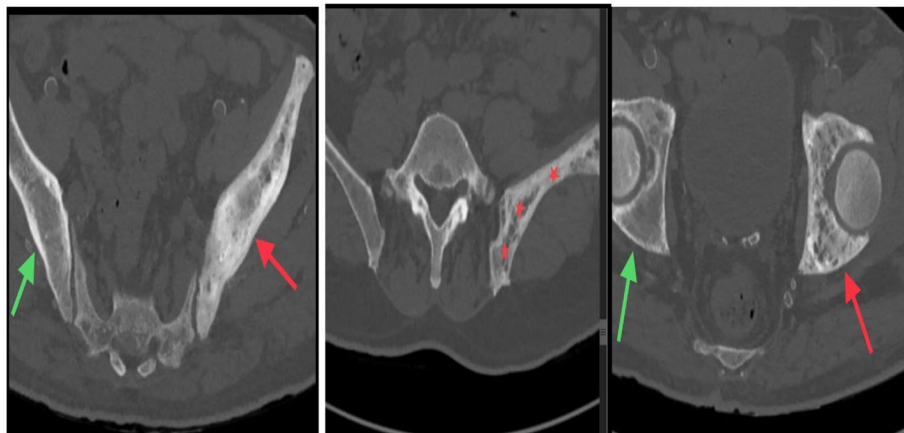


Fig. 3 Axial plane bone window MDCT redemonstrate the changes in the left ilium, ischium, and pubic bone (red arrows & stars) normal right-side bony pelvis (green arrow)

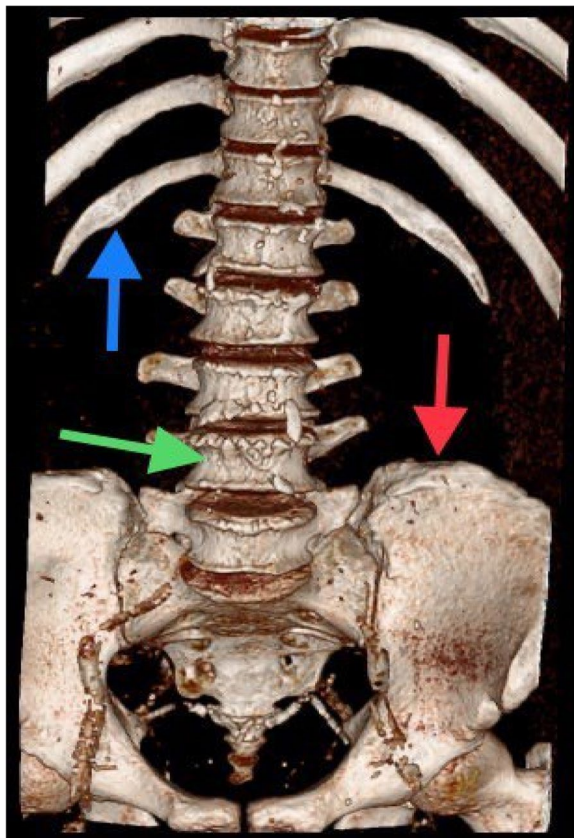


Fig. 4 Volumetric image of bone window MDCT illustrates the hypertrophied left pelvic bones (red arrow), degenerative changes in the spine (green arrow), and normal ribs (blue arrow) and right-side pelvic bones

the clinical practice. Although the low prevalence of the disease in this part of the world might be vastly attributed to genetic makeup and climatic factors, PDB is asymptomatic and hence tangible when overlooked, missed, and underreported.

Abbreviations

PDB Paget’s disease of bone
MDCT Multidetector-computed tomography

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Authors’ contributions

The corresponding author conceived the study, analyzed and interpreted the radiological findings, and established the manuscript design and organization. The co-author designed the illustrations and drafted the manuscript. The authors read and approved the final manuscript.

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Availability of data and materials

The data and materials supporting the findings of this study are available on request from the corresponding author.

Declarations

Ethics approval and consent to participate

This study was approved by the ethical committee of Hawler Medical University. Written informed consent has been obtained from the patient for the publication of this case report and accompanying images.

Consent for publication

The patient included in this case report gave written informed consent for the publication of the data and materials contained in this study.

Competing interests

The authors declare that they have no competing interests.

Conclusion

The rarity of the PDB in the Asian horizon rendered the entity worthy for an account whenever fulfilled in

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