FRIEDRICH’S DISEASE

ASEPTIC NECROSIS OF THE STERNAL END OF THE CLAVICLE

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Friedrich’s disease is a rare condition which may be misdiagnosed as osteomyelitis, arthritis or tumour. Four case reports are presented in which it is shown to be an aseptic necrosis of the epiphysial region of the sternoclavicular joint which resolves spontaneously. Correct diagnosis will spare patients unnecessary operations and unnecessary medication.

Considering the few publications in the literature, there is little doubt that Friedrich’s disease is rare. We had the opportunity to treat four such patients (one of them bilateral), in two of whom the disease was proven histologically. The disease is not well known; clinical and radiographic signs may mislead in the direction of osteomyelitis, arthritis, or tumour. Awareness of the possibility of aseptic necrosis may spare the patient unnecessary operations, and our experience with the first two patients enabled the second two to be treated conservatively (and successfully). This report presents our experience and follow-up of patients with this relatively rare disease.

Friedrich first described it in 1924 in two patients. Fischel and Bernstein (1975) reported one other patient and a review of the literature yielded 10 more (including the two reported by Friedrich). To the best of our knowledge, since 1975 there have been no other reports in the English literature.

CASE REPORTS

Case 1. A 14-year-old girl was admitted to our department after complaining for about two months of spontaneous pain and swelling in the region of her right sternoclavicular joint. She had no fever and her erythrocyte sedimentation rate was 15 and 45 millimetres in the first and second hour respectively, so it was decided that she should only be observed. Six years earlier she had had suspected familial Mediterranean fever which had never been confirmed. On examination a tender swelling measuring two by three centimetres could be palpated at her right sternoclavicular joint but there was no other abnormality. The white cell count, haemoglobin and haematocrit were normal, latex and Rose-Waaler tests were negative, as was the antistreptolysin titre (fibrinogen 350 milligrams per 100 millilitres). Liver and kidney function tests were also normal. On needle aspiration some turbid liquid was obtained which was negative for Mycobacterium tuberculosis, in all routine cultures and also on direct smear. She had not received antibiotics.

Tomography revealed bone damage in the clavicle close to the sternal joint (Fig. 1). Since osteomyelitis or a tumour were suspected, a biopsy was done. At operation the bony lesion was curetted until normal bone was seen macroscopically. Microscopy showed necrotic bone with no other findings. Nevertheless, the patient was given oxacillin 2.0 grams per day for a few days by which time recovery was complete.

She remained well until, at the age of 16 years, she again complained of spontaneous pain and tenderness, this time in her left sternoclavicular joint. Again there was no fever and the white cell count was normal. The erythrocyte sedimentation rate this time was 32 and 58 millimetres. On admission she had a tender swelling of her left sternoclavicular joint with some hyperaemia. The right, previously operated, side was without remarkable findings. All additional clinical and laboratory findings were normal. She had received no antibiotics. Needle aspiration resulted in some turbid liquid which was again negative on culture and smear. She was operated on and the lesion curetted. Microscopy showed necrotic bone with no additional findings. This time she received no antibiotics and recovery was complete. She has now been followed up for a further four years.

Fig. 1

Tomogram showing bone damage in the clavicle close to the sternal joint.
Tomograms showing destruction of compact bone at the medial inferior end of the clavicle with irregularity and density of bone tissue.

**Fig. 4**—Cystic area filled with debris of necrotic bone fragments surrounded by intact bone. (Haematoxylin and eosin, ×24.)

**Fig. 5**—Higher magnification of necrotic bone fragments. (Haematoxylin and eosin, ×150.)

**Case 2.** A 26-year-old woman presented suffering from pain and swelling at the left sternoclavicular joint, with no history of trauma or fever. The pain increased on trying to move her left shoulder. Tomography showed destruction of compact bone at the medial inferior end of the clavicle as well as some irregularity of the structure of the cancellous bone (Figs 2 and 3). Laboratory examinations, liver and kidney function tests and protein electrophoresis were all normal. Needle aspiration resulted in some turbid liquid which was negative on direct smear and on culture. Nevertheless, it was decided to operate for it was felt that a tumour had not been ruled out definitely. At operation, about 1.5 centimetres of the sternal end of the clavicle was resected. The articular surface appeared normal. Microscopy revealed aseptic necrosis of bone with hyperplastic bone marrow and focal fibrosis in the surrounding bone (Figs 4 and 5). This patient too, received no antibiotic treatment before, during, or after her operation. Recovery was complete. She has now been followed up for three years.

**Case 3.** A girl aged six years was referred to the orthopaedic clinic because of pain and swelling of the right sternoclavicular joint, which appeared spontaneously two weeks before. There was no fever. Latex, Rose-Waaler and antestreptolysin titre tests were all normal, as were haemoglobin, haematocrit and the white cell count. The sedimentation rate was elevated at 50 and 80 millimetres. Tomography revealed irregularity of bony structure and some deformity of the sternal end of the clavicle (Fig. 6). Aspiration cultures as well as direct smears were negative. Considering our experience with the earlier patients who were operated on, we decided to treat her by observation only. After about two months most of the swelling and tenderness disappeared and shoulder movement became normal. The sedimentation rate also became normal. Six months later there was little residual swelling and no pain. She has now been followed up for two years and she is doing well.

**Case 4.** A 58-year-old man presented in our clinic with tender swelling of the left sternoclavicular joint. This time there was a history of injury to the left shoulder four months previously but the left sternoclavicular pain and swelling appeared more than three months after the injury. On examination a hard (bony?) swelling was palpated at the joint. Pain
increased on abduction of the left shoulder. Routine laboratory tests were normal. Tomography revealed irregularity of bone structure at the sternal end of the clavicle suggestive of avascular necrosis of bone (Figs 7 and 8).

Again we decided on the basis of our previous experience to follow the patient without any treatment. The tenderness disappeared, and after one and a half years most of the swelling has also disappeared. Recent radiographs have revealed normal bone structure.

DISCUSSION

The aetiology of Friedrich's disease is unknown, partly because of its rarity. Fewer than 20 cases have been reported to date. It is probably due to aseptic necrosis at the epiphysial ends of some bones, as was suggested by Friedrich in 1924. The epiphysial plate may have closed already, as in our two older patients; the epiphysis of the sternal end of the clavicle begins to ossify at the age of 18 to 20 and closes at the age of 25 (Jit and Kulkarni 1976).

None of our patients had local or systemic treatment with corticosteroids. Although one patient (Case 4) had an injury four months previously, it seems unlikely that the disease resulted from this. Our first patient is probably the only bilateral case reported.

Because of its rarity, the disease is probably often confused with arthritis, osteomyelitis, or tumour, and may not, in fact, be so rare but only misdiagnosed. Awareness of the possible presence of this disease will save patients an unnecessary operation and unnecessary medication. This is also the conclusion of Köhler and Zimmer (1968).

REFERENCES