Paget's Disease of Bone

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Abstract

Background: Paget's disease of bone is a focal disorder of bone metabolism that results in enlargement of the affected bone and deformity, a process that is sometimes accompanied by pain and/or disfigurement [(1)]. The disease is common in Western Europe, USA, and Australia but very rare in Middle East and Arabian Peninsula. Here we reported seven un-relative patients with Paget's disease in Karbala between 1995-2005. Their ages ranged between 51-75 years. One patient was polyostotic and the remaining were monostotic types. Four cases presented with fracture femur, the 5th patient with undisplaced fracture tibia and the last two cases with pain in tibial bones and knee joints. Radiological criteria of Paget's disease were more obvious in cases of tibial involvement than femoral cases in which histopathological examination confirmed the diagnosis. Serum alkaline phosphatas at time of presentation ranged between 20-34 KAU/100ml, the higher level was in polyostotic case. The purpose of this paper is to shed light on this rare entity and the variety of presentations.

Key wards: Karbala, Paget's disease of bone, Serum alkaline phosphatas.

Introduction

As early as 1877 Sir James Paget described his observation on five middle-aged individuals with chronic deformity bone disease & named the condition as osteitis deformans[(2)]. Paget's disease is a focal disorder of bone turnover characterized by initially increased bone resorption leading to osteolytic lesion which is the site of subsequent increased bone formation producing sclerosis[(3)]. The exact etiology is unknown, but the slow viral hypothesis is attractive[(3)].

The majority of patients are asymptomatic. Those with symptoms commonly present with musculoskeletal complaints including pain, osseous deformities, fracture, back pain, hearing loss, neurological abnormalities, degenerative joint disease & rarely bone neoplasm. The diagnosis depends on: (1) Clinical features, (2) Radiological criteria, (3) Elevated S Alk-Ph. and (4) Biopsy is rarely indicated.

The disease is common in Western Europe, USA, & Australia[(4)] but very rare in Middle East & Arabian Peninsula[(5)]. To our knowledge, there is no previously reported study in Karbala, this study discus presentations of seven un-relative patients of Paget's disease of bone reported between Jan.1995 – Dec.2005

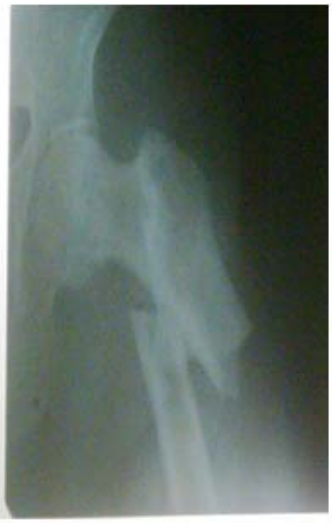
Patients

Patient No. (1) (Female) 51 years, Bedouin from (AL-Nekhab). Presented with pain & inability to walk on her Rt leg for one week duration following trivial trauma. She had a history of dull aching pain with progressive bowing of Rt leg for 10 years. Examination revealed a healthy looked but hypertensive The Rt. leg looked bent, thick, tender & warm skin. Movements of Rt. knee were painful. No family history of bone disease. X-ray showed criteria of Pagetic changes in Rt. tibia (expansion of bone,

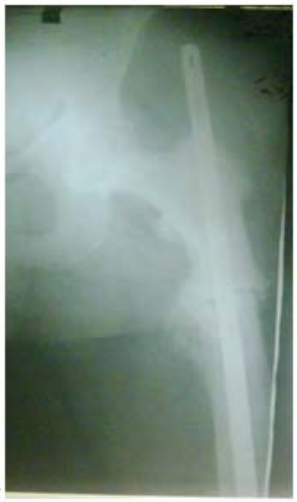
cortical thickening, coarsing of trabecular markings & lytic and sclerotic changes) with un-displaced transverse fracture in upper third. Skeletal survey showed no abnormalities apart of lumber spondylosis. S. Alk-Ph 20 KAU /100 ml. The final diagnosis depended on radiology & increased S. Alk-Ph.

Patient No. (2) (Female) 52 years, from AL-Najaf, (الشـامية) Presented with pathological fracture of Lt. femur. No history of previous localized complaint. On examination she looked healthy. No family history of bone disease. X-ray showed complete displaced transverse fracture of upper third of Lt femur. Skeletal survey showed no other abnormalities. S. Alk-Ph 28 KAU /100 ml. Treated by internal fixation using K-nail and suspicion of the diagnosis formed during operation, that (a) the bone ends were unusually hard sclerotic with partial obliteration of medulla which needed reaming to widen it (b) the prepared length K-nail was difficult to insert distally because of bone bowing that necessitated to change it by shorter nail (c) biopsy taken & the result was Paget's disease. The final diagnosis depended on raised S. Alk-Ph & histopathology.

X-ray of 1st patient



X-ray of 2nd patient



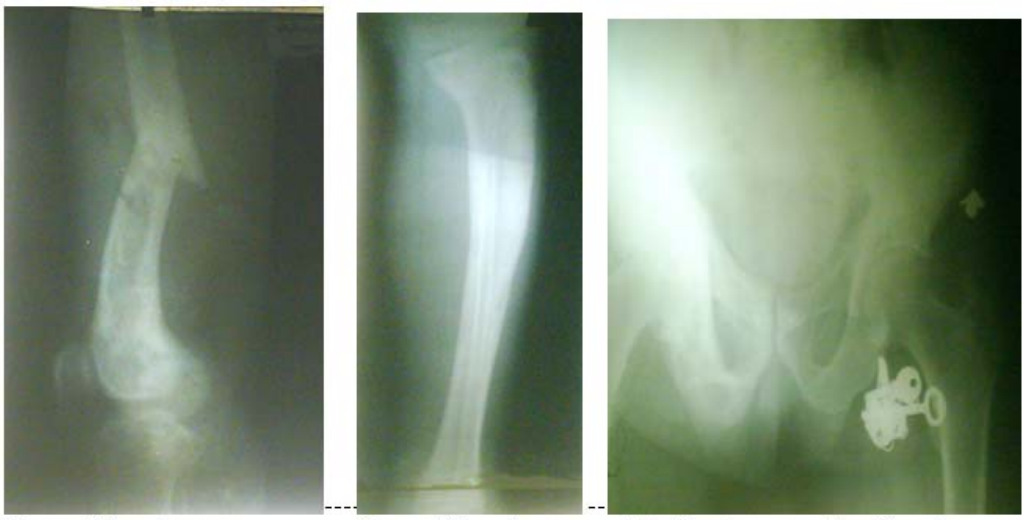
Post-op. of 2nd patient

Patient No. (3) (Male) 55 years, from Karbala, Presented with fracture Lt. femur following road accident. Examination. revealed a healthy looked but subnormal mentality. No family history of bone disease. X-ray showed oblique fracture of lower half of Lt femur. Skeletal survey showed no other abnormalities.

Treatment by internal fixation using K-nail and suspicion of the diagnosis also formed during operation, that the bone ends were unusually hard sclerotic, so piece of bone taken for histopathology & the result confirmed the diagnosis of Paget's disease. The prepared length K-nail inserted without technical difficulty. Next post-operative day, (S. Alk-Ph) was 20 KAU/100ml. The final diagnosis depended on raised S. Alk-Ph & histopathology.

Patient No. (4) (Male) 72 years, from Babel, AL-Mussiab. Presented as chronic pain in Lt. knee causing some problems with movement & aggravated at night for six years duration. Examination revealed elderly healthy-looked patient, & apart of partial deafness (for 6 years) no peripheral neurological deficit. Local examination revealed swelling, mild bowing & tenderness of upper third of Lt. leg, Movement of Lt. knee was painful & limited. X-ray showed some criteria of Pagetic changes in upper half of Lt tibia (bone expansion, cortical thickening & coarsing of trabecular markings). Skeletal survey showed Pagetic criteria of Rt pubic rami. Skull &

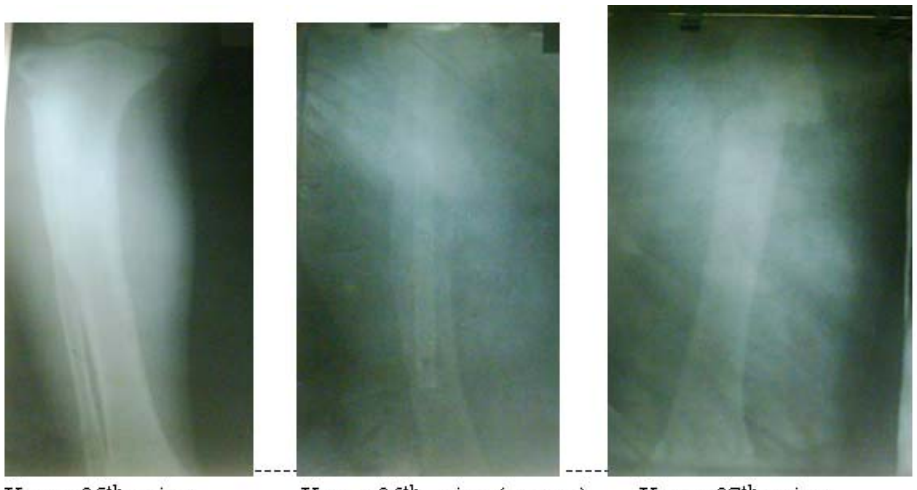
spinal x-ray not significant. S. Alk-Ph was (34 KAU/100ml). The final diagnosis depended on radiology & increased S. Alk-ph.



X-ray of 3rd patient X-ray of 4th patient Pelvis involvement of the 4th patient

Patient No. (5) (Male) 62 year, from Karbala (فريحة) Presented as chronic dull aching pain with feeling of hotness in his Rt leg for 8 years duration. The pain is aggravated by weight-bearing & relieved by simple analgesia. No family history of bone disease. Examination revealed a healthy looked, but locally there was antero-lateral bowing of Rt leg at upper third with mild tenderness, & warm skin. Movement of Rt. knee was painful. X-Ray showed criteria of Pagetic changes (bone expansion cortical thickening, coarsing of trabecular marking with antero-lateral bowing). Skeletal survey showed no other abnormalities. S. Alk-Ph, 23 KAU/100ml. The final diagnosis made depended on radiology & elevated S. Alk-Ph.

Patient No. (6) (Female) 70 years, from Karkalla (حي العامل), Presented with pathological fracture Rt. femur. No history of previous localized or generalized illness. No family history of bone disease. There was nothing remarkable on general examination. X-ray showed complete displaced short oblique fracture of upper third of Rt femur. Skeletal survey showed no other abnormalities. S. Alk-Ph. 20. KAU /100ml. Treatment by internal fixation using K-nail and operative findings were similar to case No-2-. The final diagnosis depended on raised S. Alk-Ph & histopathology.



X-ray of 5th patient X-ray of 6th patient (post-op) X-ray of 7th patient

Patient No. (7) (Female) 75 years, from Karbala, .(حي العاملل)Presented with pathological fracture Lt. femur. No history of previous localized or generalized illness. No family history of bone disease. Examination revealed nothing remarkable on general examination. X-ray showed complete displaced transverse fracture of upper third of Lt. femur. Skeletal survey showed no other abnormalities. S. Alk-Ph. 23. KAU /100ml. Treated by internal fixation using K-nail and operative findings also were similar to case No-2-. The final diagnosis depended on raised S. Alk-Ph & histopathology.

Results

Table No-1- show the results of epidemiology

Pt. No	Date	Sex	Age	Occupation	Residence
(1)	Jan--1995	F	51	House wife	Kabala(Al-Nekhab)
(2)	Dec--1996	F	52	Farmer	Al-Najaf (Shamiah)
(3)	Nov--1997	M	55	Worker	Karbala
(4)	Apr--2000	M	72	Retired military	Babel (Al-Mussiab)
(5)	Oct--2000	M	62	Farmer	Karbala
(6)	Apr--2001	F	70	House wife	Karbala
(7)	Dec--2005	F	75	House wife	Karbala

Table No-2- show the results of clinical features

Pt. No.	Presentation	Previous History	Gen. Ex.	Local Ex.
(1)	Pain & inability to walk on Rt leg	Dull pain For 10 y.	Hypertension	Bent, thick, tender Rt leg
(2)	Pathological # Lt femur	--Ve	Healthy	# Lt femur
(3)	Traumatic # Lt femur	--Ve	Subnormal mentality	# Lt femur
(4)	Pain in Lt knee	For 6 y.	Partial deafness	Bent, thick, tender Lt leg
(5)	Dull aching pain in Rt leg	For 8 y.	Healthy	Bent, thick, tender Rt leg
(6)	Pathological # Rt femur	--Ve	Healthy	# Rt femur
(7)	Pathological # Lt femur	--Ve	Healthy	# Lt femur

Table No-3- show the results of investigations

Pt. No.	S. Alk-Ph (KAU/100ml)	Radiology	Skeletal Survey	Biopsy
(1)	20	Criteria of paget's	(-- Ve)	
(2)	28	No obvious signs	(-- Ve)	(+ Ve)
(3)	23	Criteria of paget's	(-- Ve)	(+ Ve)
(4)	34	Criteria of paget's	Pelvis involved	
(5)	25	Criteria of paget's	(-- Ve)	
(6)	22	No obvious signs	(-- Ve)	(+ Ve)
(7)	23	No obvious signs	(-- Ve)	(+ Ve)

Discussion

Incidence: Paget's disease of bone is a slowly progressive metabolic disorder of bone[1] which not hormone dependent. It is common in western Europe & in countries that have colonized by western European but very rare in the Middle East that could be due to a difference in the genetic make up of the population & the geographical & climatic influence of the environment on the disease[6]. Doter has described the characteristics of 278 cases in Palestine, of which all patients were Jewish & none Arabs[7]. Woodhouse described the first Saudi Bedouin patient suffered from Paget's disease in 1988[5]. A decade since the first reported case, Mona Founda described another case in a Saudi female[6]. In symposium in Al-Najaf hospital 1985, Dr. Malik Jasim reported four cases.

Family clustering has occasionally been noted[4], (~ 15%). No similar bone disease in relative of our patients.

Age incidence is mostly developed after the age of 40-50 years & rarely below 25 years[7]. The range of age at time of presentation in this study was between 51-75 years.

Clinical features: The onset is insidious & progress is slow. It is either monostotic or polyostotic[7, 8]. The 4th patient of our series was polyostotic type (involvement of tibia & pelvis) but the remaining were monostotic at time of presentation. Any part of the skeleton may be affected[4], but the pelvis & tibia being the commonest site, the femur, skull, spine & clavicle the next commonest [7,9]. In this study, four cases with femoral bone, three with tibial bone & one with pelvis involvement.

Most patients are asymptomatic[1-4,]. The most common presenting feature is dull constant localized pain, worse in bed when the patient warms up[9] & aggravated by exercise & motion[7]. Nocturnal pain which may waken the patient is common[4,10]. Localized pain & tenderness was the main presentation for cases with tibial bones involvement in our patients.

The 2nd presentation is thickening & bowing of long bone[4,5,11]. Bowing was obvious clinically in the tibial cases, & operatively in femoral bones in which shorter K-nails inserted in 3 patients.

Osteoarthritis of joints adjacent to involved bones is extremely common[4,5] & may have 2 etiologies: (a) mechanical malalignment due to deformity, or (b) the joint may itself be distorted by expansion of an involved bone[3]. All 3 patients with tibial involvement suffered from arthritic pain of adjacent knee.

Fractures are common, especially in the weight bearing long bones[9]. Incomplete fracture can be found on the convex side[12,13], & may cause considerable pain[10], as seen the 1st patient with facture tibia, Complete fractures are either traumatic or pathological[7]. Pathological long bone fracture is the most common presentation. Although involved bones are prone to fracture because their disturbed structural architecture, which results in weakness despite increased radiological density[10]. Three of our patients with fracture femur were pathological & the 4th one was traumatic.

Radiologically: The lesions may be osteolytic or osteoblastic, & in most cases a mixture of two changes are seen[2]. The appearances are so characteristic that the diagnosis is seldom in doubt. (1) expansion of bone, (2) cortical thickening, (3) coarsing of tarbecular markings & (4) typical lytic & sclerotic changes may be found[7], as seen in all three patients with tibial bone presentation. Occasionally, the diagnosis is made only when the patient presents with a pathological fracture as happened in three cases of our patients with femoral bone fracture without classical criteria of pagetic changes, & the diagnosis confirmed by histopathology.

Biochemical parameters: S. Alk-Ph in blood with normal level 3-13 KAU/100ml (21-100I.U./l). It is increased in conditions in which there is marked osteoblastic activity. S. Alk-Ph is

the simplest useful & sensitive marker of disease activity & extent. The patients with highest levels (10 times the upper limit of normal or greater) typically have involvement of the skull as at least one site of the disorder[7]. Active monostotic disease (other than skull) may have lower value than polyostatic disease. Lower values (less than 3 time the upper limit of normal) may reflect a less extent of involvement[7]. In this study, the S. Alk-Ph ranged between 20-34 KAU/100ml & the highest level was in the 4th patient with polyostotic disorder.

References

1- The New Registry for Paget's Disease of Bone, 55 fruit Street, Bulfinch 165, Boston, MA 02114.

2- Abdul H. Zargar et al, Paget's Disease of Bone, Saudi Medical Journal (1999); 20 (8): 629-632.

3- Duthie R. B. and Bently G., MERCER'S Orthopaedic Surgery, 9th Ed. (1996), Vol. 2 (5): 326-332.

4- Stevenson, J. C., Paget's Disease of Bone, Medicine International (1985); (14): 619-621.

5- Woodhouse N. J. Y. and El-Baage T., Paget's Disease in a Bedouin, Saudi Medical Journal (1988); 9 (1): 78-82.

6- Fouda M., Paget's Disease of the Bone, Saudi Medical Journal (1998); 19 (2): 205-208.

7- Favus M. J. et al, Primer On The Metabolic Bone Disease And Disorder Of Mineral Metabolism, 2nd Ed. (1993); 83: 375-383.

8- Frohlich E. D., Pypins Basic Sciences Review, 16th Ed. (1993): 636.

9- Solomon L., Warwick D. J., and Nayagam S., Apley's System of Orthopaedics & Fracture, 8th Ed. (2001) 7: 126-129.

10- Hodkinson M., Paget's Disease of Bone, Medicine Meddle East, (1978); 4: 265.

11- Ronald L. E., Skull and Spine Imaging, An atlas of differential diagnosis, New York, (1994): 16

12- CANALES. T., CAMPBELL'S Operative Orthopaedics, 9th Ed. 1998, Vol. (1): 758.

13- Fauci, et al, Harrison's PRINCIPLES OF IMERNAL MEDICINE, 14th Ed. 1988 (358)2266-2269