Proceeding S.Z.P.G.M.I vol: 15(2) 2001, pp. 97-100.

Pagets Disease - A Rare Cause of Pain in Whole Skeleton

Shafique Ahmad, Faisal Masood, Bilquies A. Suleman, Munawar Ali, Hamid Qayum
Department of Orthopaedic Surgery, Shaikh Zayed Hospital, Lahore.

INTRODUCTION

We describe a case of patient with aches and pains in whole of the skeleton secondary to Paget's disease and its complications which is a rare etiology in our region.

The patient was diagnosed by open biopsy of cervical spine and was managed by bisphosphonates therapy. Though Paget's disease is commonly seen as a rare cause of pain in long bones and axial skeleton. It is rarest to see a disease with full blown complications and rapid spread in a patient especially in our region (South Asia).

CASE REPORT

A forty years old female patient, housewife, from Lahore presented through outpatient department with the presenting complaints of pain in lumbosacral, mid dorsal and lower cervical spine for the last 1 year. Pain was gradual in onset moderate to severe in intensity and was not associated with any symptoms like anorexia, weight loss and fever. Pain was localized without any radiation and without any neurological disturbance. Pain was decreased partially with medicine and rest but patient was never relieved fully. Patient had multiple consultations and medications from local practitioners. Her systemic review was non revealing except complaints of pain in right eye and facial skeleton in right side. A few days later, she also started complaints of decreased vision in her right eye. No history of arthralgia or urogenital disturbance.

In past history she had a road traffic accident 10 years back for which she was operated and open reduction and internal fixation was done for her fracture right radius/ulna and right humerus.

Examination

- General physical examination revealed healthy conscious young lady of average weight and height.
- There was no lymphadenopathy.
- There was no evidence of weight loss.

Her musculoskeletal and spine examination revealed an area of tenderness present at lower cervical region. Mid dorsal spine area and 2nd lumbar vertebra. Overlying skin was normal. There was no area of increased temperature there was no swelling or deformity of spine. Range of movement of spine was full with intact sensory motor and vascular supply of upper and lower limbs.

Investigations

Investigations were started from CBC, ESR, chest X-ray, Biochemical profile which were all normal. Bone scan revealed multiple areas of increased uptake resembling a picture of metastatic bone disease.

X-rays of cervical spine revealed area of complete resorptions in left transverse process of C7 (Fig. 1).

Dorsal and lumbosacral spine x-rays revealed areas of sclerosis present in D7 and L5 vertebrae (Fig. 2). There was no peri lesional sclerosis. No periosteal reaction and no vertebral collapse. Keeping in mind the age of patient and radiological findings following differentials were made[1].

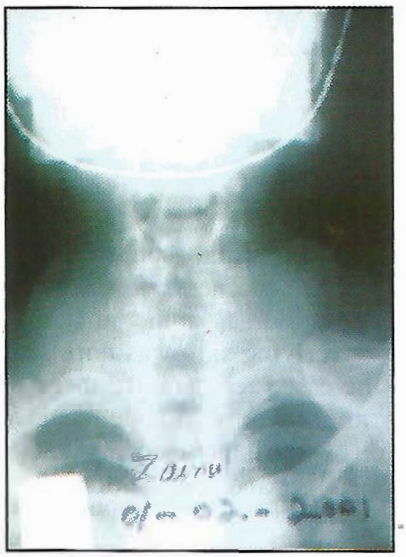
1. Tuberculosis.
2. Hyperparathyroidism
3. Lymphoma.
4. Metastatic bone disease.
5. Fibrous dysplasia

Other Investigations

LFTs, RFTs, were within normal range, only Alkaline phosphatase was high. Bone scan showed increased uptake in all areas of lesion plus increased uptake on right side of facial skeleton and base of skull alongwith above said hot areas in spine. USG abdomen did not show any visceral mass.

Complete screening for hyperparathyroidism including serum PTH and parathyroid scan was also within normal range.

Bilateral mammography was done twice to find any primary site. But it also did not revealed any suspicious areas.



1: Osteolytic lesion in transverse process of C7 vertebra.

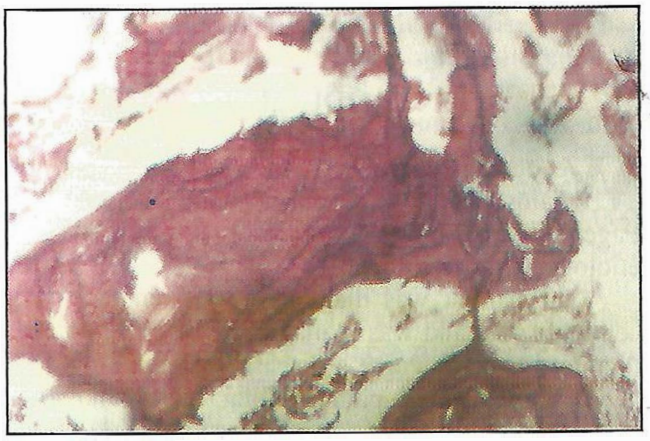


Fig. 3: Mosaic pattern of trabecular bone (H&E stain X100).

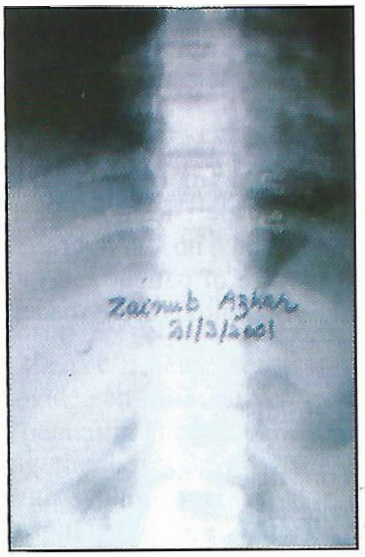


Fig. 2: Osteoblastic lesion in D7 vertebra.

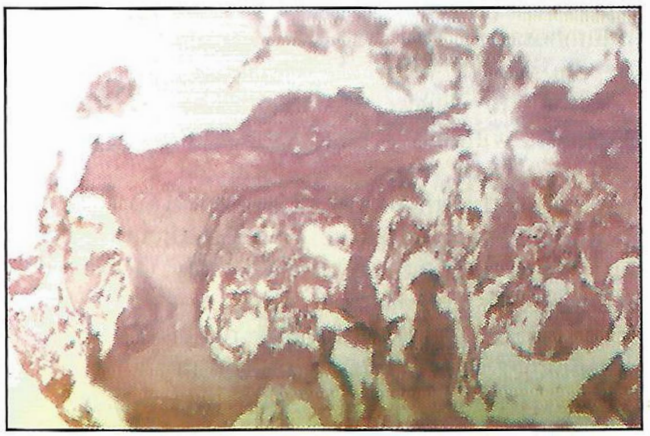


Fig. 4: Mosaic pattern with osteoblastic and osteoclastic activity (H&E stain X100).

FNA, done under general anaesthesia from L2 and D8 areas revealed only granulation tissue with out any malignant cells.

Failing the diagnosis with all these investigations patient was advised open biopsy, which was taken from C7 transverse process. The specimen included resorbed bones and soft tissues. There was no pus. Her histopathology report was highly suggestive of Paget's disease, with multiple areas of bone resorptions and bone sclerosis (Figs. 3-5).

DISCUSSION

Pagets disease is a rare etiology of musculoskeletal pain in our region as compared to western hemisphere. Incidence varies from region to region from 3.5% to 10%. Incidence also increases with age so that by age of 80 incidence may reach up to 10%[2,3]. It is a disorder that produces focal enlargement and deformity of the skeleton and was originally thought to be an

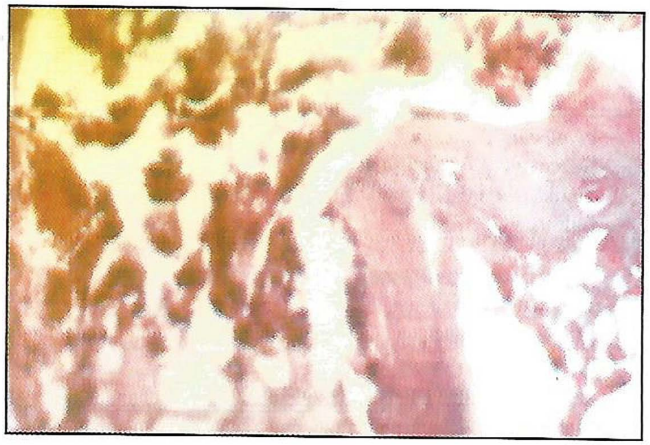


Fig. 5: Mosaic pattern with osteoblastic and osteoclastic activity (H&E stain X400).

inflammatory disorder, being termed osteitis deformans. It is characterized by excessive bone resorptions and subsequent excessive bone formation due to over activity of bone osteoclasts and osteoblasts. The disorder may be monostotic or polyostotoic[4].

Major complications include high out put heart failure, Paget's sarcoma and platybasia (remodeling of skull base) leading to blindness (as seen in our patient). Orthopaedic surgery may be advised in severe deformity of long bones and to relieve nerve compression[5,6,7].

In our patient, some secondaries seemed more likely to be the cause. But regarding the duration and slow progression of symptoms one should keep in mind rare causes like pagets disease.

Regarding the radiological features lesions in spine look like areas of sclerosis and as present in our case. Vertebral bodies may enlarge with thick margins and central coarse striations. In skull there are multiple areas of resorptions and calcification giving the appearance of "cotton wool skull"[3,4].

Histological features include excessive osteoblastic resorptive activity. Bone marrow is replaced by fibrous tissue and disorganized trabeculae. Woven bone with osteoblastic rimming is also seen[8].

Bone scintigraphy provides more specific and precise results regarding the unifocal or multifocal lesions according to patient's condition and disease severity[9,10].

As far as the medical treatment is concerned two major types of agents are available to suppress disease activity. These are calcitonin and bisphosphonates. Calcitonin acts directly on osteoclasts to inhibit bone resorptions by binding to hydroxyapatite crystals and inhibiting their growth and dissolution[11-13]. Risedronate is a new agent in this regard with promising results and few side effects.[14]

Alkaline phosphatase (from increased osteoblastic activity) can be used as an index of disease activity[9,15].

REFERENCES

1. James O Johnston, Lor Randall. Tumouurs in orthopaedic. In: Harry B. Skinner eds. Current Text Book of Orthopaedic Surgery. 2000; 316-17.
2. Pande KC, Arsford RU, Dey A, Kayan K. Atypical form of Paget's disease of bone: incidence and literature review. J Bone Spine J 2001; 68 (3): 257-66.
3. Nigel H Harris. Metabolic bone disease. In: Nigel H. Harris, Rolfe Birch eds. Postgraduate Textbook of Clinical Orthopaedics. Blackwell Science, 1995; 370.
4. Fransen P, Mestdagh C, Dardenne G. Paget's sarcoma of skull; report of 02 cases. Acta neurol Belg 1998; (4): 352-5.
5. Hadjipavlou AG, Gaitanis LN, Katonis PG, Launder P. Paget's disease of spine and its management. Eur Spine J 2001; 10 (5): 370-84.
6. Walpin LA. Emg in vertebral Paget's disease. Arch Phys Med Rehabil 1980; 61 (4): 196-97.
7. Walpin LA, Singer FR. Paget's disease; reversal of severe paraparesis using calcitonin. Spine 1979; 4 (3): 213-19.
8. Reddy SV, Kunhara N, Menaa C, Roodman GD. Paget's disease of bone: a disease of osteoblasts and osteoclasts. Rev Endocr Metabol Disord 2001; 2(2): 195-201.
9. Roodman GD. Studies in Paget's disease and their relevance to oncology. Semin Oncol 2001; 28 (4): 15-21.
10. Jesser H, Zita g, Drexler H. Bone scintigraphy in osteodystrophia deformans (Paget's disease). Frotschr Med 1997; 3: 95 (41): 2451-58.
11. Grauer A, Klar B, Kans J, Scharla, Ziegler R. bisphosphonate therapy of Paget's disease of bone with paimidronate. Med Clin 1996; 15: 91 (1): 41-49.
12. Dewis P, Prasad BK, Anderson DC, Willets S. Clinical experience with the use of two diphosphonates in the treatment of Paget's disease. Ann Rheum Dis 1985; 44 (1): 34-8.
13. Purohit OP, anthony c, Radstone CR, Owen J. High dose intravenous palmidronate for metabolic bone pain. Br J Cancer 1994; 70 (3): 554-8.
14. Unland EM, Boyee EG. Risedronate. A new oral Bisphosphonate. Clin Ther 2001; 23 (9): 1409-21.

15. Alvarez L, Gaunabens N, Peris P, Vidal S, Ros I. Usefulness of biochemical markers of bone turnover in assessing the response to treatment of Paget's disease. Bone 2001; 29 (5): 447-52.

The Authors:

Shafique Ahmad,
Associate Professor,
Department of Orthopaedic Surgery,
Shaikh Zayed Hospital,
Lahore.

Faisal Masood,
Department of Orthopaedic Surgery,
Shaikh Zayed Hospital,
Lahore.

Bilquies A. Suleman
Assistant Professor Histopathology
Shaikh Zayed Hospital,
Lahore.

Munawar Ali,
Department of Orthopaedic Surgery,
Shaikh Zayed Hospital,
Lahore.

Hamid Qayum
Professor
Department of Orthopaedic Surgery,
Shaikh Zayed Hospital,
Lahore.

Address for Correspondence:

Shafique Ahmad,
Associate Professor,
Department of Orthopaedic Surgery,
Shaikh Zayed Hospital,
Lahore.