AN UNUSUAL COMPLICATION OF PAGET'S DISEASE

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SUMMARY: A patient with symptomless Paget's disease of the bone, suddenly developed multiple osteogenic sarcoma. Obstruction to one jugular vein led to increased intracranial tension and a perforation developing in the orbital plate of the frontal bone led to uniocular symptoms and signs. With this, we believe hitherto unrecorded complication, he also exhibited some of the well known complications of Paget's disease of the bones.

Introduction

Paget's disease in all its forms has been found to occur in about 1 in 30 persons over the age of 40 (Collins, 1956). The vast majority of such cases are subclinical being detected only when specifically looked for at post mortem. Even in the clinically apparent cases the course is usually insidious and although disabling bony deformities may ensue, the disease itself rarely causes death. A small number of patients do die of complications which can be specifically related to Paget's disease. The commonest lethal complication is the supervention of an osteogenic sarcoma. There are wide discrepancies in the reported incidence of this complication but two recent series (Price, 1962, Porretta, Dahlin and Janes, 1957) give the frequency rates as 0.15 and 0.9 per cent respectively. The lower of these two figures represents a thirty fold increase in the risk of sarcoma in persons aged over forty, compared with the normal. Other lethal complications of Paget's disease include a high output cardiac failure, and renal complications of hypercalcaemia and hypercalciuria when such patients are immobilised, as for example after a pathological fracture.

Case Report

A publican aged 68 years was admitted to a Royal Air Force Hospital in October 1969 with a 10 days history of frontal headache. This was constant, day and night, and bore no relation to posture, coughing or straining; there was no nausea or vomiting and simple analgesics gave no relief. Two days prior to admission the left eye had become swollen and red, but there were no subjective visual symptoms. A few days earlier he had been seen in an orthopaedic out-patients department for a swollen painful knee. A large bowel carcinoma had been surgically removed in 1967.

The most striking aspect on initial confrontation was an enlarged head with a circumference of 62 cm. There were numerous bony "bosses" scattered over the surface of the skull and there was a marked frontal overhang. He admitted to having found difficulty in obtaining a hat to fit his head for at least the previous 15 years. The left tibia was enlarged and deformed with some bowing and the overlying skin was warmer than on the other side. The right knee was enlarged and the appearance superficially resembled osteoarthritis, but there was a full range of pain free movement.

The cardiac apex beat was palpable in the 6th left intercostal space on the anterior axillary line and had the quality of a sustained heave. B.P. was 160/100 mm of Hg. There were no signs of cardiac decompensation.

The left eye exhibited peri-orbital and conjunctival oedema and the left ocular fundus papilloedema without haemorrhages or exudates. The corrected vision was Right 6/6 and Left 6/9.

A lumbar puncture produced clear c.s.f. under a raised pressure of 280 mm of c.s.f.; compression of the right jugular vein produced the usual Queckenstedt's response, but there was no response to compression of the left jugular vein. The biochemistry and cytology of the c.s.f. were within normal limits.

Radiographs of the skull showed the typical cotton-wool appearance of Paget's disease, with the thickness of the skull of up to two cm in places. Other bones involved radiologically were the left tibia and femur, the pelvis and the lower end of the right femur, which showed a rather disorganised periosteal reaction suggestive of osteogenic sarcoma; the right knee joint showed none of the features of osteoarthritis.

Serum alkaline phosphatase level was 640 K.A. units, the highest ever recorded in this hospital's laboratory.

A radiograph of the chest confirmed an enlarged heart. Electrocardiogram showed a moderate left ventricular hypertrophy. Electroencephalogram showed changes consistent with raised intracranial pressure, with occasional delta waves seen mainly in the frontal area suggesting a left frontal space occupying lesion.

Shortly after admission to hospital, he started bleeding from the left nostril and this became increasingly difficult to control. Examination by an otorhinolaryngologist showed the source of bleeding to be an infected, friable, intensely vascular area in the left ethmoidal region. An incidental finding was a combined conductive and sensorineural deafness on the left side. Biopsy taken from the ethmoid area was technically unsatisfactory; biopsy taken from the lower end of the right femur showed "Paget's disease and considerable necrosis but no malignancy".

At this stage he was transferred to a regional neuro-surgical unit for a possible left orbital decompression, but the neurosurgeon considered any surgical interference to be undesirable and the patient was discharged home.

Five weeks later, in December 1969, he was readmitted under our care having deteriorated rapidly. The head circumference had increased from 62 cm to 64.5 cm, and the bony bosses in the skull were much more prominent. He was almost blind in the left eye. The left periorbital oedema was gross and masked a proptosed left eye. There was slight weakness of the left half of the face of the lower motor neurone variety. The enlarged right knee joint had increased further in size being extremely painful and tender. There was no further epistaxis, but there was a most foul smelling nasal discharge, to which he was mercifully oblivious. He developed an intercurrent chest infection and died three days after re-admission.

At post mortem, as the skull was opened, the cerebrospinal fluid spurted out under pressure; the bony bosses palpated externally were noted to be equally widespread on the inner surface of the cranial bones. There were necrotic areas in the frontal bones, left petrous temporal bone, the sphenoid and ethmoid bones; these were considered possibly sarcomatous and specimens were taken for histological examination. In between these areas, the cranial bones showed the typical, spongy appearance of Paget's disease. Over the sarcomatous looking areas of the cranium, the dura mater was firmly attached. The most striking finding was a punched out hole, 1.25 x 1.25 cm in dimension, in a

very necrotic area in the orbital plate of the frontal bone. This provided a direct communication between the intracranial cavity and the back of the left orbit. The optic nerves and the nerves arising from the mid-brain were encased in a necrotic mass of tissue anterior and inferior to the pituitary fossa. Section of the right femur, 7.5 cm above the knee joint, showed spongy, expanded, necrotic bone with extension through the periosteum into the overlying muscles; a specimen from this area was also taken for histology.

Histological examination confirmed Paget's disease of the bones with osteogenic sarcomatous changes in several separate areas of the skull and lower end of the right femur.

Discussion

The dual peak in the prevalence of osteogenic sarcoma is well documented; the first peak occurring in the second decade with a predilection for the metaphyses and the second peak occurring in the sixth decade with a predilection for areas of bone exhibiting Paget's disease. Coley and Sharp (1931) in a series of 71 cases of osteogenic sarcoma found 28 per cent to be associated with Paget's disease. In all their cases of osteogenic sarcoma of the skull there was a pre-existing Paget's disease. Porretta, Dahlin and Janes (1957) in a review of 128 cases of skeletal sarcoma associated with Paget's disease found 20 per cent to be in the skull. Of these 128 cases there was only one reported five-year survival.

Our patient presented with symptoms referable to multiple sarcomatous changes occurring simultaneously in several areas of the skull and in the lower end of the right femur. From being symptom free he was dead within 2 months. Although sarcomatous change has been observed in more than one situation in Paget's disease, its supervention is usually monomelic (Price, 1962). Its simultaneous development at multiple sites in a patient with long standing Paget's disease raises the theoretical possibilities of some subtle but generalised change in the osteocytes or of a humoral mediator.

The sudden and severe headache with which this patient presented may be explained by a combination of raised intracranial pressure and local, sarcomatous changes in the cranial bones. An explanation of the raised intracranial pressure offered is compression of the left jugular vein by bony changes around the left jugular foramen. This was clinically supported by the unilaterally negative Queckenstedt's test. However, the cranial nerves emerging from the jugular foramen appeared intact. Localisation of the papilloedema to the left eye could be explained by the combination of the raised intracranial pressure with sarcomatous changes in the orbital plate of the left frontal bone. But the optic nerves were observed at post mortem to be enmeshed in necrotic tissue in the base of the skull and this could have been asymmetrical to start with.

However the most striking aspect of this case was the unexpectedly sudden downhill course with a very rapid deterioration in the vision of the left eye, accompanied by marked proptosis of the left eye. Unilateral proptosis is always suggestive of local pathological changes and had previously been mentioned in Paget's disease (Mackie, 1956; Stemmermann, 1955); but such cases were usually insidious. In our case it occurred acutely over the course of a few days. Post mortem examination showed this acute onset to be due to the c.s.f., under increased pressure, communicating directly with the left orbit via a hiatus through an area of osteogenic sarcoma in the orbital plate of the left frontal bone. A review of the literature reveals no previous record of such a complication being hitherto described in Paget's disease.

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