PULSATING ANGIO-ENDOTHELIOMA OF THE INNOMINATE BONE TREATED BY HINDQUARTER AMPUTATION

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The infrequency of pulsating angio-endothelioma of bone appears to merit the publication of an individual case in a short note rather than its inclusion in a list of thirty-three personal interinnomino-abdominal (hindquarter) amputations wherein the case might more readily be overlooked. An appeal to the classical work of Ewing (1942) vouchsafes the information that the growth of endotheliomata tends to be slow but progressive, and thus they are comparatively benign. More particularly, by comparison with the carcinomata and sarcomata which they may resemble histologically, the course of endotheliomata is relatively favourable. When undisturbed by the knife the growth capacity of some of these tumours seems to be confined within certain limits which do not apply to carcinoma and sarcoma.

It is said that endotheliomata spread by local infiltration, principally along existing channels such as lymph vessels and spaces, while extension to neighbouring lymph nodes, if it occurs at all, is late. Such infiltration accounts for the difficulty of thorough extirpation and for the frequency of persistent local recurrence, each recurrence being apt to show an increasing capacity for growth. This fact encourages the belief that the appropriate treatment is complete removal of the organ involved and not merely local extirpation of the tumour.

Endothelioma of bone tends to replace pre-existing tissues with little or no increase in the size of the bone, but with perforations. Three types of bone endothelioma were distinguished by Ewing: 1) solitary, bulky telangiectatic angio-endothelioma; 2) multiple endothelioma of bone; 3) diffuse endothelioma of bone. The solitary angio-endothelioma of bone, which is the type of tumour present in the patient now reported, affects adults and principally involves the long bones. It grows steadily, perforates and invades the soft tissues and may, as a late feature, metastasize to the lungs; it may pulsate and produce a bruit. Considerable variation in size may be observed; it is usually painful and sometimes causes pathological fracture. The tumour consists of masses of large cuboidal cells in cords or pseudoalveoli enclosing freely circulating blood.

Ewing also reminds us that a somewhat similar clinical and histological picture may be given by metastatic carcinoma of renal or adrenal origin, and that such a primary tumour must always be excluded in formulating the diagnosis.

Other pulsatile osteolytic tumours may occur in bone, such as telangiectatic osteogenic sarcoma and giant-cell tumour. Telangiectatic osteogenic sarcoma usually affects children and is characterised by exceedingly rapid growth and early pulmonary metastases. Giant-cell tumour of bone, when vascular and pulsatile, may cause great difficulty in differential diagnosis, its growth being slow like the angio-endothelioma; but usually the periosteum lays down a bony capsule which gives rise to characteristic radiographic appearances.

Case Report—E. B., female, 42 years of age, was admitted to hospital on April 15, 1949, with a history that five months earlier she had noticed pain in the right hip, rather posteriorly, which persisted for a month. The pain was a dull ache, with an occasional sharper element, gradually spreading down the front of the thigh to the knee and from there to the front of the shin and the toes. In the lower leg it tended to be a shooting pain, rather sharp like a knife. It came on particularly in the morning and was eased by rest. Lying on the right side caused discomfort in the limb which prevented her from getting to sleep, but did not awaken her when she was once asleep. The pain made her limp.

On examination—There was a mass in the right iliac fossa exhibiting marked expansile pulsation which could be obliterated by compression of the aorta. A to-and-fro murmur

could be heard over the tumour. The tumour presented on both surfaces of the iliac part of the os innominatum. The diagnosis seemed to lie between a tumour of the innominate bone and an iliac aneurysm, with the odds heavily on a bone tumour. The radiograph showed destruction of the wing of the ilium (Fig. 1).

There was nothing abnormal in the cardiovascular, respiratory or central nervous systems. The appetite had been indifferent but there was no other abnormality in the alimentary system. The urinary system was normal and menstruation had ceased for the last eight months. The patient was rather pale and thin; the skin, hair and nails were normal; thyroid not palpable; no superficial glands felt. Pulse rate 76; regular; volume and tension normal: blood pressure 160/85: heart sounds normal.

FIG. 1

Radiograph showing destruction of the ala of the right ilium by angio-endotheliomatous tumour. The pyelogram shows a defect in the ureter at the pelvi-rectal junction on the right side.

Blood count (February 1, 1949)—Haemoglobin 68 per cent.; red blood corpuscles 3.85μ ; colour index 0.88; P.C.V. 34; white blood corpuscles 3,600 (polymorphonuclears 63 per cent., lymphocytes 33 per cent., monocytes 4 per cent.) hypochromia; microcytosis; anisocytosis. Erythrocyte sedimentation rate 20 mm. in one hour.

Faeces (February 8, 1949)—Occult blood—Benzidine, faintly positive; Guaiacum, negative.

Radiographs of skull, ribs, sternum and vertebrae (February 8, 1949)—No evidence of myelomatosis in the ribs, skull, sternum or spine. The opacity in the pelvis was presumed to be a solitary plasma cell tumour.

Blood examination (February 11, 1949)—Total protein, 7.0 gms. per 100 ml.; albumen-globulin ratio, 2.3 : 1; fibrinogen, 0.38 gms. per 100 ml.

Red bone marrow examination (February 21, 1949)—Total nucleated cell count 67,000 c.mm.; myeloid/erythroid ratio 3 : 1; proerythroblast 2 per cent.; early normoblast 4; intermediate normoblast 7; late normoblast 17; megalokaryocyte 0.2 per cent.; myeloblast 1; premyelocyte 3; myelocyte 9; metamyelocyte 17; polymorphonuclear 33; lymphocyte 5 per cent.; monocyte 1 per cent.; plasma cell 0.3 per cent.; no myeloma cells seen.

Intravenous pyelography—Preliminary films—The tumour in the right ilium was considerably larger than on previous radiographic examination. Both kidneys excreted the dye at five minutes. The renal drainage system on the left side was normal. On the right side there was a

FIG. 2

FIG. 3

Specimen of angio-endothelioma of innominate bone showing relation to the iliac vessels and displacement of the ilio-psoas (Fig. 2) and the same specimen on section (Fig. 3).

constant defect in the ureter at the pelvi-rectal junction (Fig. 1). The pelvis and calyces on this side were otherwise normal. The nature of the defect was obscure and the lower part of the right ureter was displaced medially as if by the tumour. An impression was also present on the right side of the bladder. Retrograde pyelogram showed the same appearances as in the intravenous pyelogram. The narrowing at the pelvi-ureteric junction could be explained by extrinsic pressure, and the deviation of the lower end of the ureter by the mass. There did not appear to be any clear evidence of hypernephroma.

Biopsy of tumour of right ilium (March 17, 1949)—An incision was made half an inch below the anterior part of the iliac crest and parallel to it. There was considerable bleeding from the subcutaneous tissue and below the deep fascia; in the muscle the bleeding became so profuse that further approach from this direction was impossible. The iliac crest was exposed and the muscle stripped off it for three-quarters of an inch. Two pieces of bone were removed for biopsy: they were macroscopically normal. There was by this time so much bleeding from the bone as well as the other tissues that further progress was not practicable. Accordingly, the bleeding was controlled by Oxycel gauze swabs and the wound closed, leaving two of these inside. The tumour could be felt as a hard mass beneath the muscle, but it was not exposed or incised. Histological examination of the biopsy fragments showed normal bone tissue, no tumour tissue being present.

Radiograph of chest (March 17, 1949)—There was some cardiac enlargement and two opacities in the right lower thoracic zone. The appearances indicated old calcified tuberculous foci. No secondary deposits seen.

Operation (April 20, 1949)—The usual preliminary operative approach to a hindquarter amputation was employed, which also gave adequate approach to the iliac vessels. Owing to the extent of the tumour the whole innominate bone required removal; disarticulation at the sacro-iliac joint was necessary (Figs. 2-3). During the operation and subsequent post-operative

period six pints of blood were transfused. The patient made an uneventful recovery from the hindquarter amputation and left hospital, June 9, 1949, walking well on crutches.

Histological Report (April 20, 1949)—Tumour 4 cms. $\times$ 3 cms. $\times$ 3 cms. eroding the centre of the iliac bone. The mass appeared to have a fibrous capsule; it had not invaded but had displaced the iliacus muscle above and the gluteus below. On section, the tumour was composed of a firm outer rim three-quarters of an inch wide with a bony vascular centre:

FIG. 4

Microscopic section of haemangio-endothelioma of the innominate bone.

there were many spicules of bone in the tumour. Section showed a cellular tumour replacing the bone: the tumour showed numerous spaces containing blood, surrounded by several layers of endothelial cells. The degree of differentiation indicated a moderate degree of malignancy (Fig. 4).

REFERENCE

EWING, J. (1942): *Neoplastic Diseases*. Philadelphia and London: W. B. Saunders & Co., **342**, 360.