SURGICAL TREATMENT OF MANUBRIO-STERNAL PAIN IN BEHÇET'S SYNDROME

Report of a Case

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The clinical triad of recurrent mouth ulcers, genital ulcers and anterior uveitis has been documented sporadically in the literature for over fifty years (Bluthe 1908), but the eponymous title by which it is now known derives from a more recent account of the symptom complex (Behçet 1937). Strachan and Wigzell (1963) pointed out that the concept of a "triple syndrome complex" has now outlived its usefulness, for the range of clinical manifestations in this condition is wide.

Joint symptoms probably occur in something over 50 per cent of patients with Behçet's syndrome (Oshima, Shimizu, Yokohari, Matsumoto, Kano, Kagami and Nagaya 1963). The arthropathy is often mild or inconspicuous, but occasionally it is the most striking feature. We have found no reference to arthropathy of the manubrio-sternal joint in this condition, although involvement of this joint is well recognised in ankylosing spondylitis. We therefore report a patient with Behçet's syndrome in whom the clinical picture was dominated by symptoms arising from arthritis of the manubrio-sternal joint.

CASE REPORT

A forty-two-year-old man, formerly a coal miner, was first seen for chest pain in April 1965. Since the age of twenty years he had suffered from recurrent ulcers in the mouth. Appearing singly, or in crops of two or three, these ulcers were about five to ten millimetres in diameter and affected any part of the oral mucosa. New ulcers appeared about once a month, lasted approximately two weeks and were always painful. A variety of topical medicaments including corticosteroid preparations had been doubtfully effective.

In December 1963 the patient developed an eruption of pustules scattered over both legs. These cleared gradually except over the medial aspect of the left upper calf where a number of pustules coalesced to form an ulcer fifteen centimetres in diameter, for which he was admitted to another hospital. The ulcer healed slowly over the next month, leaving a scar. While in hospital he suffered, for the first time, a crop of small painful ulcers on the scrotum. This coincided with an exacerbation of the mouth ulcers, and with slight urinary symptoms in the form of dysuria and mental irritation. At about the same time he first noted the chest pain which subsequently developed into the main feature of his illness. Beginning as an ache in the region of the manubrio-sternal joint, pain gradually increased until any movement of the chest produced a sharp pain in the joint. Coughing and sneezing became excessively painful; in fact any movement of the upper half of the body produced sternal pain, and he began to adopt a hunched posture. By January 1965 he had given up his work. Mouth ulcers continued, and in February 1965 there was a further episode of scrotal ulcers.

The only other joint symptoms were slight pain and stiffness in the neck and left shoulder for two months; otherwise his general health had been excellent and he had led a very active life. He had suffered from mild psoriasis since the age of five years, but had never had erythema nodosum, nor was there any history suggestive of deep or superficial thrombophlebitis. He denied any eye complaints, but his medical file at work recorded that he had blepharitis followed by a chalazion in June 1963. He had never suffered from a urethral discharge and had not had any neurological symptoms.
On examination the manubrio-sternal joint and the surrounding area were extremely tender to touch although not obviously red or swollen. Deep breathing and passive distortion of the thoracic cage were both very painful and he moved about cautiously in order to avoid the sharp manubrio-sternal pain which any chest movement provoked. Movements of the cervical spine and of the left shoulder were slightly limited by intrinsic pain. The rest of the locomotor system showed no abnormality. There were no rheumatoid nodules.

In the mouth were two painful lesions with the appearance of aphthous ulcers, one on the floor of the mouth, the other on the gingival margin. Both were oval, about ten by five millimetres in area and one or two millimetres deep. A few faint scars, about the same size as the mouth ulcers, marked the site of previous scrotal ulcers and there was a round "tissue paper" scar, about fifteen centimetres in diameter, on the inner aspect of the left calf.

The cardiovascular, respiratory and central nervous systems were normal, and abdominal palpation revealed no abnormality. Ophthalmological examination was negative.

**Investigations**—The erythrocyte sedimentation rate was nine millimetres in the first hour (Westergren), haemoglobin 15 grammes per cent and white blood cell count 5,100 per cubic millimetre with a normal differential count. Serum albumin was 4.7 grammes and serum globulin 2.6 grammes per cent with a normal electrophoretic pattern. The serum alkaline phosphatase was 8.6 King-Armstrong units, serum calcium 9.6 milligrams, inorganic phosphorus 2.8 milligrams and blood urea 40 milligrams per 100 millilitres. Three L.E. cell preparations, an anti-nuclear factor test, the latex test and serological tests for syphilis were all negative. An electrocardiograph was normal.

A standard chest radiograph was normal. Tomographs of the manubrio-sternal area failed to define the joint. A radiograph of the pelvis was normal, with no evidence of sacro-iliitis.
Histological examination—A section from a mouth ulcer showed a slightly depressed ulcer of the oral mucosa with non-specific features. Material was obtained from the manubriosternal joint by open biopsy (Fig. 1). The histology of the bone and deeper cartilage was normal, and there was no extension of ossification either in the cartilage or the capsule. In some areas the cartilage was breaking up in relation to vascular granulation tissue with a small number of polymorphonuclear leucocytes, but no frank pus. No organisms were seen in stained sections. These changes were opposite the middle of the joint. There was no evidence of necrosis in the compact cartilage nearer the bone. No rheumatoid lymphoid foci were seen around the blood vessels. Some proliferation of large mononuclear cells, locally resembling the palisade cells of a rheumatoid granuloma, occurred, but these were apparently developing into cells forming a distinctly chondroid fibrous tissue. In view of the broken-up nature of this part of the material this sequence was not completely established. The inflammation was more active and less ossifying than that seen in ankylosing spondylitis (Cruickshank 1956). There were no genuinely rheumatoid features or active immunologically competent cells. The distinctly purulent edge to the inflammation had its nearest parallel in some examples of dysenteric arthritis.

Progress—For a while the patient’s symptoms were partly relieved by phenylbutazone and later by indomethacin, but after seven weeks mouth ulcers were severe and the manubriosternal pain was worse. Skin lesions continued to appear (Fig. 2). Steroid therapy, started in August 1965 (prednisolone 10 milligrams daily) produced considerable improvement and he was able to return to work. However, the manubriosternal pain and tenderness remained troublesome and local treatment was considered essential. He was given a course of local deep x-ray therapy (125 r daily for ten treatments at 220 kilovolts), again with transitory improvement. Later, a local injection of hydrocortisone with lignocaine was tried without sustained effect. By December 1965 severe manubriosternal pain still dominated the clinical picture. It was decided to arthrodes the joint.

At operation in February 1966 the front of the sternum was exposed through a midline incision and the manubriosternal joint was excised. Its gross appearance was unremarkable, there being a generalised fibrous thickening of the anterior and lateral ligaments with a gritty mass of similar tissue in the cavity. This was evacuated and a slot prepared for an inlay bone graft taken from the left ilium; the graft was fixed securely with a Vitallium plate and screws (Fig. 3).
Histological examination of the excised material showed subacute inflammation of the perichondrium with local small purulent foci, at least one of which involved the cartilage. No rheumatoid lymphoid foci were seen, but some of the histiocytes had more than one nucleus and were a little like those of rheumatoid granulation tissue.

Immediately after operation the symptoms were markedly improved and he was discharged home within two weeks. So far as this joint is concerned he has remained completely free of pain.

The other manifestations of the disease (mouth and skin lesions and joint pains elsewhere) continue and in addition he developed a corneal ulcer. There have been episodes of dyspepsia and mild emotional abnormalities thought to be a complication of steroid therapy; attempts to reduce the steroid dosage have resulted in flares of the condition. A course of gold injections has been started.

**DISCUSSION**

There seems little doubt that this patient suffers from Behçet’s syndrome. The recurrent mouth and scrotal ulcers and the pustular lesions on the legs are typical. The eye lesion completes the clinical triad, although this has taken the form of corneal ulceration instead of the more characteristic anterior uveitis, retinitis or optic neuritis (Perkins 1961). There is mild arthropathy of the left shoulder and cervical spine, but the clinical picture was dominated by the severe pain arising in the manubrio-sternal joint.

A recent review of the joint involvement in Behçet’s syndrome (Strachan and Wigzell 1963) suggests that there are no characteristic features—large or small joints being affected singly or in groups. The arthritis may coincide with, precede or follow other features of the syndrome.

In an earlier review France, Buchanan, Wilson and Sheldon (1951) described the histological changes seen in a synovial biopsy from an affected joint. These include hyperaemia, oedema and perivascular round cell infiltration.

In the present case histology of the manubrio-sternal joint showed a subacute inflammatory reaction involving the superficial layers of cartilage. Rheumatoid features were absent, although the anatomical arrangement of this joint makes comparison with material from synovial joints difficult. The reaction differs from that seen in ankylosing spondylitis, the only condition to involve this joint with any frequency. The present findings do not clearly favour any particular etiological theory.

Joint symptoms tend to be a minor feature of the clinical picture, and the rheumatologist usually sees these patients referred by a dental, ophthalmic or venereological colleague. Occasionally arthralgia is the presenting symptom, so that enquiry about mouth and genital ulceration should be included in the investigation of all patients with joint pains. Oshima et al. (1963) collected eighty-five cases of Behçet’s syndrome in one unit in less than four years, so it may well be much more common than is at present appreciated. Certainly all cases of unexplained sero-negative arthropathy require careful investigation to exclude this diagnosis.

The non-specific clinical and histological features of the mouth ulcers are typical. Dowling (1961) suggested that the mouth ulcers of Behçet’s syndrome tend to be deeper than common aphthous ulcers, but most authors seem unable to differentiate the two. The difficulty is increased by the fact that mouth ulcers may precede the other features by many years. One cannot entirely exclude the possibility that the two conditions are the same, the common aphthous ulceration being an incomplete expression of the fuller syndrome. The final decision about this awaits the development of a diagnostic test for Behçet’s syndrome.

The occurrence of pustulation at the site of a scratch in this patient is interesting. Blobner (1937) drew attention to the undue sensitivity which these patients may show to minor traumata such as needle pricks or injections. Fellner and Kantor (1964) found that the intradermal injection of 0·1 millilitre of normal saline was followed by the appearance of a pustule. The test, which they suggested might indicate disease activity, was negative in this patient. There was also no defect of healing after operation.
Successive descriptions of the syndrome have steadily increased the list of known clinical manifestations of Behçet’s syndrome. To the original triad of eye, mouth and genital lesions, one must now add a variety of skin lesions, arthropathy, superficial and deep phlebitis, and a variety of neurological features (Rubinstein and Urich 1963). After a long, fluctuating course, often benign and spread over many years, the disease may assume a more florid form with brain-stem lesions and a fatal outcome (Pallis and Fudge 1956).

The etiology remains unknown. Behçet himself suggested a viral infection and a distant focal infection. Sezer (1953, 1956) argued the case for a virus etiology, but Dudgeon (1961) found the evidence inconclusive. Recently the possibility that the condition may be an auto-aggressive state has been raised. Studies by Oshima et al. (1963) appear to support this.

With regard to the local manubrio-sternal condition, as in other inflammations of the rib cartilages and sternum (Elson 1965), there was extreme pain; the patient was unable to bear the lightest palpation. Arthrodesis of this joint must be an unusual procedure. The method used was effective, although the texture of the posterior cortex of the manubrium was soft and it would have been easy to strip the screw threads in the bone. The immediate relief of pain suggests that the excised capsule was the locally irritated structure.

**SUMMARY**

1. A case of Behçet’s syndrome in which the main complaint was severe pain in the manubrio-sternal joint is described. Arthrodesis relieved the pain.
2. Histological material from a mouth ulcer and from the manubrio-sternal joint showed non-specific inflammatory changes but failed to throw light on the etiology.

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**REFERENCES**


