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Rare case of paget's disease with co-existing features of ankylosing spondylitis

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Abstract

Introduction: Paget's disease is an uncommon metabolic skeletal disorder characterized by increased osteoclast-mediated bone resorption & compensatory excessive osteoblastic activation. It is often an incidental finding & can also rarely co-exist with other disorders like ankylosing spondylitis.

Materials & Methods: Observational type of case study using computed tomography (CT) and X-Ray. 80 years old male came with complaints of giddiness & fall & decreased speech output. CT scans showed intraventricular hemorrhage & patient was treated for the same. However, CT scans revealed features suggesting Paget's disease with co-existing ankylosing spondylitis & a bone survey for whole body was done. Increased S. Alkaline Phosphatase (187 U/L) & low Calcium (7.5 mg/dl) was noted.

Results: CT Brain showed mixed lytic sclerotic changes with widened diploic space & coarse trabeculae involving calvarium fully giving a cotton wool appearance. Spine imaging revealed, picture framing of some vertebrae, anterior & lateral syndesmophytic fusion giving a bamboo spine appearance & diffuse osteopenia with multiple sclerotic areas. X-Ray pelvis showed of Kohler tear drop & sclerosis with near complete fusion of bilateral sacroiliac joints.

Conclusion: Paget disease of bone shows an abundance of patterns on imaging that are related to pathologic stage of disease. Recognition of imaging spectrum usually allows early diagnosis & differentiation of its complications, which helps guide therapy & improve patient management.

Keywords: Paget disease, X-Ray, CT, bony mixed lytic sclerotic, cotton wool lesions, vertebral picture framing

Introduction

Paget's disease of bone is a common, non-inflammatory & metabolic skeletal disorder of uncertain etiology characterized by increased osteoclast-mediated bone resorption & compensatory excessive osteoblastic activation. It is often an incidental finding on a radiological examination requested for a different indication.^[1] Frequent sites of involvement include skull (25%-65%), spine (30%-75%), pelvis (30%-75%), & proximal long bones (25%-30%)^[2].

Materials & Methods

Observational type of case study using MRI. 80 years male came with complaints of giddiness & fall & decreased speech output. CT scan showed intraventricular hemorrhage & patient was treated for the same. CT scans also revealed features suggesting Paget's disease with co-existing ankylosing spondylitis & a bone survey for whole body was done. Increased S. Alkaline Phosphatase (187 U/L) & low Calcium (7.5 mg/dl) was noted.

Results

CT Brain showed mixed lytic sclerotic changes with widened diploic space & coarse trabeculae involving calvarium giving a cotton wool appearance (Figure 5). X-Ray dorsal spine showed anterior & lateral syndesmophytic fusion involving visualised dorsal vertebrae giving a bamboo spine appearance suggesting Ankylosing spondylitis (Figure 1) X-Ray Chest shows bony expansion with mixed lytic sclerotic changes noted in anterior aspect of Rt 1-4th ribs (Figure 2). X-Ray pelvis AP view shows cortical thickening & sclerosis of iliopectineal & ischiopubic lines with obliteration of Kohler tear drop (Paget's). Sclerosis with near complete fusion of bilateral sacroiliac joints were also noted (Figure 3). X-Ray AP

of hand & elbow show generalized osteopenia & diffuse cortical thickening predominantly involving diaphysis of ulna. Erosive changes noted involving interphalangeal joints (Figure 4). CT Cervical spine showed exaggerated cervical lordosis, picture squaring of few vertebrae & anterior syndesmophytic fusion involving visualised dorsal vertebrae (D1-D5) giving a bamboo spine appearance (Figure 6) which were also evident on X-Rays. Diffuse osteopenia of visualised vertebrae along with few small foci of sclerotic areas noted (Paget's)

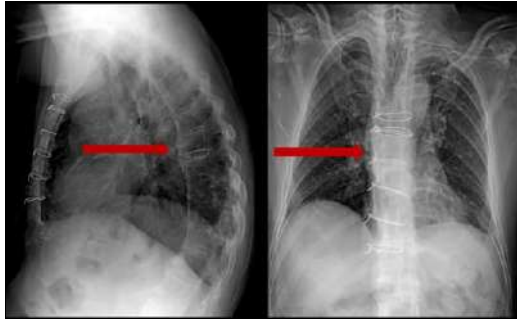


Fig 1: X-Ray dorsal spine shows anterior & lateral syndesmophytic fusion involving visualised dorsal vertebrae giving a bamboo spine appearance suggesting Ankylosing spondylitis & also seen picture framing of vertebrae.

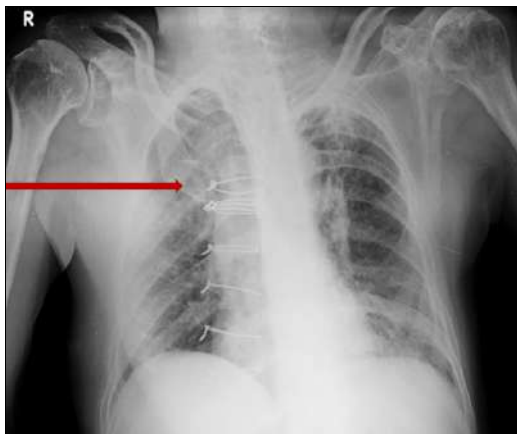


Fig 2: X-Ray Chest shows bony expansion with mixed lytic sclerotic changes noted in anterior aspect of Rt 1-4th ribs



Fig 3: X-Ray pelvis AP view shows cortical thickening & sclerosis of iliopectineal & ischiopubic lines with obliteration of Kohler tear drop (Paget's). Sclerosis with near complete fusion of bilateral sacroiliac joints noted (ankylosing spondylitis).



Fig 4: X-Ray AP of hand & elbow show generalized osteopenia & diffuse cortical thickening predominantly involving diaphysis of ulna. Erosive changes noted involving interphalangeal joints.

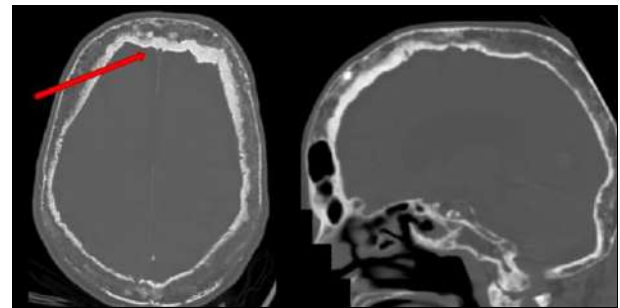


Fig 5: CT brain –bone window shows mixed lytic sclerotic changes with widened diploic space & coarse trabeculae involving calvarium fully giving a cotton wool appearance



Fig 6: CT C-spine Sag view show diffuse osteopenia of visualised vertebrae along with few small foci of sclerotic areas (Paget's). Exaggerated cervical Lordosis & anterior syndesmophytic fusion involving visualised dorsal vertebrae (D1-D5) giving a bamboo spine appearance (Ankylosing spondylitis) & few vertebrae showing picture framing.

Discussion

Paget disease is an uncommon skeletal disorder of middle aged & elderly persons characterized by abnormal & excessive remodeling of bone. An initial osteolytic phase

with a subsequent osteosclerotic phase seen on radiographs of Paget disease are diagnostic. Bone enlargement with increased radio density, enhanced trabecular pattern, & deformity is typical. Lab parameters reveal increased S. Alkaline Phosphatase (187 U/L) & low Calcium (7.5 mg/dl) which are seen in this disease. Complications include pathologic fractures, neurological symptoms, skeletal deformities, articular derangements, & secondary neoplasms ^[3]. Imaging modalities are complementary, with radiographs being vital for disease characterization & assessing complications. CT provides further detail of bony architecture when needed, particularly in spine. MRI is essential for assessment of complications, such as spinal stenosis & sarcomatous degeneration ^[4].

Conclusion

Paget disease of bone shows an abundance of patterns on imaging that are related to pathologic stage of disease ^[3]. Recognition of imaging spectrum usually allows early diagnosis & differentiation of its complications, which helps guide therapy & improve patient management ^[2].

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Author's Contribution

Not available

Conflict of Interest

Not available

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