IDIOPATHIC MULTICENTRIC OSTEOLYSIS WITH ACRO-OSTEOLYSIS

A CASE REPORT

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We report a case of multicentric massive osteolysis. A 52-year-old woman presented with a three-year history of progressive deformities of the hands. She had osteolytic lesions of the metacarpals and metatarsals, and resorption of the terminal phalanges.

During follow-up over four years osteolysis spread to affect the ribs, clavicles, mandible, and long bones. There was no family history of any bone disorder and renal function was normal. Death resulted from resorption of the rib cage and post-mortem studies failed to reveal the cause of the osteolysis.

Massive osteolysis was first described nearly 150 years ago by Jackson (1838) but Gorham and Stout in 1955 were able to find only 24 case reports in the literature. In all these cases osteolysis occurred in one bone or in contiguous bones, and angiomatous tissue was noted histologically in many of the cases. Often there was a history of trauma. No biochemical abnormalities were reported and in no case was there a family history of a similar condition.

Other cases were subsequently reported in which the osteolysis was multicentric and in which there was a family history of similar complaints (Thieffry and Sorrell-Dejerine 1958) or an associated renal disorder (Torg and Steel 1968).

A simple classification proposed by Tyler and Rosenbaum (1976) divided idiopathic massive osteolysis into "unicentric", which included all Gorham's cases, and "multicentric" groups. The multicentric cases were further divided into multicentric osteolysis with nephropathy, and hereditary multicentric osteolysis.

Lysis of the terminal phalanges, termed acro-osteolysis, is associated with a number of conditions, including exposure to vinyl chloride, scleroderma, psoriasis and hyperparathyroidism. It was first described in association with multicentric osteolysis by Gilula, Bliznak and Staple (1976) and a further case was reported by De Smet (1980). Both cases were sporadic and neither showed any evidence of renal disease.

We report a third case of acro-osteolysis and multicentric osteolysis, which was unusual in its late age of onset and its relentlessly progressive course to death.

CASE REPORT

A white woman aged 52 years presented with a three-year history of gradually progressive deformities of the hands. She had otherwise been well and had worked as a packer of cosmetics. She had flexion contractures of the fingers with ulnar deviation of the metacarpophalangeal joints, but no soft-tissue swelling or tenderness. Her full blood count and erythrocyte sedimentation rate were normal and neither rheumatoid factor nor antinuclear factor was detected. Radiographs of her hands revealed large erosions of the periarticular cortex of the metacarpophalangeal joints, with some subluxation (Fig. 1). The terminal phalanges of the thumbs were eroded. Radiographs of her feet showed large periarticular erosions at the interphalangeal joints of the great toes, with resorption of the terminal phalanges (Fig. 2).

The initial diagnosis was of probable seronegative rheumatoid arthritis and she was given ibuprofen and occupational therapy. The function of her hands continued to deteriorate, and two years later she had operations for Silastic replacement of the metacarpophalangeal joints of the fingers of the right hand. A chest radiograph at this stage showed some lysis of several ribs (Fig. 3). The operation did not help and when next seen two years later, at the age of 56, she complained that her hands were useless, her back was painful and she was short of breath on exertion.

On examination. The patient's hands were grossly deformed, with flexion deformities of all of the interphalangeal joints and boneless thumbs (Fig. 4). Micrognathia and atrophy of the temporalis muscle were also noted (Fig. 5). She had marked kyphosis of the dorsal
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Figure 1—Radiographs of the hands at first presentation showing periarticular erosions at the metacarpophalangeal joints and resorption of terminal phalanges. Figure 2—Radiographs of the feet showing erosions of the hallux interphalangeal joints and resorption of terminal phalanges.

Figure 3—Chest radiograph showing lytic lesions of ribs 7, 8 and 9 on the right, and rib 7 on the left. Figures 4 and 5—Photographs of the hands showing gross deformity, and of the face to show micrognathia caused by resorption of the superior rami of the mandible.

Figure 6—Lateral radiograph of thoracic spine showing the gross kyphosis; the spinous processes tapered into sharp points. Remarkably, no vertebral body throughout the spine showed collapse or even significant wedging. Figure 7—Radiograph of the chest showing lytic lesions of the left clavicle and of many ribs, especially on the right. Figure 8—Radiograph of both knees showing large periarticular erosions at the lower ends of the femora and the superior pole of the right patella. The left knee is subluxated.

spine (Fig. 6) with a large bursa overlying the apex of the curve, within which the sharp end of a partially eroded spinous process could be felt. Her chest expansion was very limited, and radiography confirmed further lysis of the ribs, with developing lysis of the left clavicle (Fig. 7). Investigations. These gave the following results: haemoglobin 13.1 g/dl; erythrocyte sedimentation rate (Westergren) 5 mm in the first hour; liver function tests, alkaline phosphatase, calcium phosphate, urea, 24-hour urinary calcium and hydroxyproline all within normal limits.
Tests for rheumatoid and for antinuclear factor were both negative. Protein electrophoresis revealed a discrete band in the gamma region, but immunoglobulin levels were within normal limits and no Bence-Jones protein was detected in the urine. Vitamin D and vitamin C assays showed no deficiency of either. Thyroxine, parathormone and cortisol levels were normal, but oestriadiol was slightly high at 2.9 nmol/l (normal 0.5–2.1). All these results were felt to be within appropriate ranges for a menopausal woman. As indicated above, her respiratory function was limited, forced vital capacity being only 0.45 litres and FEV₁ 0.4 litres.

Management. Since bone resorption might be caused by osteoclastic overactivity, a trial of calcitonin by injection was made. The patient could not tolerate a daily dose of 100 iu, and no improvement was found on 50 iu daily, reducing to 50 iu three times weekly for three months. Her condition gradually deteriorated. Further osteolysis occurred, affecting mainly the ribs and the knees, where large lytic lesions developed at the site of some ligament and tendon insertions. Eventually the left knee dislocated posteriorly (Fig. 8).

The patient died at the age of 57, of cardiorespiratory failure caused by lysis of most of the ribs.

Post-mortem examination. This revealed that the bones of many parts of the body had disappeared, leaving only soft tissue replacing them. The fourth to the eighth ribs on both sides had largely disappeared, the mandible was severely affected, and whole phalanges, metacarpals and metatarsals had vanished.

The synovium and articular surfaces of all of several joints which were examined were normal. Histological examination of the soft tissue which had replaced the vanished bone revealed no excessive or abnormal vasculature. No occult malignancy was revealed by careful examination of the organs, and iliac crest biopsy showed normal bone histology. The distal end of the left clavicle had shown partial resorption on radiographs, but there was no abnormality of the histology of the remaining bone, though this ended abruptly at some distance from the acromioclavicular joint. The soft tissue surrounding this was also normal.

DISCUSSION

This case presents several very unusual features. It does not fit into either half of the classification of Tyler and Rosenbaum (1976) as there was no positive family history and no evidence of renal disease. It may be that our patient’s condition was caused by a spontaneous mutation. Such sporadic cases have been reported (Beals and Bird 1975; Amin and Evans 1978), but the onset of symptoms in their cases has usually been in infancy.

Our patient noticed the first skeletal abnormality at the age of 49 years and died as a direct result of gross osteolysis only eight years later at the age of 57. Such widespread osteolysis, progressing inexorably and directly causing death, has not previously been described.

REFERENCES