THE ARTHROPATHIES.

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The occurrence of arthropathies in connection with chronic disease of the nervous system was first commented upon, in 1831, by J. K. Mitchell who formulated the theory of "trophic centres" controlling the nutrition of bones and joints. Charcot, in 1868, described the trophic disorders of *tabes dorsalis*, including the joint lesions which bear his name, the first case in this country being recorded by Clifford Allbutt during the following year. Some twenty years later, similar arthropathies were recognized and reported in association with syringomyelia.

Tabes Dorsalis.

In *tabes dorsalis*, arthropathies occur in about 4 per cent. of all cases and are most frequent in the lower limbs, especially the knee joint, while the wrists and finger joints are very rarely affected. Thus, of 268 tabetic arthropathies collected by Chipault, no less than 207 were arthropathies of the lower limbs, 120 with the knee and 57 with the hip joint affected. The wrist was involved in only three cases and the finger joints in two. The temporo-maxillary joint was affected in one case only. Of other authors, Gowers' series gives the relative frequency of the joints affected as follows: knee 45, hip 20, shoulder 11, tarsus 8, elbow 5, ankle 4; he does not mention the wrist. Collier and Adie give the frequency of the joints affected in the following order: hip, shoulder, elbow, ankle, small joints of the hands and feet and spine.

Occasionally, the arthropathies are multiple and are then as a rule symmetrical. A joint that has been damaged or has been subjected to undue strain is specially liable to be involved. It is probable that Edinger's "Aufbrauchtheorie"—fatigue or strain from excessive use—applies to many cases. Painlessness is a pathognomic sign of this form of joint lesion; exceptions to this rule are very few and even then the pain is usually slight or transient.

From the anatomical standpoint, an arthropathy shows the following stages: (1) Enlargement of the joint owing to increase of the synovial fluid together with thickening of the synovial membrane and stretching of the joint capsule. (2) Enlargement of the ends of the bones forming the joint with atrophy of the cartilages and occasionally some exostosis. (3) The effusion may spread to the surrounding tissues and produce a characteristic solid oedema. In appearance the skin over the joint is white and shining, the subcutaneous veins are unduly visible and full mobility of the joint is retained. (4) After several weeks or even months, absorption of the effusion and oedema tends to occur, with rarefaction and absorption of the articular surfaces of the bones. This atrophy is especially predominant in the ball and socket joints while in the hinge joints some hyperplasia usually occurs.

In rare instances of hip joint arthropathy, hyperplasia instead of atrophy may predominate and the clinical condition resembles that of a new growth in the region of the joint. Most frequently, however, the atrophic process in the hip joint leads to a rapid destruction of the head and neck of the femur with consequent displacement Fig. 1 (Plate 1).

In the case of the knee and elbow, hyperplasia usually occurs and especially in the head of the tibia Fig. 3 (Plate 1) and Fig. 4. As a rule, the lower end of the femur also shows hyperplasia but occasionally the femoral condyles may

atrophy. In any case, the usual result is abnormal mobility of the joint although ankylosis occurs in a very few cases.

Fig. 1 (Plate 1) illustrates an arthropathy of the right hip joint in a case of tabes dorsalis. There is an advanced degree of destruction of the joint and the head of the femur and the acetabulum have disappeared. There is a wide and shallow articulation between the neck of the femur and the upper part of the former acetabulum together with a new surface on the ilium. Several loose bodies are also seen within the enlarged joint capsule. This joint lesion developed in an ordinary case of tabes dorsalis of some four years' standing in 1919 when the patient was aged 44. Continued antisyphilitic treatment by means of repeated courses of novarsenobillon and bismuth injections eventually led to a negative Wassermann reaction in the blood and cerebrospinal fluid and the tabetic signs showed no advance. This patient, now aged 62, is still able to get about satisfactorily with the aid of an arm crutch.

FIG. 2.—Tabetic arthropathy of right knee joint.

Fig. 2 shows an arthropathy of the right knee in a case of tabes dorsalis. The patella is dislocated laterally from the femoral condyles which are both visible and palpable. The head of the tibia is also displaced laterally and is freely movable on the femur both laterally and anteroposteriorly. The lateral ligaments of the knee joint are very slack, allowing a considerable range of movement from the long axis of the limb.

The patient is a woman aged 65 with a history of ataxia for the past twelve years. The arthropathy of the knee has been present for two years. She shows unequal pupils, both of which are of the Argyll Robertson type, loss of deep sensation and vibration sense in the legs, absent arm and ankle-jerks as well as absent left knee-jerk. The Wassermann reaction is weakly positive in both blood and cerebrospinal fluid, and the latter contains two small lymphocytes per c.mm, total protein 0.05 per cent. and shows a Lange reaction of the luetic type.

Fig. 3 (Plate 1) is a skiagram of an arthropathy of the left knee-joint in a patient aged 63 with tabes dorsalis of 34 years' standing. The knee-joint had been affected for six years. Clinically, the joint was enormously enlarged, especially posteriorly. It showed full mobility and was quite painless. The skiagram shows lipping of the bones with massive formation of new bone (hyperplasia) affecting both femur and tibia. There are several loose bodies lying in the posterior part of the joints which show partial calcification.

FIG. 4.—Tabetic arthropathy of right wrist joint.

A tabetic arthropathy of the right wrist joint is shown in Fig. 4. The left wrist and the first and second metacarpo-phalangeal joints of the left hand were also involved. The onset of the joint lesions occurred at the age of 40 in the right wrist, gradually and without pain. About a year later the left wrist became affected and the metacarpo-phalangeal joints began to enlarge. These arthropathies were the first indication of tabes. The only neurological signs present were double Argyll Robertson pupil, slight inequality of the knee jerks, and ulnar and tendo

Achillis analgesia. Ten years later (1928), when the photographs were taken, the following is a description of the physical examination: Right wrist irregularly enlarged and much deformed. On the dorsum towards the ulnar side is a large swelling containing fluid with bony thickening beneath. A similar but smaller swelling is situated on the radial side of the joint. The enlargement is neither painful nor tender. Flexion and lateral movement at the joint are moderate and extension is limited. X-ray examination shows total disorganization of this joint, the carpus having almost entirely disappeared and the lower ends of the radius and ulna, as well as the bases of most of the metacarpals, being absorbed. Also, the metacarpal bones are dislocated ventrally on to the lower ends of the radius and ulna Fig. 5 (Plate 1). The left wrist is considerably swollen, especially towards the ulnar side, and deformed but is without pain or tenderness; flexion and extension is fairly good. The skiagram shows an early stage of Charcot arthropathy with little bone damage.

The metacarpo-phalangeal joints of the left index and middle fingers are also swollen and enlarged but are freely movable. X-ray examination shows signs "suggesting early Charcot joints." In the right axilla are several much enlarged glands which are neither painful nor tender. Epitrochlear glands are palpable in each arm, the left one being larger than the right.

Central nervous system: Right pupil larger than left; neither pupil reacts to light but reacts on accommodation-convergence. Other cranial nerves normal. Some ulnar and tendo Achillis analgesia. Arm-jerks and abdominal reflexes normal. Knee-jerks present, right slightly brisker than left; ankle-jerks normal; plantars flexor. No definite inco-ordination. Blood: Wassermann reaction positive. Cerebro-spinal fluid: sixteen lymphocytes per c.mm; globulin increased; Wassermann reaction positive; colloidal gold reaction shows curve of luetic type.

The influence of this patient's occupation—that of a painter—is almost certainly of importance in determining the arthropathies of the wrists. The alternate flexion and extension, continued over long periods, constitute the factors of strain and fatigue. When the right wrist became affected he began to paint with his left hand, with the result that this joint also became involved. Other features of interest in the case were the enlarged axillary and bilateral epitrochlear glands.

I have now had this patient under observation for nearly twenty years; the joints remain in much the same condition and he is still able to continue his painting. Originally, he was treated with novarsenobillon intravenously and bismuth intramuscularly (two courses per year of 12 injections of each substance) until the Wassermann reactions were negative in the blood and cerebrospinal fluid—which occurred in 1932. There has been no addition to the tabetic signs.

Syringomyelia.

In syringomyelia, the joint lesions are practically identical with those of tabes, but differences exist in their relative frequency and in the actual joints affected. Arthropathies occur in about 10 per cent. of cases of syringomyelia as compared with 4 per cent. of cases of tabes dorsalis. Also in tabes 75 per cent. of the arthropathies occur in the lower limbs, whereas in syringomyelia the reverse occurs—the upper extremities being affected in nearly 80 per cent. of cases. Schlesinger found that in 97 cases of syringomyelic arthropathy, 29 involved the shoulder joint, 24 the elbow and 10 the wrists.

Skiagrams of a case of syringomyelic arthropathy of both wrists are shown in Fig. 6 (Plate 1). This was a woman, aged 43, with pain and numbness in both hands and arms. Numbness of the left hand had begun five years previously

PLATE 1.

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FIG. 1.—Tabetic arthropathy of right hip joint.

FIG. 3.—Tabetic arthropathy of left knee joint (lateral view).

FIG. 5.—Tabetic arthropathy of right wrist joint (anteroposterior view).

FIG. 6.—Syringomyelic arthropathy of both wrists.

FIG. 7.—Syringomyelia, Morvan type.
Left hand.

The terminal phalanges of thumb and index finger
have disappeared.

FIG. 8.—Syringomyelia, Morvan type.
Skiagram of hand seen in Fig. 7.

FIG. 9.—Hypertrophic pulmonary arthropathy of both feet.

and had gradually involved the arm. Three years ago a deformity of the left wrist developed. The sensation of the right hand became affected eighteen months previously and three months later the wrist was deformed. Also she had had a recent painless burn of the left hand.

Physical examination showed dissociated anæsthesia—loss to pain, heat and cold—over the right hand extending to wrist, and over the left hand and arm and extending on to the adjacent part of the chest and root of neck (5th cervical to 3rd dorsal areas).

In the right arm there was weakness and atrophy of the intrinsic muscles of the hand, especially of the first dorsal interosseous and thenar muscles. Subluxation of the metacarpo-phalangeal joint of the thumb was present with bony prominence of the carpo-metacarpal joint towards the radial side. The wrist joint was deformed and thickened, the whole hand being deflected towards the ulnar side with limitation of movement. In the left arm, there was similar weakness and atrophy of all the intrinsic muscles of the hand with subluxation of the metacarpo-phalangeal joint of the thumb and thickening of carpo-metacarpal joint. The wrist joint was thickened and showed limited flexion and lateral movement towards the radial side. The whole hand was deflected towards the ulnar side. There was general wasting of the arm muscles, but no actual paresis. The arm-jerks were moderate on the right and unobtainable on the left side. The lower limbs were normal with brisk but equal knee and ankle-jerks and flexor plantar reflexes.

X-ray examination showed: Right wrist—Dislocation of metacarpal bone of thumb from trapezium and hyperextension of the first phalanx. Left wrist—Same condition of thumb as on right, together with considerable destruction of the carpal bones Fig. 6 (Plate 1). Scoliosis in dorsal region with C-shaped curve to left and some lumbar lordosis.

The patient was a milliner, working chiefly with wire in the making of hat-shapes, an occupation in which the wrists are much used. Edinger's "Aufbrauchtheorie", therefore, might also apply to this case.

Another trophic disturbance which occurs in syringomyelia is that of painless whitlows in the fingers and which may give rise to deep ulceration and even necrosis of the terminal phalanges (Morvan type of the disease).

Fig. 7 (Plate 2) illustrates the left hand of a case of the Morvan type of syringomyelia—a girl aged 25 years—and Fig. 8 (Plate 2) the skiagram of the same hand. The disease was of three years' standing. The terminal phalanges of the left thumb and index finger had practically disappeared; the remaining fingers showed flexor contracture (*main-en-griffe*). Dissociated anæsthesia was present in the left arm and adjacent part of the thorax as far as the mid-line (fifth cervical to second dorsal spinal segments) as well as exaggerated knee and ankle jerks, ankle clonus and bilateral extensor plantar reflexes.

Osteo-arthropathies.

The term "oste-arthropathy" is usually applied to the very rare vertebral lesions met with in *tabes dorsalis*. Clinically, the onset is insidious, occasionally sudden, with a gradually increasing spinal curvature which may be either a kyphosis or a scoliosis. In *tabes*, the lumbar region is usually affected. Pain is entirely absent or slight and locomotion is very little impaired.

In syringomyelia, some degree of spinal curvature is almost the rule and, in contrast to *tabes*, the cervico-dorsal region is usually affected. In some cases

the scoliosis may result from weakness of the spinal muscles, but the pathological investigations of Borchardt and Nalbandoff showed that in the majority of cases the condition was a true osteo-arthropathy.

The morbid anatomy of vertebral osteo-arthropathy consists in a central destruction of the vertebral body with peripheral hyperplasia about the various vertebral processes. The bone, both old and recent, is usually soft and spontaneous fracture or even collapse of a vertebra may occur.

A special type of osteo-arthropathy is known as the "tabetic foot" first described by Page in 1881 and later by Charcot in 1883. It usually occurs at a considerably earlier stage of tabes than the ordinary arthropathies. The foot acquires a swollen truncated appearance with disappearance of the arch and thickening of the malleoli. The soft tissues show a hard oedema. The metatarsus may be dislocated either above or below the proximal part of the foot and the calcaneum may collapse. The condition is painless and may be accompanied by perforating ulcer.

Hypertrophic Pulmonary Arthropathy.

The term "hypertrophic pulmonary osteo-arthropathy" was applied by P. Marie to a condition, first described by Bamberger, of enlargement of the joints of the fingers and toes and occasionally of the lower ends of the radius and ulna and of the tibia and fibula. The terminal phalanges are much enlarged and show curving both in the longitudinal and transverse directions; the nails are enlarged and may be curved over the ends of the phalanges. In practically all cases, the arthropathy is associated with chronic disease of the lungs, bronchi or pleura—new growths, chronic bronchitis, bronchiectasis, pulmonary tuberculosis and chronic empyema—and occasionally with congenital disease of the heart. It has also been met with in syphilitic disorders and is most frequent in adult males. Thayer collected 59 cases of which 43 showed preceding pulmonary disorders. Of the remainder, 3 were associated with syphilis, 3 with heart disease, 2 with chronic diarrhoea, one with spinal caries and in 3 cases no cause could be determined.

The exact causation is not known. It has been suggested by Marie that toxins of the pulmonary disease are absorbed into the circulation and have an irritant action on the bony and articular structures at the extremities causing an ossifying periostitis. Thorburn's suggestion of a benign and chronic tuberculous affection is most unlikely.

Fig. 9 (Plate 2) shows a skiagram of both feet in a case of hypertrophic pulmonary arthropathy complicating a new growth of the lung. The splayed appearance of the terminal phalanges is very characteristic.

Other Arthropathies.

Joint lesions in association with hemiplegia have been reported from time to time since Alison's first recorded case in 1847. The shoulder is almost invariably affected. The joint is usually painful, sometimes tender, shows crepitus and movement is very limited. The lesion probably results from purely local causes especially paralytic subluxation of the head of the humerus. The morbid anatomy consists in subacute synovitis with villous overgrowth and hyperæmia.

Arthropathies have also been reported in association with progressive muscular atrophy and amyotrophic lateral sclerosis (motor neurone degeneration), and I have recently seen a case of peroneal muscular atrophy (Charcot-Marie-Tooth type) with an arthropathy of both knees associated with almost total disappearance of both patellæ.