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Massive pelvic osteolysis in the Gorham-Stout syndrome

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Summary. *Massive osteolysis is a very rare tumour-like lesion usually affecting young adults. There are 132 reported cases which we have reviewed, including 31 single cases of pelvic involvement. The term "massive osteolysis" is based on the typical radiological findings, such as increasing translucency and loss of bone density. The diagnosis of the Gorham-Stout syndrome must be confirmed by the microscopic finding of intramedullary vascular structures. Our patient was a 23 year old white Caucasian, who had the full clinical signs of Gorham-Stout syndrome. She had achieved 2 normal pregnancies and deliveries in spite of her massive pelvic osteolysis.*

Résumé. *L'ostéolyse massive est une lésion pseudo-tumorale extrêmement rare, qui atteint habituellement les adultes jeunes. Il en existe 132 cas publiés, qui ont été passés en revue, parmi lesquels 31 cas d'atteinte limitée au bassin. Le terme «d'ostéolyse massive» est basé sur les images radiologiques typiques, telles que l'augmentation de la transparence et la diminution de la densité de l'os. Le diagnostic de syndrome de Gorham-Stout doit être étayé par des constatations microscopiques concernant les structures vasculaires intra-médullaires. Notre malade est une européenne blanche de 23 ans qui présente le tableau complet du syndrome de Gorham-Stout. Elle a eu deux grossesses et deux accouchements normaux malgrè son ostéolyse massive du bassin.*

Introduction

The cause of primary idiopathic osteolysis is unknown. Osteoclasts usually resorb bone in association with tumours, osteomyelitis and biochemical disturbances. Trauma may also lead to localised bony destruction, for example at the lateral end of the clavicle [33] or in the pelvis [1]. Nerve lesions, alimentary disorders, microfractures or biochemical disorders are recognised as pathogenetic factors in osteolysis [29, 42].

So-called massive osteolysis is very rare and was first described by Jackson in 1838 [31]. Individual bones or localised bony regions may be totally decalcified and replaced by fibrous tissue [33, 36, 37, 42] leading to disappearance of the bone. In standard radiographs the affected parts of the skeleton can no longer be identified.

More than 30 years ago Gorham and Stout reviewed 24 cases published by several authors, and found that this type of disappearing or vanishing bone disease is associated with intraosseous vascular changes [18, 19, 20]. The Gorham-Stout syndrome came to be recognised as a definite entity to be differentiated from primary idiopathic osteolysis.

Since the first description, 132 cases of massive osteolysis have been published. In this paper we report a case of Gorham-Stout syndrome of the pelvis occurring during pregnancy.

Case report

In 1983 a healthy 23 year old woman complained of ill defined pelvic pain during her first pregnancy, which resolved after delivery. No further investigations were carried out.

In 1985 she developed a similar pain and radiographs showed osteolysis of the right pubis and ischium. Her symptoms were mild and she refused to have any other investigations. During her second pregnancy in 1986, insufficiency of the cervix was treated by cerclage and there were no other complications, although pelvic pain had increased significantly. She agreed to have further investigations in 1987.

She then had a normal gait, but she was unsteady when standing on one leg. There was marked weakness of the right

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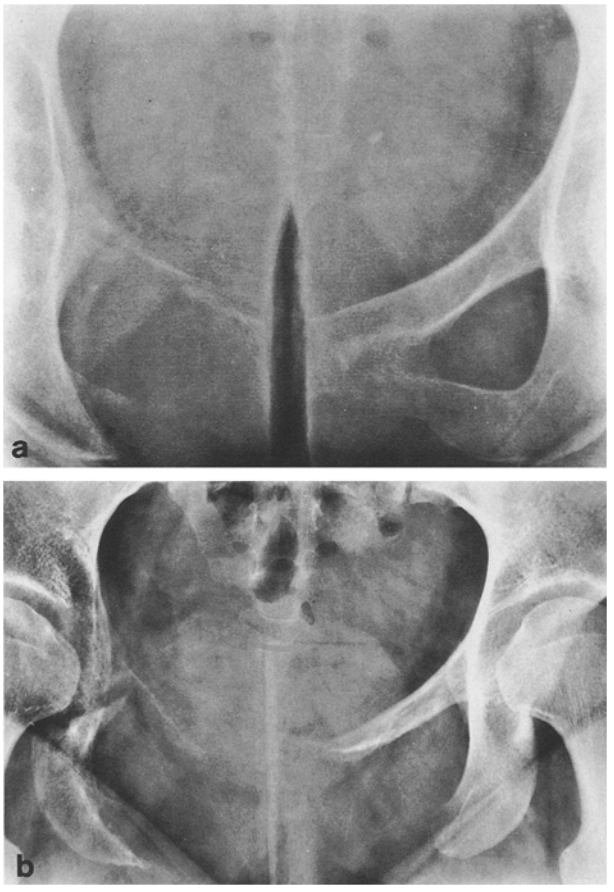

Fig. 1a,b. Radiographs of the patient in 1985 (**a**) and 1986 (**b**)

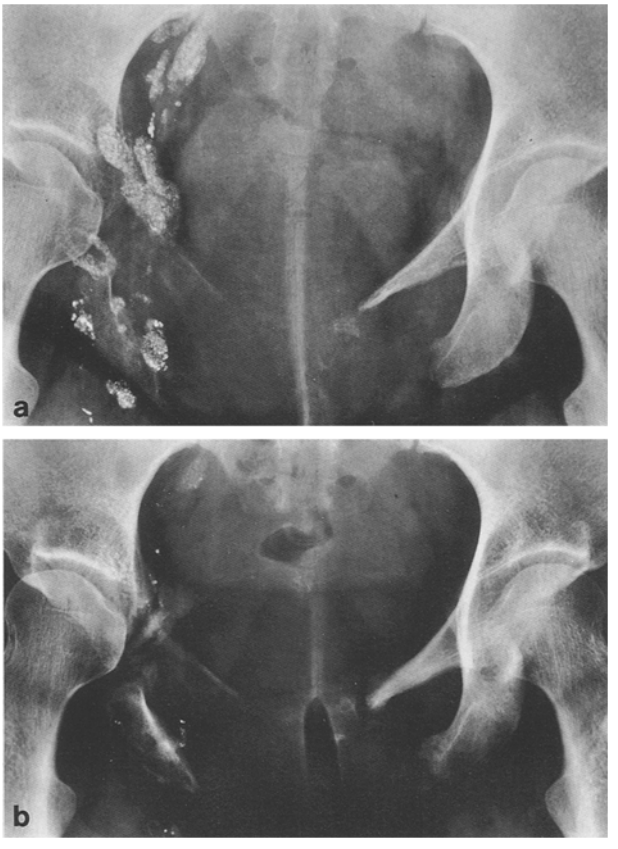

Fig. 2a,b. Pedal lymphangiography in 1987 before radiotherapy (**a**) and radiographs one year later (**b**)

thigh and she needed help with dressing. She was tender in the right groin and had pain on extremes of flexion, abduction and internal rotation of the right hip. The circumference of both thighs was the same, and there were no abnormal neurological or vascular signs.

Radiographs showed that osteolysis was present in 1985 (Fig. 1) with translucency in the right pubis and ischium. There was complete loss of density of the cancellous and cortical bone, and changes had occurred in the adjacent left side of the pelvis in 1986. By the following year, the changes had reached the right acetabulum without affecting the joint itself. Nuclear scanning was almost completely inconclusive as in a 3-phase study only late appositions revealed some increase in activity in the osteolytic area. Pelvic angiography, done as a non-selective and hyperselective study of the hypogastric artery, did not demonstrate any vascular abnormality. Pedal lymphography of the right leg showed unusual opacities in the lymphatic vessels in the area of osteolysis (Fig. 3a). A CT scan confirmed complete demineralisation; Houndsfield units in the area were consistent with fibrous tissue, and the surrounding soft tissues were completely normal. There were no signs of malignancy. CT guided intraosseous angiography demonstrated angiomatous structures within the affected bone (Fig. 3b). MRI studies showed that the lesion was benign; a significantly prolonged T_1 and T_2 relaxation time was consistent with surrounding soft tissue.

Open surgical biopsy was carried out at the junction between normal and osteolytic bone. Soft cancellous bone containing yellowish fluid was present. There was no bleeding from the marrow. The surrounding soft tissue, periosteum and cortical bone were macroscopically and microscopically normal. The central cancellous bone showed areas with wide vascular structures on microscopy, and these were associated with infiltration by lymphatic and plasma cells, and by fibrous tissue. There were signs of only minimal osteoclastic or osteoblastic activity.

A diagnosis of the Gorham-Stout syndrome was made, which was especially supported by the lymphangiomatous changes on microscopy and by the CT guided intraosseous angiography. The clinical course was also consistent with this diagnosis.

Fractionated irradiation of the hips and ischiopubic areas was carried out over 5 weeks in 1987. The bladder and intestine were shielded by lead and 5×2 Gy/week was applied with opposing AP/PA fields to a total dose of 40 Gy. Pain disappeared 4 to 5 weeks after the end of treatment and the affected area was no longer tender. She was able to do housework and did not use crutches. There had been no changes in the laboratory tests (blood count, electrolytes, alkaline phosphatase and LDH) during radiotherapy or in the following weeks. Quantitative nuclear bone scanning showed that the quotient of activity in the ischiopubic region and in the uninvolved areas showed a decrease in metabolic activity. A sign of improvement was the appearance of faint radio-isotope apposition in the lateral osteolytic area, more on the left side than on the right.

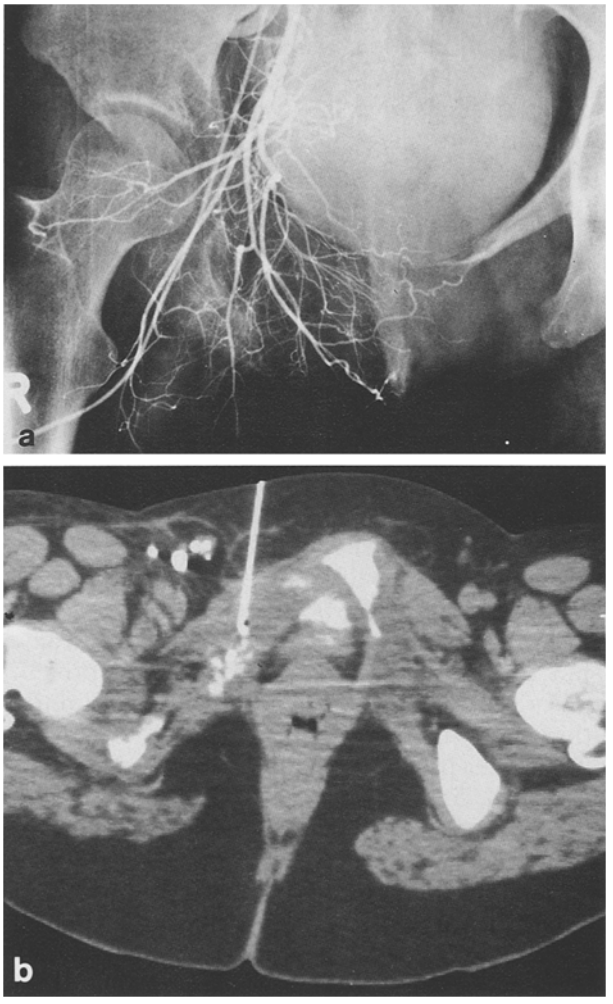

Fig. 3a, b. Pelvic angiography shows no evidence of the kind vascular changes seen in bone tumours (**a**). CT guided instillation of contrast medium demonstrates angiomatous structures within the osteolytic bone (**b**)

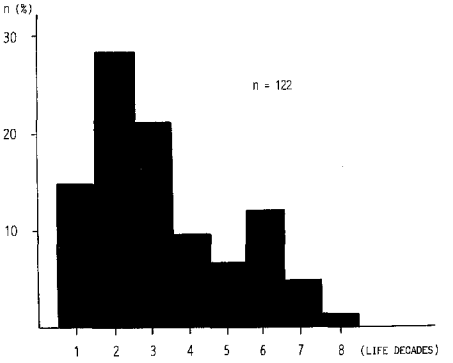

Fig. 4. Histogram showing the age distribution of 122 patients reported in the literature (given in %)

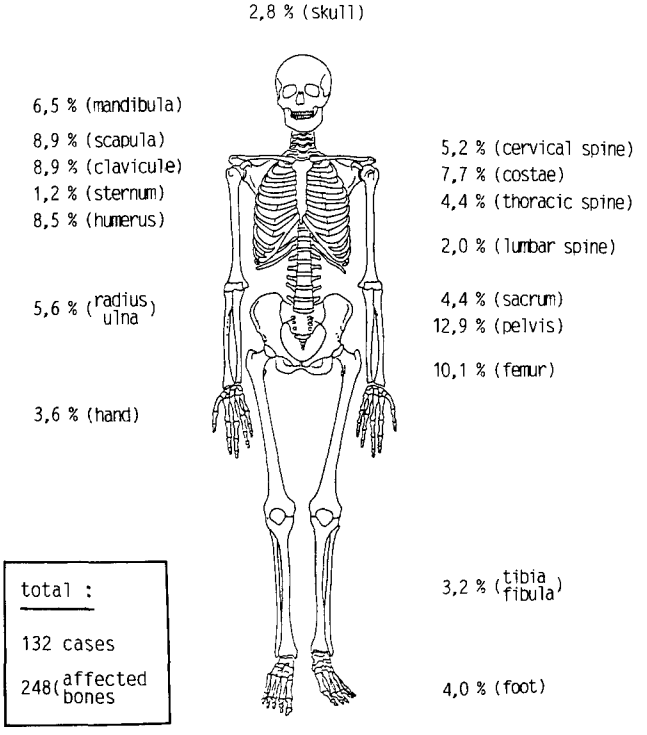

Fig. 5. Diagram showing the distribution of the skeletal lesions in 132 cases in the literature

Discussion

There have been more than 100 single case reports of massive osteolysis, and some reviews have added more cases [5, 11, 18, 19, 30, 63, 72]. Most publications come from Europe and North America, but there are some from Africa, Israel, India and Hawaii [10, 12, 14, 16, 43]. Various synonyms are used to describe the condition such as massive osteolysis [36], vanishing bones [68], disappearing bone disease [1], phantom and disappearing bone [11], the Gorham-Stout syndrome [21], and Jackson-Gorham disease [58].

Massive osteolysis is most frequently seen in young adults (Fig. 4) [10, 11, 23, 26, 37, 71] with slightly more males than females (55:45). Most often the condition is restricted to a single area, but multifocal cases [17, 54] and a malignant metastatic course [21] have been described. The most common bony areas affected are the pelvis and the shoulder, but any part can be involved (Fig. 5) [45]. We found 31 patients with massive osteolysis of the pelvis in the literature [2, 3, 4, 15, 23, 24, 25, 26, 27, 28, 34, 35, 38, 39, 40, 49, 51, 54, 56, 58, 59, 60, 62, 68, 71, 72, 73, 74]. The bilateral involvement in our case represents a different feature and 3 comparable cases have been published [25, 34, 35]. Our patient became pregnant, and the increased pelvic pressure from the growing child may have influenced the course of the condition

and could have been responsible for the spread to the other side of the pelvis. Mechanisms in the pathogenesis of massive osteolysis include trauma, osteomyelitis, hormone disturbances, auto-immunological changes and even congenital vascular defects [7, 17, 19, 20, 24, 37, 52, 53, 74].

Typical symptoms such as loss of strength, increasing pain and walking difficulties, are surprisingly mild compared to the changes seen in radiographs. Most patients do not complain of severe symptoms and a pathological fracture may lead to the diagnosis [7, 34, 65]. There may be pallor of the skin and swelling of the affected area [36, 39, 58, 65, 66], shortening of a leg, bowing of an extremity or scoliosis [3, 20, 23, 30, 31, 32, 36, 42, 72]. Joint function is almost always normal [3, 7, 20, 37, 39]. Neurological abnormalities are rare, but may occur when there is vertebral osteolysis [13, 15, 23, 42, 71, 75]. When the chest is affected there may be recurrent pleural effusions and diminished lung function [13, 23, 37, 46, 75]; a chylothorax has been reported [26].

In most cases laboratory findings are normal [4, 23, 39, 46, 65], but occasionally the sedimentation rate is raised and eosinophil granulocytes are increased. Alkaline phosphatase and the $\alpha 2$-globulin fraction may also be higher than normal [30, 36, 42, 63, 70].

The diagnosis is possible from standard radiographs provided other causes of osteolysis, such as tumours or osteomyelitis, are excluded. In the early stages, monostotic intramedullary translucencies, of variable size and with illdefined borders, are present [39, 66, 72]. No reactive bony changes are seen [36, 39, 66, 72]. Later, the affected bony region appears to be "rubbed out" [67]. Changes at the borders lead to a destructive pattern due to additional periosteal resorption [34, 36, 72, 74] and this was found in our case.

Standard angiography and lymphangiography usually fail to show the intraosseous angiectasis which occur in the Gorham-Stout syndrome [28, 36, 45, 66] and the intravascular changes seen in massive osteolysis do not seem to have any major communications with extraosseous vessels. Intraosseous angiography under fluoroscopy or CT control [28, 36, 41] is an important diagnostic technique which demonstrated the nature of the disease in our case. Pedal lymphangiography showed only a small focus with unusual lymphatic vessels, which was in contrast to the large amount of bone loss.

Massive osteolysis is a radiological and morphological description, but to diagnose the Gorham-Stout syndrome there must be microscopic evidence of intraosseous angiomatous changes which involve blood vessels more often than lymph vessels, although mixed types occur. The abnormal vessels lie in areas of lymphocytic infiltration. Eosinophilic granulocytes may also be present which suggest that there might be an immunological basis for the condition [6, 23, 30, 34, 36]. In the late stages the bone is almost completely replaced by fibrous scar tissue. The angiomatous changes are no longer present, so the diagnosis may become difficult [14, 36, 42]. Pathogenesis includes pressure-induced erosion by ectatic vessels and local disturbances of bone biochemistry, such as changes in pH [15, 19]. Other findings are increased perivascular resorption by mononuclear osteoclasts [5, 28], pathological abnormalities of the osteoblasts [4, 48], or modified rheumatic bone destruction [47, 48, 74]. Neoplastic characteristics justify classification of the condition as a tumour-like bone lesion [44].

The treatment of the Gorham-Stout syndrome is not well established and none of the present regimes are always successful [7, 45]. Calcitonin, mithramycin and diphosphonates have been suggested; other previously tried drugs have not been effective [4, 5, 7, 57, 70]. Surgical resection with the implantation of an endoprosthesis is more effective, but homogenous bone grafting often failed [7, 9, 23, 30, 45, 61, 65, 76]. Irradiation seems attractive since it sometimes stimulates reossification [36, 37, 41]. The dosage is under discussion and it is not clear whether between 6.5 and 68 Gy will induce beneficial obliterative endarteritis [28, 34]. There does appear to be an unknown intrinsic mechanism producing self-healing. If radical treatment is not feasible, conservative management using orthoses or other similar devices is justified as spontaneous remission can occur [8, 24, 25, 26, 34, 36, 44, 45, 53, 62].

Although complete re-ossification, with or without treatment, is uncommon the scarring process may improve stability in the later stages. Life span is rarely limited and only associated with complications secondary to thoracic or vertebral involvement [20, 23, 37, 55].

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