SYPHILITIC SPONDYLITIS

WITH A REPORT OF TWO CASES

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Syphilitic spondylitis is due to the *Treponema pallidum* invading the periosteum and bones of the vertebral column in tertiary syphilis, and should not be confused with Charcot's joints of the spine which may occur in tabes dorsalis.

Syphilitic spondylitis is well recognized but is rare, and there are few references to the condition in recent literature. This appears to be due partly to the difficulty in proving the diagnosis in the absence of histological or autopsy proof of the syphilitic aetiology. In 1914 Hunt reviewed the literature and stated that there were about a hundred well-authenticated cases recorded. Since that time other cases have been described, and in more recent years three cases were described by Freedman and Meschan (1943), and three cases by Sgalitzer (1941). Autopsies were performed upon two of Freedman and Meschan's cases.

The marked predilection of syphilitic spondylitis for the cervical vertebrae has been repeatedly stressed. Ziesché (1910) reviewed eighty-eight cases and found that the cervical region was affected in 70 per cent. of the cases. The cervical vertebrae were also affected in all six cases described by Freedman and Meschan (1943), and by Sgalitzer (1941). Abernethy (1931) recorded two cases of syphilitic spondylitis and the cervical spine was affected in both.

The symptoms and signs of syphilitic spondylitis do not differ essentially from those that accompany other forms of spondylitis or affections of the spine. There is localized pain at the site of the affected vertebrae, and the pain may be worse at night. The head is held stiffly and movements are avoided. The normal lordosis of the vertebral column may become straightened out. Local tenderness may be detected on palpation of the diseased vertebrae. Hunt (1914) was impressed by the frequency of neurological features in the extremities corresponding to the spinal region involved. Of one hundred cases analysed, neurological manifestations

were detected in twenty-six. They consisted of root pains, paraesthesiae, or localized paralysis. Hunt considered that this group of features was particularly frequent in the cervical region and could simulate pachymeningitis hypertrophica cervicalis.

Coexisting syphilitic lesions in other bones are common. In Freedman and Meschan's cases (1943) there were gummatous lesions involving the sternum, ribs, humerus, and skull. In Sgalitzer's cases (1941) the humerus, ilium, and skull were affected. Other syphilitic lesions may also be present, for example, syphilitic aortitis, gumma of the liver, and diseased cerebral vessels.

The blood Wassermann reaction is positive in syphilitic spondylitis. It is impossible to determine whether the reaction may ever be negative in syphilitic spondylitis, as the positive reaction is one of the clues to the correct diagnosis, and cases with a negative serology are not described.

Radiological Appearances

In many of these cases of syphilitic spondylitis there are certain common characteristics that allow the radiologist to suggest the correct diagnosis. The photographs published in the literature have a certain similarity which should lead to the diagnosis if the entity is kept in mind.

There is usually evidence of destruction of the vertebral bodies, particularly in the anterior part of the bodies. In addition there is a marked tendency to the formation of osteosclerosis and hyperostoses, and the vertebrae ultimately become surrounded by more or less marked spur formation. These productive changes usually predominate over the destructive changes, and collapse of the vertebral bodies is extremely rare, due to the pronounced sclerosis.

The lesion in the cervical spine is usually extensive, but only one vertebral body may be affected, or two or three adjacent vertebrae. The involvement

of the dorsal and lumbar regions yields a less suggestive radiological appearance than the diffuse type of cervical involvement.

There is usually calcification in the anterior and lateral ligaments, leading eventually to complete ankylosis. The intervertebral space is usually well preserved, as in Sgalitzer's three cases (1941). However, the intervertebral disks may rarely become destroyed and the intervertebral spaces become reduced in width. The vertebral endplates may be smooth or moth eaten. Sgalitzer (1941) states that paravertebral abscess is never found in syphilitic spondylitis, but Cofield and Little (1925) described a case with cervical involvement and a retropharyngeal abscess.

Treatment

Antispecific therapy caused rapid symptomatic improvement in Gill and Frazer's case of syphilitic lumbar spondylitis (1933). Abernethy has described two cases of cervical involvement (1931) which improved radiologically after antispecific therapy. Symptomatic improvement, and sometimes radiological improvement, was also noted by Freedman and Meschan (1943) and by Sgalitzer (1941) in their cases. In other cases the radiological appearances have remained unaltered.

Diagnosis

If it is remembered that syphilis may be a cause of spondylitis, then the diagnosis should not cause much difficulty. The condition has to be differentiated from tuberculous spondylitis, pyogenic spondylitis, other forms of infective spondylitis, osteoarthritis, and ankylosing spondylitis. The predilection for the cervical vertebrae is an important differentiating point in the diagnosis, and the radiological appearances are striking and suggestive. The Wassermann reaction is positive. There are often associated syphilitic lesions in other bones or in other viscera. Finally, there is relief of symptoms after antispecific therapy, and sometimes there is corresponding radiological improvement.

In view of the comparative rarity of the condition the following two cases are reported.

Case Reports

Case 1.—A European, aged 55, was admitted to Groote Schuur Hospital, Cape Town, on June 14, 1946. He complained of severe pain in the back of his neck. The pain had been more or less constantly present for the past three years. It was worse at night, and was aggravated by movement of the neck. The pain sometimes radiated down the medial aspect of both arms, and these sites felt numb. The pain in the neck was accompanied by progressive stiffness. The left hand had

slowly become weak during the two weeks before his admission.

EXAMINATION.—The patient looked well. He held his neck very stiffly and was unable to turn his head to either side. The normal lordosis of the cervical spine was lost, and the spine was tender on palpation. There was also marked rigidity of the dorsal and lumbar regions of the vertebral column, but there was no associated tenderness. The peripheral joints were normal.

There was marked wasting of the small muscles of the left hand, associated with some weakness of the interossei and flexor muscles of the fingers. Sensory examination revealed anaesthesia and hypo-algesia of the skin of the medial two fingers and medial half of the left hand. The reflexes were normal. There were no other abnormal neurological findings.

The cardiovascular, respiratory, and alimentary systems were normal. The blood pressure was 160/100 mm. Hg. The urine was chemically and microscopically normal. The blood count was normal.

The blood Wassermann reaction was positive (this was confirmed a second time).

The cerebrospinal fluid was clear and colourless. There was no block on lumbar puncture. Protein was 50 mg. per 100 c.cm., globulin 1 plus. No cells were seen in 1 c.mm. of fluid, and the Wassermann reaction was negative.

The Widal and the Brucellin agglutination reactions were negative.

Radiographs.—There was marked spondylitis of the cervical vertebrae (Fig. 1). The normal trabeculation of the vertebral bodies was replaced by small areas of translucency associated with much spur formation and osteosclerosis. There was bony fusion between the vertebral bodies, with calcification of the surrounding ligaments.

The thoracic region was not so severely affected, but most of the vertebral bodies showed hypertrophic spur formation with some calcification of the lower dorsal interspinous ligaments.

The lumbar spine was also involved. It showed marked "lipping", osteophytic outgrowths, and calcification of the anterior ligament (Fig. 2). The sacroiliac joints were normal (Fig. 3).

The features of spondylitis, especially the marked affection of the cervical vertebrae, suggested the diagnosis of syphilitic spondylitis.

No syphilitic osseous lesions were present in skull, tibiae, humeri, femora, hands, ulnae, radii, or feet.

TREATMENT AND PROGRESS.—A course of deep x-ray therapy was given to the vertebral column from July 1, 1946, until Aug. 8, 1946, as it was initially considered that the diagnosis was an atypical form of Marie-Strumpell's ankylosing spondylitis. The patient was in bed while the treatment was given. By the end of treatment there was a very slight decrease in the amount of pain, but the patient was still experiencing considerable discomfort in the neck and in the arms.

Antispecific therapy was begun on Aug. 7, 1946. He received potassium iodide orally and was given 6 intravenous injections of neoarsphenamine at weekly

intervals. Bismuth oxychloride ("bisoxyl") was administered bi-weekly intramuscularly for five weeks. After his discharge from hospital, two further courses of "bisoxyl" were given.

He was re-examined as an out patient in August, 1947, and in November, 1947. His neck was still stiff but was quite painless and there was no tenderness on palpation or on attempting movements. There was no pain in the arms. The left hand felt less numb and, objectively, the degree of sensory loss had decreased in comparison with the findings on admission. The power in the left hand had returned to normal. These improvements began a few weeks after the institution of the antispasmodic therapy. The blood Wassermann reaction was still positive. The radiographs of the spine were repeated but no changes could be detected in the features seen in the original films.

Case 2.—The patient, a non-European, aged 28, was admitted to Groote Schuur Hospital, Cape Town, on Feb. 10, 1947. His neck was painful and stiff. The symptoms had begun eight years previously and were becoming progressively worse. His head was slowly becoming depressed on to his chest and he was unable to open his mouth completely. There had been occasional pains in the dorsal and lumbar regions of the spine for the past year. For the past three years there had been constant severe pains in the right knee and in the right upper arm, and the left clavicle had become painful for the past few months. He began coughing a few days before admission to hospital.

EXAMINATION.—The patient was pyrexial with a temperature of 101°F. , and looked very ill and toxic. He was very dyspnoeic, and there was pitting oedema over the sacrum. The neck was rigid, and the chin was practically in contact with the anterior chest wall. There was barely any movement of the neck in any direction. The cervical spines were tender on palpation. The lumbar and dorsal spines were also rigid and tender, but to a much lesser degree than the neck.

The right tibia, the right humerus, and the left clavicle were visibly and palpably thickened. They were very tender on palpation. There was a large effusion in the right knee, with wasting of the right quadriceps muscles. The movements at the right knee joint were limited by pain and by swelling. There was limitation of movement at the right elbow. The skin overlying the right tibia and right humerus was thickened. There was a fracture in the proximal third of the right humerus, with abnormal mobility.

The blood pressure was 115/70 mm. Hg. On auscultation the second sound at the aortic area was loud and ringing and had a hollow character.

There were bilateral basal crepitations in the lungs.

The abdomen was distended. The liver was enlarged 4 fingerbreadths below the right costal margin. It was firm, rounded, and tender.

No abnormalities were detected in the central nervous system.

The specific gravity of the urine was 1010. A heavy albuminuria was present. Microscopy showed numerous pus cells, several red blood cells, and a moderate number of granular and hyaline casts.

The blood count was: erythrocytes, 3,500,000 per c.mm. of blood; leucocytes: 11,200 per c.mm. There were 63 per cent. polymorphonuclear leucocytes; 34 per cent. lymphocytes; 2 per cent. monocytes; and 1 per cent. eosinophils.

The blood Wassermann, Kahn, Berger, and Rappaport reactions were all positive.

The cerebrospinal fluid was clear and colourless. There was no block on lumbar puncture. There was 35 mg. per 100 c.cm. of protein, no globulin, and 1 lymphocyte per c.mm. The Wassermann reaction was negative.

Radiographs.—There was marked osteosclerosis of the cervical vertebrae, with bony fusion of several of the bodies (Fig. 4). In the lumbar region there was slight erosion of the adjacent bodies of the fourth and fifth vertebrae, with surrounding bony sclerosis. There was slight calcification of the anterior longitudinal and interspinous ligaments. The sacro-iliac joints were normal (Fig. 5 and 6).

The right tibia and right humerus were diffusely affected by syphilitic osteitis. There was osteosclerosis with many areas of erosion (Fig. 7 and 8).

A radiograph of chest and ribs showed the aorta to be dilated; there was a large area of pneumonitis in the midzone of the right lung, and a smaller area of pneumonitis at the left base (Fig. 10). Destructive changes were noted in the left clavicle (Fig. 10).

Serum calcium was 9.6 mg. per 100 c.cm. of blood, serum inorganic phosphorus 4.6 mg. per 100 c.cm., serum alkaline phosphatase 15.1 Bodansky units, serum albumin 2.5 g., globulin 4.7 g., and cholesterol 610 mg. per 100 c.cm.

Bence-Jones proteinuria was present in the urine.

Van Slyke's urea clearance test gave the following results: blood urea 55 mg. per 100 c.cm.; first period urea clearance 22 per cent. of mean normal; second period 24 per cent. of mean normal.

The sedimentation rate was 105 mm. in 1 hour (Westergren).

Sternal puncture showed the bone marrow to be normal.

Of the Congo red present in the serum after 4 minutes, 65 per cent. remained after 1 hour.

No tubercle bacilli were found in the sputum. Gram-positive cocci resembling pneumococci were grown on blood agar.

TREATMENT AND PROGRESS.—Penicillin, 40,000 units, was administered three-hourly, by intramuscular injection, from Feb. 24, 1947, for seven days. A total amount of 2 million units was given. A second course of penicillin was administered from May 1, 1947, for eleven days. He received 50,000 units three-hourly until a total of 4,256,000 units were given.

"Bisoxyl," 2 c.cm., was given intramuscularly bi-weekly for five weeks. Potassium iodide was administered orally, 10 gr. three times a day, for approximately three months.

Many changes developed after the commencement of this therapy. The area of pneumonitis in the right lung decreased rapidly and was almost absent three weeks later (Fig. 11, March 3, 1947). A small effusion

FIG. 1.—Case 1: cervical vertebrae, showing loss of normal curvature, small areas of erosion, marked osteosclerosis, and bony fusion between some vertebrae.

FIG. 2.—Case 1: lateral lumbar vertebrae, showing lipping, osteophytic outgrowths, and calcification of ligaments.

FIG. 3.—Case 1: lumbar vertebrae, showing marked osteophytic outgrowths and normal sacro-iliac joints.

FIG. 4.—Case 2: radiograph of cervical vertebrae, taken Feb. 11, 1947, showing marked osteosclerosis with bony fusion of several vertebrae.

FIG. 5.—Case 2: radiograph of lumbar vertebrae, taken Feb. 11, 1947, showing slight erosion of adjacent bodies of fourth and fifth vertebrae, marked liping, and early calcification of anterior longitudinal ligament.

FIG. 6.—Case 2: radiograph of lumbar vertebrae, taken Feb. 11, 1947, showing bony sclerosis surrounding the fourth and fifth vertebrae, and early calcification of some interspinous ligaments.

FIG. 7.—Case 2: radiograph of right tibia, taken Feb. 11, 1947, showing diffuse osteitis with areas of translucency and marked osteosclerosis.

FIG. 8.—Case 2: radiograph of right humerus, taken Feb. 11, 1947, showing multiple areas of erosion with some osteosclerosis, and fracture of the shaft in the upper third of humerus.

FIG. 9.—Case 2: radiograph of right humerus, taken April 28, 1947 (after antisyphilitic therapy). There is much more osteosclerosis and less bony erosion as compared with the previous radiograph (Fig. 8).

FIG. 10.—Case 2: chest radiograph taken Feb. 11, 1947, showing areas of erosion in the left clavicle. The aorta is dilated. A large area of pneumonitis can be seen in the mid-zone on the right side, and a small area of pneumonitis at the left base.

FIG. 11.—Case 2: chest radiograph taken on March 3, 1947. The area of pneumonitis present on the right side has decreased considerably. There is a left-sided pleural effusion.

FIG. 12.—Case 2: radiograph of chest taken on May 15, 1947. The lungs are completely clear. Thickened pleura can be seen in the left costophrenic region. The left clavicle is almost normal. The aorta remains dilated.

developed at the left base (Fig. 11). A later chest radiograph taken on May 15, 1947, showed that the lungs were completely clear, with thickened pleura at the left base. The aorta remained dilated (Fig. 12).

There was an initial exacerbation of the features of congestive cardiac failure, and the dyspnoea and the oedema increased in extent. Then the patient's general condition improved steadily and progressively and the features of congestive cardiac failure decreased and disappeared. He was discharged from hospital in June, 1947.

The vertebrae and the affected bones were again x-rayed on April 28, 1947. Regeneration of bone was noted in the right tibia and in the right humerus (Fig. 9), and the left clavicle had become almost normal in appearance (Fig. 12). The cervical spine remained unchanged.

Clinically, the pain and tenderness in the spine and in the affected bones decreased considerably after the institution of the antisyphilitic therapy. There was no alteration in the rigidity of the spine.

Albuminuria was constantly present during the patient's stay in hospital, but the Bence-Jones proteinuria disappeared. The specific gravity of the urine varied between 1004 and 1012 during the ten weeks' observation. The blood urea remained raised. There was impairment in the water concentration and water dilution tests. The serum proteins returned to normal. The intravenous uroselectan examination revealed no local renal abnormality.

The patient was seen again as an out patient in August and in October, 1947. The right knee was still painful and swollen and he was unable to walk properly. Radiological examination revealed that this was due to backward subluxation of the tibia at the knee joint. There were no further radiological changes in the vertebrae or in the tibia. The blood Wassermann, Berger, and Rappaport reactions were still positive.

Discussion

Although there is no biopsy or pathological proof of syphilitic spondylitis in these two cases, the diagnosis is almost certainly correct in view of the characteristic clinical, radiological, and serological features.

The first case presented with the characteristic complaints of pain and stiffness of the neck. In addition there were neurological manifestations similar to those described by Hunt (1914)—root pains in the arms, with sensory and motor disturbances. The radiological appearances of the cervical vertebrae were strikingly similar to those seen in the photographs of the cases recorded by Freedman and Meschan (1943). The radiological appearances in the lumbar and thoracic regions were much less suggestive, and resembled changes seen in ankylosing spondylitis and in osteo-arthritis. The sacro-iliac joints were perfectly normal. There were no other syphilitic osseous lesions in spite of

extensive radiological investigations. The blood Wassermann reaction was positive. After antisyphilitic therapy there was a rapid and progressive improvement in the clinical features of the cervical spondylitis. There were, however, no changes in the radiological appearances.

The second case had severe involvement of the vertebral column. There was marked pain and tenderness in the spine, especially in the cervical region. The radiological changes were similar to those encountered in other cases of syphilitic spondylitis and were maximal in the cervical vertebrae. Syphilitic lesions were present involving the right tibia, right humerus, left clavicle, and left tenth rib. The second sound at the aortic area suggested the probability of dilatation of the aorta, and this was confirmed radiologically. The blood Wassermann reaction was positive. After antisyphilitic therapy there was a decided and marked improvement in the clinical features and the affected vertebrae became less painful and less tender. There was no improvement in the degree of rigidity of the spine. There was regeneration of bone in the right humerus (Fig. 9) and in the left clavicle (Fig. 12). The vertebrae remained unaltered. The right knee joint remained painful and swollen on account of a subluxation of the right tibia.

The presence of Bence-Jones proteinuria in the second case is interesting. Bence-Jones proteinuria is a well-known feature in multiple myelomatosis but may also be present in other conditions with widespread bone and bone-marrow involvement. It is sometimes present, for example, in leukaemia and in secondary carcinomatosis of bone. Bence-Jones proteinuria has also been noted in Boeck's Sarcoidosis (Harrell, 1940; Horwitz, 1948), due probably to the widespread involvement of the bones and bone marrow by sarcoid tissue. Its presence in a case of very widespread and gross syphilitic osteitis is thus not inexplicable.

The nature of the renal lesion remained unelucidated. Amyloidosis may occur as a complication of extensive generalized visceral syphilis, and this seemed to be the best clinical suggestion. Fishberg (1939) describes a type of amyloid disease of the kidneys which clinically and chemically resembles cases of chronic nephritis. It is, therefore, possible that the persistent albuminuria, azotaemia, and impaired renal function tests are an indication of amyloidosis of the kidneys. However, this could not be confirmed by the Congo-red test.

Pneumonitis was present in the second case on admission. It decreased markedly in size after three weeks. The clinical and radiological features were suggestive of a bacterial pneumonia resolving with penicillin therapy. An alternative possibility,

which is less likely and which remained unproved, is that it was a rare example of gummatous involvement of the lung which responded to the penicillin and to the other forms of antisppecific therapy employed.

There was no clinical evidence of aortic regurgitation in the second case, and it is uncertain what precipitated the congestive cardiac failure. The exacerbation of the congestive features after the commencement of antisppecific therapy raised the question of a Herxheimer response.

Summary

1. Two cases of syphilitic spondylitis are presented. The diagnoses were made on the characteristic clinical and radiological features.

2. The cervical region of the spine was the site mainly affected in both cases.

3. There were associated syphilitic osseous and visceral lesions in the second case.

4. The blood Wassermann reaction was positive in both cases.

5. There was a prompt response clinically in both cases to antisppecific therapy. Pain and tenderness of affected bones decreased considerably. There was no change in the radiological appearances of the vertebrae.

6. Bence-Jones proteinuria was noted in the second case.

7. The diagnosis of syphilitic spondylitis may be made readily from the clinical and radiological manifestations if the condition is considered in the differential diagnosis of all cases of spondylitis. Response to antisppecific therapy affords confirmatory evidence.

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Spondylite Syphilitique: Avec Compte-rendu de deux Observations

RÉSUMÉ

1. L'auteur a présenté deux cas de spondylite syphilitique. Les diagnostics ont été établis d'après les caractères cliniques et radiologiques typiques.

2. C'est le segment cervical de la colonne vertébrale qui était particulièrement atteint dans les deux cas.

3. Le second malade présentait également des lésions syphilitiques osseuses et viscérales.

4. La réaction de Wassermann dans le sang était positive dans les deux cas.

5. Dans les deux cas le traitement antisypilitique a donné un résultat clinique rapide. La douleur et l'hypersensibilité dans les os atteints diminuèrent considérablement, mais l'aspect radiologique des vertèbres ne s'est pas modifié.

6. On n'a pas observé la présence d'albumine de Bence-Jones dans l'urine d'aucun de ces malades.

7. Le diagnostic de spondylite syphilitique peut être établi aisément à partir des manifestations cliniques et radiologiques si l'on pense à la possibilité de la syphilis parmi les causes de spondylite. Les résultats du traitement antisypilitique confirment le diagnostic.