Lesson of the Week

Salmonella osteomyelitis presenting as "hand-foot syndrome" in sickle-cell disease

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Sickle-cell disease is occurring more often in Britain than before. Of 70 patients with this disease in the London Borough of Brent, more than half were born in Britain.¹ Homozygous sickle-cell disease commonly presents first with recurrent swelling of the hands and feet owing to bone ischaemia—the "hand-foot syndrome." The severity and duration of the swelling are variable and resolve spontaneously. The course of the disease, however, may be complicated by osteomyelitis, possibly with associated septicaemia and then death, or with chronic suppuration and discharging sinuses. Such complications are rare in Britain. In a recent review from the Children's Hospital, Birmingham,² no case of osteomyelitis had been recorded for 10 years.

I report on a case of salmonella osteomyelitis presenting as hand-foot syndrome in an infant who has homozygous sickle-cell disease.

Case report

A 20-month-old half-caste West Indian girl presented with a five-day history of painful red swelling of the left hand in the region of the thenar eminence. During the previous three weeks her other hand and both feet had been swollen and painful, but never before that. She had been admitted at 6 months of age with fever and gastroenteritis when the diagnosis of sickle-cell disease was made, and also at 12 months with fever and pneumonia.

When seen at 20 months she was afebrile and clinically well. Her haemoglobin concentration was 7.9 g/l. An x-ray film of her left hand showed soft tissue swelling only. Analgesia with paracetamol was given as required. Five days after admission she became febrile, the tense red swelling showed signs of becoming localised, and she was reluctant to use that hand. Under general anaesthesia pus was drained and the culture grew *Salmonella schwarzengrund*, which was sensitive to ampicillin and gentamicin: *Salmonella* was also cultured from the stools. Treatment was started with intravenous ampicillin and gentamicin. The gentamicin was discontinued after 17 days, but the ampicillin was continued for six weeks. An x-ray film taken four days after drainage showed complete destruction of the first metacarpal joint (fig 1). Subsequently a new, shortened bone formed (fig 2). She has recovered complete function and remains well.

Salmonella osteomyelitis may cause the painful swelling of the hands and feet that fails to settle after an episode of sickle-cell disease

FIG 1—X-ray film of left hand, four days after pus was drained.

FIG 2—X-ray film of left hand three weeks after fig 1, showing new bone formation and shortened first metacarpal bone.

Discussion

The hand-foot syndrome arises from the vaso-occlusive sickle crisis. There are always a few irreversible sickled cells in the circulation in sickle-cell disease. Hypertonicity, acidosis, or hypoxia fosters further sickling, but there is not always a distinct event that precipitates such a crisis and many ischaemic episodes remain subclinical. Bone ischaemia may progress to non-septic

bone infarction with changes seen on x-ray examination that are indistinguishable from those of osteomyelitis.

In a review of a large number of cases of hand-foot syndrome in New York³ no salmonella osteomyelitis was reported, and the authors favoured conservative treatment rather than prolonged antibiotic treatment. The experience in Birmingham was the same. In Nigeria, however, 63 cases of salmonella osteomyelitis occurred over eight years and many presented as hand-foot syndrome—57 of the patients had sickle-cell disease.⁴ Adeyokunnu and Hendrickse suggested that salmonella osteomyelitis may occur because the organism spreads from the intestine, helped by devitalisation of the gut caused by intravascular sickling, and bone infarcts then become infected by transient bacteraemia. In the infant reported here the history suggested that sickling episodes recurred for three weeks and that the episode that resulted in salmonella osteomyelitis coincided with excretion of salmonella in the stools. This lends weight to Adeyokunnu and Hendrickse's hypothesis.

Although rare in Britain, salmonella osteomyelitis should be

considered in any patient with the hand-foot syndrome that shows no signs of improving after a few days of conservative treatment.

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MATERIA NON MEDICA

Winter fungi

One of my more interesting colour slides of fungi, at least to me, shows a group of glistening ink caps, *Coprinus micaceus*, which I photographed early one December morning. The ink caps were attractively covered in thick rime, after an overnight frost; yet when the ground thawed as the sun rose, so the fungi shrivelled to a few moist, black cords. Of course, one could have predicted the effect of freezing on the ink caps: fungal cells are delicate and people do not normally look for mushrooms or toadstools in December.

The December of 1981 was one of the coldest and most wintry that many can remember. I was surprised, therefore, when walking on the Quantock Hills at Christmas time, to see a snow-surrounded, dead elm stump which was covered with a thriving colony of colourful tawny and yellow agarics. After collecting some of the caps, which were sticky with broad, pale yellow gills, I placed them on coloured paper for a few hours. On investigation, I found that dense, radial spore patterns had been produced, so clearly these winter fungi were active and seemingly unaffected by subzero temperatures. They proved to be velvet shanks, *Flammulina velutipes*, which are, apparently, normally seen growing during winter, from November to March. When removed from their tough, wiry stalks the caps are good to eat, so they are worth considering as a tasty and original addition to the Christmas fare, always provided they have been identified with certainty.

Another healthy and living fungus I found that chill day was the Jew's ear, *Auricularia auricula*, sprouting from a dead elder branch. These fleshy, ridged, ear-like and purple-brown fungi also grow and produce spores under wintry conditions; indeed, when the large, white sausage-shaped spores were inspected under the microscope, they looked viable enough. Jew's ear, like the velvet shank, is well worth eating and thus is a valuable find in the countryside at a season when there are not many edible items for which to search.

So Arctic had been the recent weather that it was obvious that both fungal species had an efficient antifreeze mechanism, each having tissues capable of considerable supercooling. The substances elaborated by the fungi must surely have significant and interesting properties; if they have not been investigated already, is any plant chemist looking for a research project?—PHILIP RADFORD, West Bagborough, Somerset.

Materia Medica Ecclesiastica

A close examination of the decoration and furnishings of many English churches reveals that the mind of the mediaeval Englishman was no less occupied with his ailments than is the mind of his twentieth-century counterpart.

Toothache apparently plagued our mediaeval forbears. Wells

cathedral has a series of carvings illustrating the agonies of this condition—said to be relieved by prayer at the tomb of a local bishop. Saint Apollonia, patron saint of dentists, is shown in several rood screens, such as those of Barton Turf and Ludham in Norfolk, where she stands holding a pair of forceps grasping a tooth. Another patron saint of the sick is Saint Roche, a fourteenth-century healer depicted with a plague bubo on his right thigh. He can be seen on a sixteenth-century screen in Romsey Abbey and as a figure carving in All Hallows by the Tower, London.

For the neurologist, facial palsies abound. A top-hatted gargoyle leers lopsidedly at visitors to Winchcombe Parish Church, and a similarly gruesome corbel is seen in the Temple Church, London. Neither is ophthalmology neglected. A bespectacled Saint Michael is shown on the fifteenth-century rood screen of St Agnes, Cawston, and another Temple Church corbel has a pronounced convergent squint. Rheumatologists will delight in the many representations of Master John Schorne. A Buckinghamshire rector of the late thirteenth century, he was famed for cures of the ague and gout.¹ Screens such as those of Cawston and Suffield depict him conjuring from his boot the goutish imp—an evil horned dragon. They may also speculate on the disability of the woman with crutches shown in a doom painting at Oddington, or on the illness of the boy with contracted joints whose cure is illustrated in the Saint William window of York Minster. For the gastroenterologist, a stained glass window of 1220 in Canterbury Cathedral illustrates a brisk haematemesis. The church door at Hadstock, Essex, possibly the oldest in England,² is reputed still to have nailed to it the skin of a sacrilegious Dane who was punished for his crime by flaying.

What of the cures available? Apart from prayers said at specific tombs, such as that of Saint Thomas of Canterbury, which was widely acclaimed for its therapeutic effects, there were the physicians—they are satirically represented in misericords as monkeys, often clutching urine bottles.³ A fine example can be seen in the choir stalls of Manchester Cathedral. Sometimes the church was forced to limit its parishioners' attempts at self-medication, thus explaining the existence of locks on the font rims of churches such as Stoke Charity and Bosham. On Easter Day the font water was blessed and unless securely locked was gradually purloined, to be used as a healing fluid.

For all that, cures must have been few and far between. The many examples of offspring holding a skull while kneeling at the tomb of their parents (thereby indicating their death in youth) and the memorial brass to a baby of Laverham are moving reminders of the early death of many mediaeval Englishmen.—ALASTAIR TURNBULL, medical student, London.

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