MASSIVE OSTEOLYSIS—DISAPPEARING BONES

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This is the record of a case in which there was complete disappearance of the left scapula and the outer third of the clavicle and partial absorption of several ribs. Death finally occurred from chylothorax due to obstruction of the thoracic duct by fibrous tissue.

Twenty-six such cases have been recorded before, but only one in England (Jackman 1939). Opportunities for complete pathological study have been rare, although a fair amount of biopsy material has been available. From the literature it appears that one other patient died as a direct result of the disease, and in that case also chylothorax occurred after disappearance of the right clavicle, most of the scapula and several ribs (Gorham, Wright, Shultz and Maxon 1954). These authors studied two cases of their own and sixteen others from the literature.

The condition was the subject of a further review, with full references, by Gorham and Stout in 1955, when the total number of cases tabulated was twenty-four. Gorham and Stout's account is particularly important because the authors studied the original sections from a total of eight patients and concluded that on histological grounds there was a common, basic pathological lesion. Different authors have described the histological changes as "connective tissue and capillaries," "fibrous tissue rich in blood vessels," "marrow replaced by fibrous vascular tissue," "resembles fibrocystic disease" and "abundant vascular channels." Gorham and Stout found a convincing similarity in the histological pictures seen in this condition, and stated that progressive osteolysis was always associated with angiomatosis of blood vessels, and sometimes of lymphatics, which seemingly was responsible for it. They were unable to state whether massive osteolysis occurred as the direct result of active hyperaemia, of mechanical causes, of slight changes in pH, or from some other cause. Since Gorham and Stout's report Krikler (1955) and Marie *et al.* (1956) have each added a further example.

It is clear that massive osteolysis or disappearing bone disease is quite distinct from atrophy after injury or from disuse. The osteolysis progresses until the affected bones are represented by fibrous tissue. It is unnecessary to repeat the details given in the full reviews mentioned above, but a few general points may be considered. The condition is most common in adolescents and young adults, and there is often a history of minor injury. The progress of the bone resorption is gradual, and is often accompanied by surprisingly little disturbance of function. Constitutional disturbance is usually slight, and there has been no significant alteration in the blood calcium, phosphorus, phosphatase or proteins when these have been recorded. Sometimes the disease has undergone spontaneous arrest: certainly there is no known effective treatment. It is not accompanied by any definite neurological abnormality.

CASE REPORT

The patient was a man born in 1928. The first record of trouble with the left shoulder was at the age of twenty-three, when he was admitted to a military hospital. The documents state that he was swinging three floor bumpers the previous day when the left arm gave way. It is also noted that three years previously he had "put out the left clavicle and went to a bone setter who put it back. Since then the left shoulder has always been weak." According to the notes, examination showed marked dropping of the left shoulder, the lower angle of the scapula being an inch lower on the left than on the right. There was marked wasting

of the supraspinatus and infraspinatus muscles and slight wasting of the deltoid. Marked wasting of the upper arm was noted. He was unable to raise the arm from the side or to maintain the passively abducted arm. There was no sensory loss, but he complained of acute tenderness over the scapula just below the spine. Radiographs are reported to have shown an apparent crack in the scapula about two inches below the neck. Unfortunately the original radiographs have been destroyed and only the reports remain. A report made one month after his first attendance noted an old united fracture of the left clavicle. A further report a year later indicated that the fracture of the neck of the scapula had healed. He was still having difficulty in raising the arm but there were no sensory changes; tenderness over the scapula persisted.

Records do not show when he left the Army, but the remark "deserted later" appears. It is recorded that he was at work in a quarry as a labourer for a year or more before he was

FIG. 1

Radiograph showing the large effusion in the left pleural cavity. The attenuated end of the left clavicle is also shown.

first seen, at the age of twenty-eight, by one of us (R. L. M.). He had noticed loss of weight for two years. He sought advice on account of pain in the left chest with shortness of breath and swelling under the left arm. He stated that the left shoulder had never been strong since onset of the trouble five years before, though he had been able to work quite well with it.

On his admission to Hawkmoor Chest Hospital he had marked dyspnoea, with a large chylous effusion in the left side of the chest (Fig. 1). There was a large fluctuant swelling in the left scapular area which could be compressed into the chest by pressure from the hand. There was considerable wasting of the left deltoid and spinati muscles; he was unable to raise the arm, though a limited range of passive movement was possible. There were no abnormal neurological signs. Chest aspirations were performed on twenty-six occasions on the left side during the ensuing month, a total of 45.6 litres of milky fluid being withdrawn. Towards the end of the month fluid began to accumulate in the right pleural cavity and seemed to diminish in the left. In the succeeding week three aspirations of the left pleural cavity

yielded 2·3 litres and three of the right pleural cavity 4·3 litres. Thus about 52·2 litres of chylous fluid were lost to the patient in forty days. This is equivalent to 116 lb., which was very nearly his body weight on admission to hospital.

FIG. 2

Radiograph taken after removal of chylous fluid from the left pleural cavity. There is a faint shadow in the region of the glenoid cavity. The attenuation of the clavicle is well shown. There is nothing to indicate the position of the scapula.

FIG. 3

Radiograph showing the erosion of the ribs.

When the fluid had been removed it became clear that he had no left scapula, and on palpation there was nothing to indicate where it should have been. Radiographs showed only a faint shadow in the position of the glenoid cavity and no evidence of any other part of

the bone. The outer half of the left clavicle had also largely disappeared (Fig. 2). There was erosion of the ribs in the position where the scapula should have been, and it was here that the communication existed between the chest wall and the pleural cavity (Fig. 3).

Investigations—Biopsy was done on the outer end of the left clavicle, and the histology of this is considered later. The pleural fluid was examined on several occasions for tubercle bacilli both culturally and biologically, with negative results. *Blood count* showed: haemoglobin 107 per cent; white blood cells 7,400 per cubic millimetre (polymorphonuclears 87 per cent, eosinophils 4 per cent, lymphocytes 8 per cent, monocytes 1 per cent). *Urine*—Bence Jones protein was not present. *Serum proteins*—Albumin 2.22 grammes per cent; alpha 1 globulin 0.35 gramme per cent; alpha 2 globulin 1.03 grammes per cent; beta globulin 0.83 gramme

FIG. 4

The head of the left humerus and the glenoid cavity. The glenoid cavity consisted of fibrous tissue and a little cartilage; outside this was some fibrous tissue but no evidence of a scapula.

per cent; gamma globulin 5.20 grammes per cent. *Fluid from the chest* showed large numbers of mononuclear cells but no acid-fast bacilli. Protein was 2.62 grammes per cent, and fat 1.105 grammes per cent. Fat globules were present after extraction with ether. The Wassermann and Kahn reactions were negative.

The patient died from cachexia seven weeks after his admission to hospital.

Post-mortem findings—The body was that of a greatly wasted man.

Skeletal abnormalities—The outer third of the left clavicle was missing, and in the fibrous tissue around the end of the clavicle there were two hard nodules probably containing bone. The part of the clavicle nearest this outer third was thin and tapered almost to a point. The shoulder girdle was dissected but no trace of the left scapula could be found, nor was there an excess of fibrous tissue in the position where the scapula might have been. A bunch of muscle was present but this did not appear to contain any bone or calcareous elements.

FIG. 5

Three of the left ribs. They were matted together at their angles: the bone was thinned and easily fractured.

FIG. 6

Tissue from the angle of an affected rib. Bone trabeculae separated by large amount of interstitial tissue containing large capillary channels. (H. and E., $\times 40$.)

FIG. 7

Periosteum of rib and underlying fibrous tissue, showing large numbers of dilated capillaries. (H. and E., $\times 40$.)

FIG. 8

Fibrous tissue from around the outer third of the clavicle, showing large anastomosing vascular spaces. (H. and E., $\times 80$.)

FIG. 9

Tissue from around the outer third of the clavicle. Plasma cells and eosinophils present in focal collections. (H. and E., $\times 320$.)

It was difficult to imagine that there had been a scapula in this position and that it had disappeared entirely. The uppermost third of the left humerus was removed for further examination (Fig. 4). The head was well formed and of normal size, but on its inferior surface there was a considerable amount of erosion, leaving a rough corrugated surface. The capsule of the joint contained a little cartilage on the glenoid side but no bone. The ribs on the left side appeared to be normal anteriorly and laterally, but at their angles they were matted together. Three which were removed for examination showed that the bony portion of the rib became gradually thinner towards the neck of the bone (Fig. 5), and they were very easily fractured. When the anterior parts of the bodies of the vertebrae were removed the bony structure was normal.

Heart—The heart weighed 180 grammes. Though small, it was in keeping with the general physique. There was no valvular disease and the coronary vessels were healthy. The pericardial sac contained about four ounces of turbid, milky fluid. There was no excess of fibrin on the surface of the heart.

Lungs and pleural cavities—The right pleural cavity was full of turbid, milky fluid of chylous type and the lung was compressed. There was no evidence of neoplasm and the ribs appeared to be normal. On the left side, at the entrance to the chest cavity, surrounding the subclavian vessels, there was a mass of dense fibrous tissue which may well have obstructed the thoracic duct. This was removed for further examination. The lung was completely collapsed against the vertebral column, dark in colour and semi-solid, but there was nothing to suggest neoplasm. The chest cavity contained no fluid but did contain a large mass of fibrous material which was adherent both to the ribs and to the surface of the lung.

Other organs—The liver was normal in size and showed slight congestion. The spleen was about one and a half times the normal size: the cut surface appeared normal. The adrenals were normal. The kidneys showed no naked-eye evidence of disease. The alimentary tract showed no abnormality. The brain was not examined.

Histology—The tissues examined histologically included: 1) material obtained at operation from the outer third of the left clavicle; and 2) the remainder of the left clavicle, parts of ribs and the fibrous tissue surrounding the left subclavian vessels taken at necropsy.

The various bone specimens each showed a similar structure (Figs. 6 to 9). There was evidence of osteoclasts, the bone being replaced by fibrous tissue. The periosteum was thickened, and on its deeper surface it merged into fibrous tissue in which were many dilated capillaries. Similar large capillaries were present in the interstices of the bone. The degree of this vascular change was not even throughout the material. Evidence of new bone formation was scanty.

The material from around the subclavian vessels consisted of fibrous tissue containing large nerve bundles, sympathetic nerve ganglia and several large arteries and veins, some of which showed various stages of thrombosis and recanalisation. An interesting feature of this tissue was the presence of numbers of eosinophils and plasma cells in large groups, sometimes with a perivascular pattern. This feature was not found in the bone lesions. Small capillaries were not so prominent in this tissue, but large anastomosing vascular spaces, like those described by Gorham and Stout (1955), were much in evidence.

DISCUSSION

These clinical and pathological features appear to us to place this case in the rare group which shows massive osteolysis. The outer third of the clavicle and the whole of the scapula had disappeared, and portions of several ribs were in process of vanishing.

In particular, comparison of the histological findings with those described and fully illustrated by Gorham and Stout, in their study of eight examples of this condition, leaves no doubt that we are dealing with one of the very few cases in which the disease itself led to the death of the patient. The presence of eosinophils in the material from near the site of the clavicle raised the possibility of an eosinophilic granuloma, but further study of the vascular pattern, and especially of the reaction in the bones undergoing absorption, made it appear that this chronic inflammatory reaction was not the essential part of the disease process.

We do not feel able to suggest the etiological factor except to agree with Gorham and Stout that the osteolysis seems to be related to the haemangiomatous condition and may be a direct result of the consequent hyperaemia.

Injury was mentioned in the history of this patient, but it does not appear to us to be the cause of the condition. One can imagine an injury to the shoulder initiating a destructive lesion in the clavicle or the glenoid cavity, but complete disappearance of the scapula from this cause is most unlikely. Injury certainly cannot be the explanation of the osteolytic changes in the ribs. Trophic nerve lesions are not known to cause multiple foci of bone destruction of this magnitude.

SUMMARY

1. The clinical and pathological features are given of an example of the rare syndrome of progressive and massive osteolysis associated with haemangiomatous changes in the bones affected.
2. Twenty-six cases have been previously reported, but this is only the second from Britain. Only one other patient is considered to have died as a direct result of the disease.

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