A CASE OF PROGRESSIVE JOINT DISORDERS CAUSED BY INSENSITIVITY TO PAIN

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Rapidly progressive degeneration of joints in persons whose sensory function is impaired is well known in tabes dorsalis, syringomyelia, diabetic neuropathy and other diseases of the nervous system. The occurrence of such degeneration as a result of simple insensitivity to pain is less common.

CASE REPORT

A woman, born in 1926, has shown from birth a consistent lack of appreciation of pain. There is, and has been, no known similar disturbance in other members of her family. In 1948, at the age of twenty-two, she was investigated by Dr F. L. McNaughton at the Montreal Neurological Institute, but no evidence was found of any defect in her nervous system, and she was able to distinguish pinpricks for what they were, though to her they were quite painless. Temperature sense was normal, but extremes of temperature produced no pain.

Stimuli that would normally produce pain evoked neither verbal report of pain, nor wincing, nor withdrawal of the part tested, and the blood pressure, pulse rate and respiratory rate were not affected. There was no evidence of psychological defect or of personality disorder. The electro-encephalograms were normal.

When twenty-one months old she had had a large painless abscess over her occiput. At three years of age she had had an equally painless osteomyelitis of her right calcaneum, and a pinch grafting operation for a painless burn which she had sustained when kneeling on a radiator. At the age of six she had acute pyelitis and osteomyelitis of the left femur, again without pain. At seventeen she noticed loose bodies, which were removed from her left knee (Fig. 1). At the same time, a thoraco-lumbar kyphosis was noticed, and radiographs

FIG. 1
Radiographs showing loose bodies in patient's left knee at age of seventeen.
FIG. 2
Upper lumbar spine at age seventeen.

FIG. 3
Figure 3—Right hip at age of twenty-two. Figure 4—Right hip at age of twenty-three. Figure 5—Appearance of the right hip after "shelf" operation.
of the spine showed some bone resorption in the upper lumbar bodies with condensation and osteophyte formation in the same region (Fig. 2). By that time her tongue was scarred and the soles of her feet bore numerous scars of painless injuries.

In 1948, at the age of twenty-two, she noticed "crunching" noises in her right hip and swelling in this region. Radiographs showed loss of substance in the acetabulum and in the femoral head (Fig. 3). Within four months she was limping. The hip then showed an abnormally free, painless range of movement in all directions, with grating. There was a considerable bony swelling palpable in Scarpa’s triangle and the leg was two inches shortened. Her shoulders could now be subluxated and reduced at will. A year later, radiographs of the hip showed further destruction, with subluxation of the femoral head (Fig. 4). A "shelf" operation was undertaken, but it did not correct the limp.

In 1951 her legs began suddenly giving way under her without warning, and pyramidal signs appeared with diminished sensation in all forms below the eighth-ninth thoracic level, and paralysis of the bladder sphincter. Radiographs now showed a destructive lesion at the T.10-11 interspace, with a soft-tissue mass around the involved vertebrae (Fig. 6). A lumbar myelogram showed a block at the tenth thoracic level, with a filling defect at the level of the tenth thoracic disc. Operation (right hemi-laminectomy, by W. V. Cone and J. G. Petrie) revealed compression of the cord at that level, with much granulation tissue of varying consistency in the extra-dural space. The cord was decompressed by removing bone and granulation tissue, and pulsation was restored in the theca. The spine was fused from T.8 to L.1. The histology of the granulation tissue revealed nothing specific. Progress—The patient made some improvement. Bladder function has returned, but she still has a severe degree of paraparesis.

DISCUSSION

Though some recent authors (Casagrande, Austin and Indeck 1951) have ignored loss of pain-sensation as an important factor in the cause of neuropathic joint disease, there is no doubt that this function is abnormal in the large majority of joints so affected. In this case changes identical with those of "Charcot’s joints" in the knee, hip and spine occurred in a person with no other defect in her nervous functions until the spinal lesion itself produced a paraplegia. It seems probable that these joint changes were due solely to the lack of protection usually given by the sensation of pain.

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REFERENCE