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Congenital shortening of one femur in one identical twin

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Congenital shortening of the femur has been described by Ring (1961) as one of a group of congenital abnormalities of the femur. In a series of nineteen such cases, Ring (1959) includes twins where the presence of congenital shortening of one femur in one twin led to the examination and discovery of the same defect in the other twin. The purpose of this paper is to record the presence of congenital shortening of one femur in only one of identical twins and to assess the significance of this as regards the aetiology of this condition.

Case report

Mrs D.C., a 27-year-old West Indian, having previously been delivered of three normal male children, was spontaneously delivered of female twins. The pregnancy lasted 42 weeks and had been entirely normal. There was no pyrexial illness in the first trimester. The mother had taken no drugs, she had had no glycosuria, her Wasserman Reaction was negative and her blood group was O, rhesus positive.

The single placenta had two amniotic sacs and one chorion. The older child weighed 5 lb 15 oz with a body length of 20 in. and a head circumference of 13 1/2 in. The younger child weighed 6 lb 15 oz with a body length of 20 in. and a head circumference of 14 in.

After delivery, it was noticed that the elder twin appeared to have shortening of the left leg above the knee. The left hip abducted to only 90°. There were no other abnormalities detected clinically. X-ray at that time (Fig. 1) showed shortening of the shaft of the left femur with lateral bowing. The cortex of the diaphysis was thickened, almost obliterating the medullary cavity. The centre of ossification for the lower femoral epiphysis was present on both sides, but the centre for the upper tibial epiphysis was present only on the normal side. The hip joints on both sides appeared normal.

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The younger twin appeared entirely normal but was not X-rayed at the time.

The affected child was followed up in both the paediatric and orthopaedic clinics. Her general progress and weight gain were entirely satisfactory. After she started to walk the shortness of the left leg was compensated for by appropriately raising the shoe with a patten. When last seen this child was nearly 8 years old. The only abnormality detected was 2 in. of shortening in the shaft of the left femur. There was a full range of hip and knee movements. The opportunity was taken to examine the other twin. Her legs were entirely symmetrical and identical in their measurements to the normal leg of the affected child. The twins’ blood groups were compared and were found to be identical in all groups that were tested (Table 1). It was not thought justifiable to perform cross skin grafting to confirm this.

**Discussion**

According to Ring's classification (1961), congenital abnormalities of the femur vary from simple hypoplasia to complete absence. These abnormalities may occur alone, or they may be associated with other congenital defects, particularly with other defects of the skeleton. Such congenital abnormalities may be genetically determined or they may be due to extra-foetal factors arising during development. These extra-foetal factors may be chemical, e.g. drugs or heavy metals, or they may be biological in origin, e.g. hormones or substances of viral origin. Such factors are collectively described as 'teratogens' and presumably must reach the foetus through the placental circulation. It is difficult, therefore, to postulate a teratogen having different effects on twins sharing the same placenta.

Similarly, if it is accepted that these twins are in fact 'identical', then they must be genetically identical and this abnormality cannot have been genetically determined.

One of the few influences that could affect one limb of one identical twin is a mechanical one. Simple birth fracture must be excluded by the concomitant delay in the appearance of the centre of ossification for the upper tibial epiphysis and the subsequent behaviour of the femur itself. Compression of the thigh at a time when ossification of the femoral diaphysis is most active could lead to a state where the limb 'would be in a condition of reduced arterial supply combined with venous stasis' (Browne, 1936). The X-ray appearances of the short curved femur with thickening of the cortex almost obliterating the medullary cavity would suggest endosteal new-bone formation and Brookes (1960) has shown experimentally that this can follow occlusion of the principal nutrient vessels of the femur. I submit that this abnormality could have been caused by external mechanical

**Table 1**

The identical blood groups

<table>
<thead>
<tr>
<th></th>
<th>ABO</th>
<th>Rhesus phenotype</th>
<th>Probable genotype</th>
<th>MN</th>
<th>S</th>
<th>PI</th>
<th>Luα</th>
<th>K</th>
<th>Fyα</th>
</tr>
</thead>
<tbody>
<tr>
<td>Affected twin</td>
<td>O</td>
<td>CcDE</td>
<td>CDe/cDe</td>
<td>M</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>Normal twin</td>
<td>O</td>
<td>CcDE</td>
<td>CDe/cDe</td>
<td>M</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
</tbody>
</table>
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compression of the foetus and that mechanical factors must be considered amongst the causes of congenital abnormalities of the limbs.

Acknowledgment

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References


A case of oat-cell carcinoma of the lung with the carcinoid syndrome

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It is now well recognized that malignant neoplasms sometimes give rise to various syndromes by causing over-secretion of hormones. One of the most interesting of these is the carcinoid syndrome which is usually due to excessive serotonin or 5-hydroxytryptamine (5-HT). This has been described in association with carcinoma of the pancreas (Peart et al., 1963), thyroid (Moertel et al., 1965) and islet-cells (Van der Sluys et al., 1964). Williams & Azzopardi (1960) were the first to record a proved case of carcinoid syndrome caused by an oat-cell carcinoma of the lung though Harrison et al. (1957) had previously observed some features of the syndrome in a patient with this carcinoma. Gowenlock et al. (1964) reported another case, but with a slight difference in that the tumour produced 5-hydroxytryptophan (5-HTP) instead of 5-HT. Azzopardi & Bellau (1965) added a third case and there were two more described by Majcher et al. (1965) one of which had an oat-cell carcinoma and the other an undifferentiated one. The following is a report on a case which is probably the sixth recorded so far.

Case report

Mr H.S., a 45-year-old warehouse porter, first saw his doctor in April 1967 because of moderately severe, persistent burning pain in his right foot. This was his only symptom. He was a married man with four children and had had no previous illnesses. There was nothing significant in the family history. He smoked fifteen to twenty cigarettes a day and was a teetotaller. He was sent to another hospital as an outpatient. After X-rays of his lumbar spine and pelvis he was given a low spinal brace and was instructed to wear it. But the pain persisted and soon involved the other foot as well. However, he was able to continue his work, lifting heavy parcels. Towards the end of May 1967 he developed a macular rash over his legs below the knees but this disappeared in a few days. The following month he went to see his doctor again and complained of a slight cough and tiredness in addition to the pain in the feet. He was now also having bouts of diarrhoea over periods of 3–4 days with four or five stools per day. An X-ray of the chest showed a faint ill-defined opacity in the right lower zone and the patient was admitted to the Western Hospital on 11 July 1967.

His chief complaint still was the burning pain in the feet. It was constant, with no aggravating factors, and was occasionally severe enough to wake him up from sleep. He had no breathlessness but complained of feeling weak in the legs on walking about 100 yd. He had no diarrhoea at the time but had had several bouts of it at home. He had lost a few pounds in weight.

On examination he appeared fairly well. There was marked telangiectasia of the face. The skin over the feet and ankles was erythematous, rough and somewhat scaly. There was no finger clubbing or joint swelling and no lymph glands were palpable. Pulse regular, 88/min, and BP 180/100 mmHg. All peripheral pulses were felt. Heart sounds were normal and there were no murmurs. The percussion note was impaired and breath sounds diminished over the right lower chest posteriorly. The liver was enlarged to just below the umbilicus and was nodular and slightly tender. There was no abnormality in the central
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