A CASE OF CONGENITAL ARTERIOVENOUS ANEURYSM INVOLVING THE FEMUR

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This rare condition is being described for the unintentionally dramatic way in which it was discovered and, in a sense, there is also a Cautionary Tale. If there is a moral to the story, it is this: Always put a stethoscope on a suspected bone tumour and look at the radiographs more than once.

So far as I know, there is no description of a non-traumatic arteriovenous aneurysm involving, or arising in, one of the long bones.

Congenital arteriovenous aneurysms are not uncommon and are fully described in the medical literature. Various authors have described different lines of treatment, but basically it consists in radical excision of the aneurysm and arteriovenous fistulae, more especially so if there is a progressive apical systolic murmur indicating a progressive left heart failure. When the condition affects an arm or a leg there is usually an increase in the length and size of the affected limb, and this causes more disability when the leg is affected. The discrepancy in length in the young can be overcome by epiphysial arrest, if the vascularity of the area of operation allows it.

These unusual congenital deformities are ascribed to a defect in the vascular mesoderm at an early stage of development; so there is no reason why the nutrient vessels to a bone should not be affected.

CASE REPORT

A girl aged nine, an only child, complained of intermittent pain in the left thigh for six months. There had been no injury. Up to the time of onset she had enjoyed normal health, and she was still able to play games and to lead a full life at school. The pain kept her awake at night, especially after exercise, and later she started to limp and to get slightly out of breath. There was no relevant abnormal family history.

Examination—The child walked with a slight limp affecting the left leg. She indicated the middle third of the left femur as the site of the pain. There was local tenderness at that point, with slight thickening: the girth of the thigh at its mid-point was increased by one inch. Movements at the hip and knee were full and painless. I did not measure the length of the limbs or use a stethoscope at this initial examination.

A radiograph showed a multiloculated cortical erosion of the middle third of the posteromedial part of the left femur (Fig. 1). The appearance was that of a benign tumour. Nevertheless the clinical picture was suspiciously malignant; so biopsy was advised, with a provisional questionable diagnosis of monostotic fibrous dysplasia.

FIG. 1
Figure 1—Initial plain radiograph. Figure 2—Arteriograph showing the arteriovenous fistula involving the bone.
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Operation—I exposed the femur through a lateral incision, unmindful at the time that the radiograph showed the part involved to be the postero-medial aspect of the femur. This providential mistake proved the true nature of the pathology to my naked eye and forefinger in a direct, as well as an indirect, sense. For, on making a small opening in the cortex of the normal looking femur, I noticed an underlying membrane which had the glistening appearance of the lining of a bone cyst. The fluid in the cyst was bright red, a finding that baffled me at the time. However, thinking it was an unusual type of bone cyst, I ruptured the membrane with a curette and was greeted by a torrential gush of bright red blood. This was no venous oozing, but massive arterial bleeding. The tip of my forefinger fitted the hole nicely, whereupon all bleeding stopped dramatically, and it was then I first felt the strange fluttering of the arteriovenous fistula. The slightest release of pressure from the hole allowed recurrent arterial bleeding under considerable pressure. In fact I was now dealing with a strange situation: that of a bony pipe flowing with arterial blood under pressure, and with a half-inch hole in the pipe. It badly needed a bung of some sort, and I was considering a sterile cork when the third tube of Horsley's wax finally plugged the hole. After stitching up the wound I put a stethoscope on the thigh and we all listened to the strange cavernous rumblings of an arteriovenous aneurysm involving bone, at the same time noting that the patient had a fairly pronounced apical systolic murmur.

Progress and further treatment—The girl made an uneventful recovery from the operation and later a femoral arteriograph was performed by my colleague, Dr B. M. Maxwell. This showed
the multiple ramifications of the arteriovenous anastomosis involving the femoral artery and vein, with obvious involvement of the bone at the site of the multiloculated area of erosion (Fig. 2). Mr D. Ll. Griffiths, whose advice I sought, suggested as complete an excision as possible because: 1) there were signs of left heart failure; 2) the femur was structurally weaker at the affected point and might break with disastrous results; and 3) the plugging of the hole in the femur might not be as permanent as we hoped. If this excision did not prove successful an amputation might be necessary.

With the aid of Mr A. B. Birt, my surgical colleague, I proceeded to ligate all the anastomosing arteries from below upwards, starting just above the knee (Fig. 3). It is surprising how many have to be ligated, and this time it was not until we were well into the groin, having ligated five arteries below the inguinal ligament, that we found a large artery overlying an enormously distended vein which seemed to be the main culprit. After this artery and the underlying vein were excised the thrill stopped with dramatic suddenness, and the limb became silent. The anaesthetist noticed that the systolic murmur had also disappeared.

The circulation of the left leg appeared normal at the conclusion of the operation and there was normal pulsation in the popliteal and femoral arteries.

Recovery was uneventful and the child was allowed up three weeks later. At this stage she had a slightly increased systolic thrumming over the left femoral artery, just below the inguinal ligament to the medial aspect of the thigh, but otherwise sounds were normal.

She has been back at school for four years and serial radiographs showed that the affected area of the femur filled in rapidly and has appeared almost normal for three and a half years, except for the hole that I made. This hole, presumably, is still plugged with Horsley's wax, and unlikely to disappear (Fig. 4).

When I last examined her, four years after the operation, her legs were equal in length. The left thigh was still larger in girth by two and a half inches than the right one. She walks without a limp and leads a normal active life. Examination of the heart shows no abnormality.

**DISCUSSION**

The diagnosis is not in question, but how I might have arrived at the diagnosis more easily certainly is. Those who advocate arteriography for all bone tumours would be satisfied, and this may be the answer, because in competent hands it provides additional evidence without risk. However, after this experience I consider that auscultation of suspected bone tumours should be an important part of the routine clinical examination. It is interesting that the localised changes in the femur could not be distinguished radiologically from cortical monostotic fibrous dysplasia, and the dramatic improvement in the appearance of the bone after ablation of the aneurysm was striking.

There is no doubt that there was a minor arteriovenous fistula not involving bone, because there was one localised area of thrumming just above and lateral to the knee. This
area has been tender, but recently all tenderness has disappeared and the bruit has stopped, indicating a spontaneous obliteration of the fistula.

At one time, two years after the operation, this child showed an increase of an inch in the length of the left leg, but the limbs are now equal in length, showing that growth is taking place normally. Although she will be under observation for some years to come, she is no longer living on the edge of a catastrophe such as might have befallen her in the event of a pathological fracture of her femur.

A further femoral arteriograph (Fig. 5) was performed by Dr B. M. Maxwell three years after the operation and this showed a nearly normal vascular picture and no evidence of an arteriovenous aneurysm over the outer aspect of the leg. The tracing shows the original area of anastomosis faintly outlined to demonstrate the extent of ablation. This may be because the arterial supply comes from an enlarged gluteal or aberrant obturator or profundus femoris artery, beginning above the inguinal ligament and therefore not filled with the radio-opaque medium.

**SUMMARY**

1. A case of congenital arteriovenous aneurysm involving the left femur, with dangerous cystic changes in the shaft of the bone, is described.
2. After ligation of the anastomosing fistulae the radiological appearance of the femur became normal.

My sincere thanks are due to my colleagues, Mr A. B. Birt and Dr B. M. Maxwell, for their help and assistance. I also am grateful to Mr D. Lloyd Griffiths for his valuable advice on the treatment of this patient.