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CASE REPORT

Idiopathic multicentric osteolysis : A case report and literature review

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INTRODUCTION

Osteolysis is defined as destruction of bone by resorption. Usually this resorption is associated with some underlying disorder and classified as secondary osteolytic syndromes.

Primary idiopathic osteolysis is rare. It is characterised by the spontaneous onset of bone resorption without known causative factors. Bones which previously appeared normal begin to undergo partial or complete resorption. This process continues for years, until eventually it ceases spontaneously. The exact pathogenetic mechanism of this osteolysis remains unknown.

We present a unique case of idiopathic osteolysis, that does not fit into the previously described patterns of the disease.

CASE REPORT

A 64-year-old previously healthy Caucasian male presented with a 6-month history of pain in his knees and right groin. Radiograph of the pelvis revealed complete osteonecrosis of the femoral head with secondary changes in the acetabulum consistent with secondary osteoarthritis. No apparent cause for the osteonecrosis was found.

The patient underwent a cemented Charnley total hip replacement. Postoperative recovery was uneventful. A sample of the capsule was sent for histological examination, which revealed fibroblastic changes.

A few months later, the patient presented with a dislocated hip. This was uneventfully reduced under anaesthesia. In the following months he presented with recurrent dislocations. Radiographs revealed progressive bone resorption adjacent to the femoral component.

This mysterious resorption of bone was responsible for the recurrent dislocation which ultimately led to resection arthroplasty. This represents one of the rarest causes for recurrent dislocation in the literature.

At 10 months follow-up he complained of pain and weakness of the right shoulder with inability to raise his arm above his head. There was no history of trauma, fever, night sweats or weight loss.

Clinical examination revealed loss of contour of the right shoulder with moderate deltoid wasting. Shoulder movements were restricted. The neurological examination of the upper limb was unremarkable.

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Table I. — Classification of idiopathic osteolysis according to Hardegger *et al* (15)

1.	Hereditary multicentric osteolysis with dominant transmission	Between the ages of two and seven years, spontaneous pain and swelling begin in the hands and feet. Carpo-tarsal osteolysis occurs over the period of a few years. Progression ceases normally in adolescence
2.	Hereditary multicentric osteolysis with recessive transmission	Similar to Type 1, but may be associated with severe generalised osteoporosis.
3.	Non-hereditary multicentric osteolysis with nephropathy	Appears in childhood. There is a gradual disappearance of the carpus with the tarsal bones involved but to a less degree, and an association with proteinuria. Death occurs usually due to renal failure and malignant hypertension.
4.	Gorham's massive osteolysis (Gorham-Stout syndrome)	Monocentric occurrence in any part of the skeleton may start at any age. Normally "hemangiomatous" tissue is found in the osteolytic region. It has neither a hereditary pattern nor an associated nephropathy. The disease is benign and the osteolysis usually stops after a few years.
5.	Winchester syndrome	Autosomal recessive transmission. Rare childhood carpo-tarsal osteolysis in association with contractures, shortness of stature, skin lesions, corneal clouding and osteoporosis without nephropathy.

Radiographs of the shoulder revealed bilaterally symmetrical erosion of the humeral head with resorption of the lateral end of the clavicle. A skeletal survey also revealed resorption of the medial condyle of the left tibia which had progressed rapidly over 10 months. An MRI scan of the brain and spinal cord were obtained to rule out syringomelia as a possible cause.

Blood investigations were done to rule out neurosyphilis, rheumatoid arthritis, autoimmune disorders including systemic lupus erythematosus and diabetes. Investigations revealed a normochromic normocytic anaemia. He also had hypogammaglobulinaemia and hypoalbuminaemia with a low plasma viscosity. A biopsy was sent from the left shoulder joint which was reported to be fibrotic synovium with a few hyperplastic surface synovial lining cells and containing a small quantity of brown pigment consistent with haemosiderin. An aspirate from the shoulder joint demonstrated mixed inflammatory cells and occasional synovial lining cells possibly due to inflammation. The aspirate was also sent to investigate for mycoplasma arthritis commonly seen in patients with hypogammaglobulinaemia ; this was found to be negative.

DISCUSSION

Lytic lesions of bone are a common radiological finding. Usually the destruction is associated with some underlying disorder. From these so-called secondary osteolytic syndromes, the rare entity of idiopathic osteolysis can be differentiated. Local and systemic conditions which are associated with secondary osteolysis include disuse atrophy, acute inflammatory atrophy associated with trauma (Sudeck's atrophy or algodystrophy), primary and metastatic tumours, hyperparathyroidism, gout, congenital pseudoarthrosis, granulomatous diseases (sarcoidosis), rheumatoid arthritis, diabetes mellitus, psoriatic arthritis, osteomyelitis, systemic mastocytosis, aseptic necrosis, neurogenic arthropathy, prolonged steroid therapy, bone aneurysm, cystic angiomatosis of bone and occupational exposure to vinyl chloride.

Idiopathic osteolysis comprises a heterogeneous group of rare diseases, characterised by the spontaneous onset of mostly peripheral osteolysis, without obvious cause. It must be differentiated from familial and sporadic cases and from multicentric and unicentric cases. Hardegger *et al* (*15*) proposed a classification with five types of idiopathic osteolysis, based on the reports of Torg *et al* (*31*) and Macpherson *et al* (*21*) (table I).

Idiopathic osteolysis was first described in 1838 and again in 1872 by Jackson (*18, 19*) who reported a case of "boneless arm". The humerus of an 18-year-old man disappeared completely in the course of 11 years, during which he sustained a spontaneous fracture of the bone. In spite of the disease he

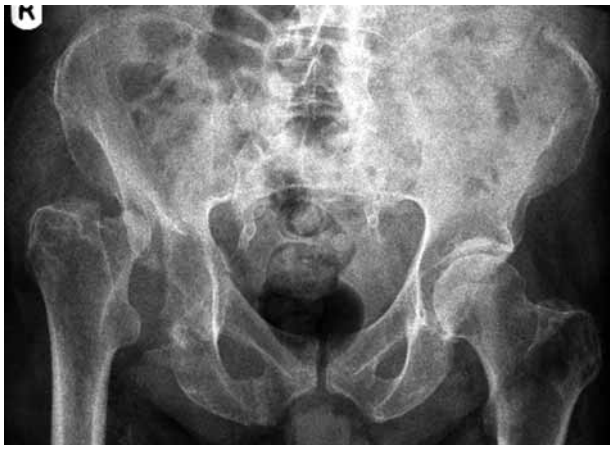

Fig. 1. — Radiograph of hip showing complete osteonecrosis of femoral head.

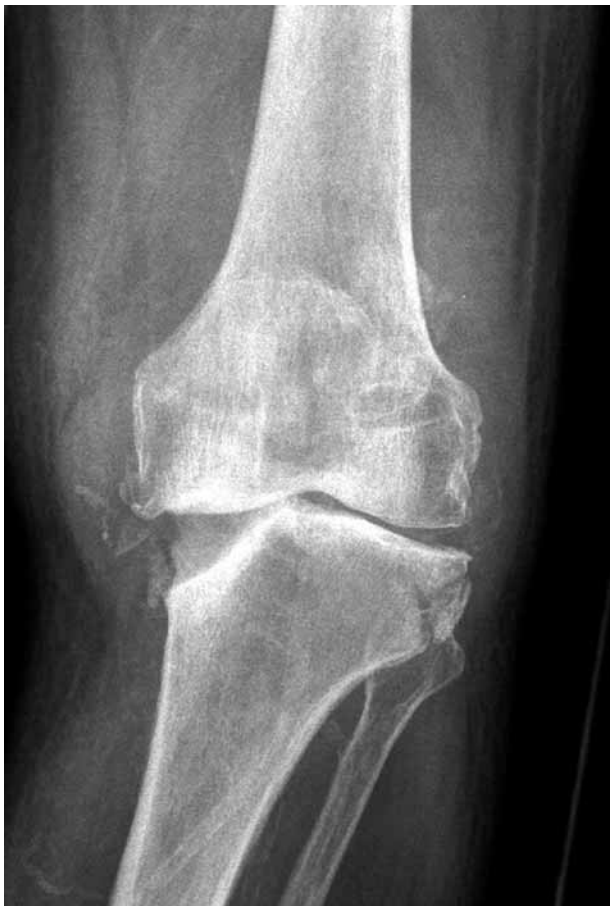

Fig. 2. — Radiograph of the knee showing resorption of the medial tibial condyle.

was able to do manual labour until his death at the age of 70 years.

Multicentric osteolysis can be divided into a hereditary form with either dominant or recessive transmission and a non hereditary variety with nephropathy. They usually present in childhood or adolescence. The monocentric type of osteolysis comprises of Gorham Stout syndrome and Winchester syndrome.

Most cases of multicentric idiopathic osteolysis are accompanied with the resorption of carpus and tarsus (Carpo-tarsal osteolysis) – table I. Although this variety of osteolysis can affect a variety of bones and joints, involvement of the clavicles or the shoulder has not been described (*34*).

In 1955 Gorham and Stout (*11*) defined a specific disease entity and reviewed 24 cases from the literature. The Gorham Stout syndrome or "vanishing bone disease" is a rare, chronic condition characterised by widespread skeletal destruction complicated by lymphangiomatosis or haemangiomatosis usually with neurological and respiratory impairment. It presents as progressive idiopathic osteolysis of one bone or contiguous bones, usually unicentric. It may affect any part of the skeleton, but most commonly involves the skull, shoulder and pelvic girdle (*8, 11, 12, 17*). It is a rare skeletal disorder of which the aetiology and pathogenesis remain unknown. There may be dominant inheritance, yet this has not been documented (*33*). In no case was there a family history of a similar condition (*11*). Spontaneous fractures are common. Regeneration of the bone does not occur even when the osteolysis ceases to progress (*30*). Affected patients range in age from 1.5 to 72 years, but in most cases the disease occurred in the second or third decade. The sex inheritance is equal, no racial predominance has been found and there is lack of evidence of pre-existing disorders (malignancy, infections, neurologic, endocrine or metabolic).

The diagnosis is essentially one of exclusion and must be based on the combined clinical, radiological and histopathological findings.

No treatment to date has proven effective in arresting this disease. Spontaneous arrest of

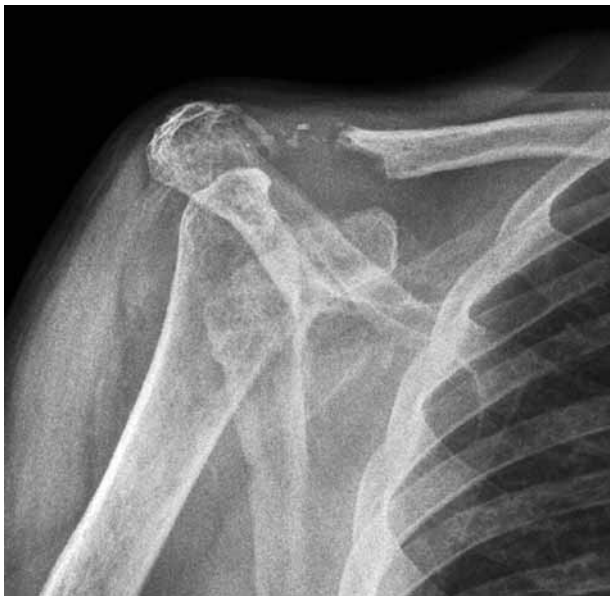

Fig. 3. — Radiograph of the right shoulder showing destruction.

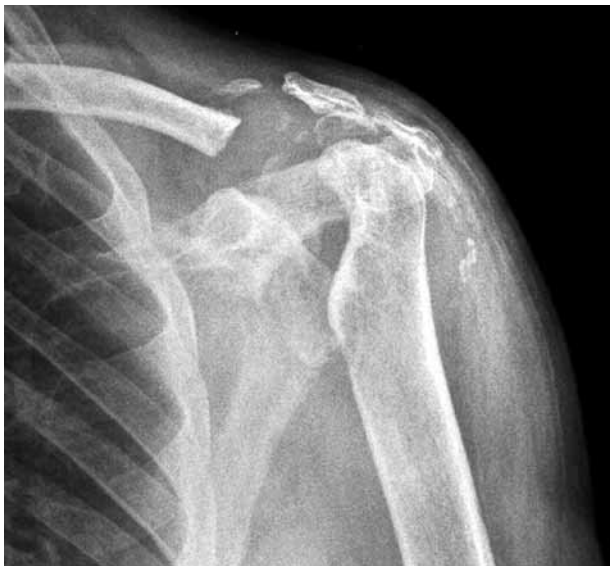

Fig. 4. — Radiograph of the left shoulder showing destruction

osteolysis after years of bone destruction is common (*11*). The end result is severe deformity and functional disability.

Drug therapy is limited to analgesics, while the benefits of bisphosphonates are unconvincing (the presence of osteoclasts is controversial) and alpha-2b interferon (used for vessel proliferation disorders) is being evaluated. If the process is progressive or the osteolysis extensive, treatment by local resection, with or without replacement by a prosthesis, radiotherapy or even amputation has been tried. Curettage with incomplete resection rarely cures the disease (*4, 25*). Bone grafts failed to arrest the disease and were resorbed (*8*), but resection and artificial bone replacement seem to be effective. Depending on the extent of the disease, amputation may be required (*1*). Only three successful cases of local resection and bone grafting, including one vascularised fibular graft have been reported (*3, 24, 33*). Most grafts are resorbed and incorporation is rare. Other forms of surgical treatment, including placement of endoprostheses for cases involving long bones have met with some success.

Radiation therapy, alone or combined with surgical stabilisation is equally promising. Radiotherapy using total doses from 30 Gy up to 45 Gy has been reported to arrest the osteolysis (*6, 8, 10, 14*) but not all cases respond (*5, 13, 29*). Immobilisation of the affected bone does not influence the prognosis (*16*). Improvement did not occur after the administration of oestrogen, androgen, magnesium, calcium fluoride, adrenal extracts, Vitamin D, aluminium acetate solution, ultraviolet radiation, ionised calcium, somatotrophin, placental extracts, Vitamin B12, amino acids or transfusions of placental blood or blood from young growing children (*3, 23, 28*).

Persistant chylothorax secondary to chest wall involvement often leads to death with circulatory failure and cachexia (*8*). Deaths from the disease have occurred in patients in whom the process was localised to the rib, mandible or vertebral bodies leading to fatal complications from respiratory failure, obstruction of the airway or compression of the spinal cord (*7, 12, 30*).

Apart from idiopathic osteolytic syndrome, another close possibility is “Rapidly Destructive Arthropathy”, also known as rapidly progressive hip disease. This is a rare and incompletely understood disorder characterised by dramatic destruction of a previously normal-appearing hip joint

over a relatively short time period. The disease was first described in the European literature by Forestier in 1957 (*9*) but was not known in the American literature until Postel and Kerboul's description in 1970 (*26*). Although the pathogenesis is poorly understood, Komiya *et al* (*20*) have found elevated levels of IL-1ß in the joint fluid of affected patients as well as increased secretion of matrix metalloproteinases by the affected synovium (as compared with conventional osteoarthritis), suggesting that excessive production of these bone resorptive factors may mediate the rapid joint destruction.

The two largest published clinical series are those of Rosenberg *et al* (*27*) and Bock *et al* (*2*), containing 27 and 22 patients, respectively. In both series, women outnumbered men, the average of the patients was in their 70s, and more than 80% of cases were unilateral. In one series, 5 patients had involvement of joints other than the hip, suggesting that the disorder may represent a focal manifestation of a systemic disease process. In all cases, the radiographic progression was from a normal-appearing joint to significant collapse, typically over a course of less than 12 months. Patients typically presented with incapacitating pain but maintained good range of motion of the hip. Both investigators describe a characteristic "hatchetlike" deformity of the femoral head that develops as it collapses, with small or absent osteophytes.

Several types of crystal-induced arthropathy can also cause rapid hip destruction (*22*). Calcium pyrophosphate dihydrate (CPPD) deposition is typically characterised by prominent osteophyte formation, sclerosis, and subchondral cysts ; patients will often have a history of recurrent pseudogout attacks. Apatite deposition can produce a similar syndrome, most commonly in the shoulder ("Milwaukee shoulder"). Detection of crystals in the joint fluid confirms the diagnosis.

Because of the degree of joint deformity and the patient's level of disability, the typical treatment of rapidly destructive arthropathy is total hip arthroplasty. Several investigators have reported excellent pain relief with this procedure, with outcomes at 5 years comparable to those of total hip arthroplasty performed for conventional osteoarthritis (*26, 32*).

CONCLUSION

This case is unique in that it does not fit into any of the yet described patterns of osteolytic syndromes. The presence of multicentricity and absence of lymphangiomatosis or haemangiomatosis virtually rules out the possibility of "The Gorham Stout syndrome". Whether it is a multicentric variant of "Rapidly destructive arthropathy" or a new disease entity cannot be said with certainty.

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