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CHRONIC ADHESIVE SPINAL MENINGITIS ASSOCIATED WITH LUMBAR NAEVUS AND DIMPLE

BY

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Chronic meningitis confined to the spinal theca has been described frequently during the last fifty years; it has been known by various synonyms—arachnitis adhesiva circumscripta, arachnoidite cloisonnée, arachnoid cysts, circumscribed serous spinal meningitis, and chronic spinal meningitis.

The aetiology of the condition is obscure in most cases: Ford (1945) states that it may follow various types of meningitis, spinal analgesia, and, in the opinion of most authorities, trauma to the spine. The reason for this obscurity seems to be that chronic spinal meningitis is usually the end-result of a previously unrecognized inflammatory process, and there is a clearly defined picture in the later stages only, either clinically or at operation or necropsy.

The subject has been reviewed by Horsley (1909), Mauss and Krüger (1918), Elkington (1936), and Rocovich (1947); but these authors do not mention congenital dermal sinuses as an important cause of chronic spinal meningitis. Mallory (1892) described coccygeal sinuses and cysts without meningeal or spinal cord involvement, and discussed their pathogenesis, with particular reference to abnormal foetal development.

Moise (1926) states that the sacro-lumbar region is a common site for developmental anomalies, among which are included congenital dimples, sinuses, cysts, and tumours. These cysts and tumours may be dermoid or epidermoid (List, 1941). There is only occasionally a connexion between the spinal canal and the skin surface. Also there may be instances of spina bifida, with all gradations from an unnoticed spina bifida occulta with no evidence of an external defect to a fusion of the spinal cord with the integument. The cases showing a connexion between the spinal canal and the exterior do not as a rule survive infancy. Moise was the first to report a case of meningitis (*Staphylococcus albus*) secondary to a

congenital dermal sinus; there were no symptoms until the age of 18; there was spina bifida and a direct connexion between the skin and the spinal cord, and the patient recovered after laminectomy with drainage.

Ripley and Thompson (1928) described a case of *Staph. albus* meningitis in which a pilonidal sinus was the route of infection. Walker and Bucy (1934) reported seven cases in which a congenital dermal sinus, appearing externally as a small dimple in the midline of the back, had become infected (two with *Staph. albus*); all the cases but one were associated with spina bifida occulta.

Other cases have been recorded by Stammers (1938), Boldrey and Elvidge (1939), List (1941), and Tizard (1950). O'Connell's case (1942), that of a child aged 4, had a lumbo-sacral port-wine stain and a small central pit extruding sebaceous material on pressure; this was complicated by recurring attacks of meningitis (coliform organisms) from the age of 8 months; the sinus was traced through a lumbar spina bifida to a large intradural abscess lying among the roots of the cauda equina.

Intramedullary abscess of the spinal cord complicating congenital dermal sinuses has been recorded by Kooistra (1942) and Shenkin and others (1944). Mount (1949) has been the first to describe a case with intracranial abscess as a similar complication.

The following case is reported because the causation of the condition and the route of infection raise some interesting problems, and because in several features it resembles previously reported cases, especially that of O'Connell.

Case Report

A girl aged 9 had backache for two weeks in 1947. In February, 1948, she again had severe backache, low in position, with delirium and pyrexia for one week, and was admitted to a hospital near her home.

Her own and her family history revealed nothing relevant. She was pale and thin; the posterior cervical and inguinal lymph nodes were enlarged; and flexion of the left hip with straight leg raising was limited. The white cells numbered 5,000 per c.mm. (neutrophils 39%), the E.S.R. (Wintrobe) was 7 mm. in one hour, and the Mantoux test (1 in 1,000) was negative. Her temperature and pulse became normal within three weeks; she received only sedatives.

In March, 1948, recrudescence of pain in the back and fever led to her readmission to the local hospital. There was limited mobility of the lumbar spine with spasm of the erector spinae. Swelling of joints, nodules, and skin rashes were absent; the white cells numbered 9,000 per c.mm. (neutrophils 50%), the E.S.R. (Wintrobe) was 19 mm. in one hour, the blood Wassermann reaction was negative, and blood culture was negative. Radiography showed spina bifida of the eleventh thoracic, fourth and fifth lumbar, and first sacral vertebrae. A tentative diagnosis of rheumatic fever was then made. She was kept flat in bed for four months and was in hospital for eight months; she was given salicylates and phenobarbitone.

In December, 1948, pain in the back and right lower limb and pyrexia reappeared.

In March, 1949, she was admitted to Great Ormond Street. She was miserable and wasted generally; there was a circular brown pigmented naevus about 2.5 cm. in diameter with a shallow dimple or pit about 3 mm. in diameter over the lumbar spine. Walking was painful and difficult. Sitting with her knees fully flexed was the position she adopted in bed. There was no neck rigidity. In the upper and lower limbs muscular development, tone, and power were poor and impaired equally with the rest of the body and on the two sides. There was no localized wasting; co-ordination was normal. The biceps, supinator, and triceps jerks were absent on the right and present on the left. The abdominal reflexes were present and equal. There was still gross limitation of straight leg raising—to 45 degrees. The right knee-jerk was exaggerated and the left

absent. Both ankle-jerks were absent and the plantar responses were both equivocal. There were no sensory changes or evidence of a sensory level.

Special Investigations

Radiography revealed no abnormality in the skull, lungs, heart outline, and mediastinum.

On March 19 the white cells numbered 12,650 (neutrophils 60%); the E.S.R. was 4 mm. in one hour; agglutinations against *Salmonella typhi*, *Salm. paratyphi* A and B, all with O and H antigens, and *Brucella abortus* were all negative. A catheter specimen of urine was normal.

On March 21 a lumbar puncture produced a dry tap from 3/4 and 4/5 lumbar spaces, but a faintly cloudy colourless fluid with clot from the 2/3 space; no resting pressure could be read because of crying. Examination of the fluid showed: 42 cells per c.mm. (98% monocytes); no organisms seen; culture sterile; protein, 275 mg. per 100 ml.; sugar, 50 mg. per 100 ml.; insufficient for chlorides; Wassermann reaction negative.

On March 25 a lumbar puncture in all the above spaces resulted in a dry tap. Cisternal puncture gave a slightly turbid colourless fluid with a clot: cells 25 per c.mm.; culture sterile; protein, 150 mg. per 100 ml.; sugar, 56 mg. per 100 ml.; chlorides, 735 mg. per 100 ml. On March 30 a post-nasal swab (for meningococci) contained a few colonies of *Staph. albus*.

The provisional diagnosis at this time was chronic adhesive arachnoiditis following old meningitis, possibly meningococcal. The patient was transferred to the Atkinson Morley Hospital under Mr. Wylie McKissock for further investigation.

On April 11 a cisternal encephalogram revealed good and normal filling of the ventricular system. Lumbar myelography (1 ml. of "myodil" introduced into the cisterna magna) showed poor filling of the theca, and the appearances suggested an arachnoiditis with block at the level of the second lumbar vertebra (Figs. 1 and 2). With the head down, the theca was filled with air and the existence of spinal block was confirmed.

Mr. Wylie McKissock performed laminectomy of the second, third, and fourth lumbar vertebrae on April 19. The theca was

FIG. 2.

tense and bulging and unusually firm to the touch. The dura was found to be particularly thick and adherent to the underlying granulation tissue. The whole subdural space was filled with a mixture of granulation tissue, pus, and fibrous adhesions, in which most of the nerve roots were embedded. The dura was left open and the wound closed in layers over it. No sinus between the skin and spinal theca was found. A light growth of coliform organisms was obtained from a swab at operation.

In the immediate post-operative period penicillin was given four-hourly for three days (total 440,000 units) and streptomycin 4 g. over five days, both by intramuscular injection, and sulphadimidine by mouth for seven days (total 18 g.).

Post-operative Progress

The wound healed by first intention. Full movement in both lower limbs had returned 10 days after operation. The patient was free from pain in the back and legs. There was only occasional urinary incontinence, and the spine remained rather rigid. During the first few weeks the tendon-jerks in the right upper limb returned and equalled those on the left; the left knee-jerk returned and was brisk, the right knee-jerk remained exaggerated; both ankle-jerks could just be elicited; plantar responses were flexor; Rombergism was absent; her gait was normal except for some evidence of apprehension.

From May to September, 1949, she was symptomless except for a discharging sinus near the laminectomy scar. Her nutrition and muscular development improved greatly and she had even learned to swim.

On her fourth admission to hospital on September 26 (Atkinson Morley Hospital), for exploration and excision of the residual sinus, there was very slight weakness of the muscles moving the left knee- and ankle-joints. There was no wasting, and the muscular tone was good. All tendon-jerks were brisk, the right knee-jerk being slightly greater than the left; the plantar responses were flexor. Spinal movements were good and painless; there was no tenderness on pressure over the spine; the gait was normal. Pus was discharging from two small sinuses

FIG. 1

in the laminectomy wound and from the small pit in the naevus. Radiography of the lumbar spine showed no myodil residues and no evidence of bone infection. *Staph. aureus* and *Neisseria catarrhalis* were cultured from the sinuses.

On October 4 the sinus was found to extend as deep as the anterior surface of the erector spinae. The wound healed, but discharged again intermittently. Swabs were repeatedly sterile on culture.

On January 24, 1950, the original wound, together with the naevus, was completely excised down to the block of fibrous tissue which covered the open theca; no true granulation tissue was discovered at any point.

During the next six months the sinus in the operation scar discharged intermittently, but the patient's general health was excellent; she remained symptomless and free from pain in the back and lower limbs. In July, 1950, there was a small discharging sinus in the lumbar scar; spinal movements were full except for slight limitation of flexion; straight leg raising was limited to 80 degrees (right and left) by pain in the hamstrings; right hip flexion was slightly weak; the right knee-jerk was very brisk, the left was normal; the right ankle-jerk was absent, the left was present on reinforcement; the plantar responses were flexor; sensation and gait were normal.

Discussion

This case has several features of interest, particularly from the aspect of diagnosis and the mode of infection.

Diagnosis of Site of Lesion.—Negative evidence included absence of a history of trauma and of radiographic signs of vertebral disease, and therefore indicated that the lesion was intradural: absence of involvement of the long tracts excluded an intramedullary lesion. Positive evidence depended chiefly on the presence of bilateral and asymmetrical changes in the tendon reflexes in the upper as well as the lower limbs, showing that spinal nerve roots in the cervical region as well as the cauda equina were involved; also, the existence of spinal block was demonstrated after lumbar puncture and contrast myelography. Spinal block was complete at the level of the second lumbar vertebra and was partial above the lumbar region.

Diagnosis of Nature of Lesion.—The long history of recurring bouts of pyrexia and back pain over more than twelve months strongly suggested an inflammatory process. An equally extensive intradural neoplastic lesion was only likely to be due to "seedling" metastases derived by gravity from an intracranial tumour, but there was no clinical or radiographic evidence of such a space-occupying lesion. Negative serological tests excluded syphilis as a cause.

The appearances at myelography were those of arachnoiditis, but all authorities agree that a pre-operative diagnosis of chronic adhesive spinal meningitis is seldom made, and that clinical distinction from spinal tumour is difficult. Mackay (1939) states that, clinically, such cases differ from those of extramedullary spinal tumour in the evidence of a diffuse disturbance of the spinal cord and roots, such as widely scattered paraesthesiae and pains, irregular and inconstant sensory disturbances, muscular atrophy and large fascicular twitches, painful muscular spasms, a mixture of exaggerated and diminished tendon reflexes, and arrest of iodized oil at multiple levels. There is absence of agreement concerning sensory disturbances, since Beaumont (1950) states that in spinal arachnoiditis the posterior roots are much more likely to be affected than the anterior. It is not clear why the first operation was followed by a return to normal of the upper-limb tendon-jerks; it has been suggested that this was an effect of chemotherapy. There remains the possibility that a dermoid is still present among the roots of the cauda equina.

Mode of Infection.—This is the chief problem of the case. In considering the possible routes by which infection reached the spinal meninges, there was no evidence of septicaemia or a primary infective focus elsewhere, at or before the onset of symptoms. There was no history of a lumbar puncture for diagnosis or spinal analgesia (Kremer, 1945). The presence together of a pigmented naevus with a dimple in the midline over the lumbar spine, spina bifida occulta of the eleventh thoracic, fourth and fifth lumbar, and first sacral vertebrae, and chronic adhesive arachnoiditis suggest that infection may have reached the meninges via a sinus, though such a sinus was not demonstrated at operation. Some support for this claim is provided by the close resemblance to the case described by O'Connell (1942).

I am indebted to Dr. Bernard Schlesinger and Mr. Wylie McKissock for permission to record the case that was under their care. I should like to thank Dr. Schlesinger for his interest and helpful criticism of this report, and Dr. E. J. Glynn for his case notes.

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The following statement about the prophylactic administration of penicillin in certain cases has been approved by the American Council on Rheumatic Fever of the American Heart Association: "Following dental extractions and removal of tonsils and adenoids, bacteria are frequently present in the blood stream for short periods of time. In rheumatic individuals or in patients with congenital heart disease these bacteria may lodge in the heart valves and cause bacterial endocarditis. Although a variety of bacteria cause this disease, the majority of cases are due to alpha streptococci (*Streptococcus viridans*). Alpha streptococci are usually resistant to sulphonamide drugs. Penicillin is, therefore, recommended for prophylaxis as follows. (1) Except in emergencies operative procedures in rheumatic individuals should be deferred until there is no clinical evidence of rheumatic activity and laboratory tests indicate that the rheumatic process is subsiding. (2) Patients should be free from upper respiratory infection. (3) Minimum dosage of penicillin: (a) 300,000 units of aqueous penicillin intramuscularly 30-60 minutes before extraction or operation. (b) 300,000 units of procaine penicillin in oil injected intramuscularly at the same time in a different site. Penicillin prophylaxis is not necessary for the extraction of deciduous incisors or bicusps unless infection of the gum is present. It should be used in the extraction of deciduous molars and all permanent teeth, and for tonsillectomy and adenoidectomy. In most instances it is best to extract one tooth at a time; multiple extractions should be avoided. In cases of extensive gum infection or severe root infections (apical abscesses) it is advisable to give several doses of penicillin starting the day before operation and continuing one or two days thereafter. Women with rheumatic or congenital heart disease should receive penicillin prophylaxis at the time of delivery. It is also recommended for patients requiring gastro-intestinal surgery."