Case 8.—Girl aged 3. Pain, synovial swelling, and effusion in left knee in July, 1949. Warm, tender, moderate swelling and effusion, and almost full movement, at start of treatment. X-rays showed no osseous focus and normal joint space. Biopsy showed inguinal glands, superficial and deep, to be positive for tuberculosis on December 9, 1949. Focus right hilum in July, 1949. *Treatment*: 12 thrice-weekly injections of 0.25 g. of streptomycin (December, 1949, to January, 1950). Two months' traction in Thomas splint. *Result* (19 months after treatment): Knee cool, no swelling; Full movement. X-ray film as before. *Comment*: An early case; proved infection overcome with local streptomycin therapy.

Case 9.—Boy aged 16. Pain and swelling of right knee in December, 1947; then treated conservatively for two years. At time of admission moderate synovial swelling; movement 20°. X-ray films showed coarse trabeculation, slightly reduced joint space, and irregularity of posterior part of femoral condyles. Biopsy showed inguinal gland (January 18, 1948) to be positive for tuberculosis. *Treatment*: 12 thrice-weekly injections of 1 g. of streptomycin (December, 1949, to January, 1950). Two months' immobilization in Thomas splint. *Result* (19 months after treatment): Moderate swelling persisted; 40° movement. January, 1951: Charnley's arthrodesis. X-ray film showed bony fusion. *Comment*: Local streptomycin failed, but moderately advanced erosion of joint surfaces was present at start of treatment.

Case 10.—Male, aged 44. Pain and synovial swelling right knee since March, 1949, x-ray film showing rarefaction. Pulmonary tuberculosis, March, 1949; positive sputum. *Treatment*: 12 thrice-weekly injections of 1 g. of streptomycin (April to May, 1950) plus 2 g. of P.A.S. intra-articularly thrice-weekly for same period. Eight months in walking calliper. *Result* (15 months after treatment): Pain and synovial swelling persisted. Charnley's arthrodesis, February, 1951. Synovial membrane at time of operation positive for tuberculosis. February 12, 1951, x-ray film showed bony fusion. *Comment*: Local streptomycin failed.

Discussion

The affected knee is indistinguishable from normal in Cases 1 and 8. Cases 3 and 4 are progressing well, have 140 and 80 degrees of movement and no pain, wear no support, and are back at work. Quiescence was attained in Case 5, but only 25 degrees of movement was regained. Quiescence was attained temporarily in Case 6, with 90 degrees of movement. Reactivation occurred 21 months later. Quiescence was not attained with local streptomycin in Cases 2, 7, 9, and 10. It is perhaps not without significance that treatment was discontinued after four injections in Case 2.

Reactions were few. One case developed a rash which did not interfere with treatment. A positive sputum in this case became negative on completion of treatment. A slight local reaction with increased temperature and tenderness not lasting more than 24 hours was occasionally noted, but this occurred only when difficulty was encountered in entering the joint and some of the streptomycin was injected into the soft tissues.

It will be at least two years before enough cases have been treated and followed for any final opinion on this method of treatment to be given. We feel justified in publishing this interim report in the hope that other surgeons seeing these cases early will try to establish a diagnosis and begin treatment before cartilaginous damage takes place.

The average case admitted to an orthopaedic hospital has had symptoms for a year or more.

Summary

The results obtained with intra-articular streptomycin in synovial tuberculosis of the knee are encouraging.

Early diagnosis by gland and/or synovial biopsy is essential.

Four of our ten cases have progressed well, and in two normal function has been re-established.

REFERENCES

- Ahern, R. T. (1950). *Lancet*, 1, 443.
 Arden, G. P. and Scott, J. C. (1947). *British Medical Journal*, 2, 87.
 Bickel, W. H., Young, H. H., Pluetze, K. H., and Norley, T. (1948). *J. Amer. med. Ass.*, 137, 682.
 Jones, G. B. (1948). *J. Bone Jt Surg.*, 30B, 106.
 Smith, A. D., and I-Sen Yu, H. (1950). *J. Amer. med. Ass.*, 142, 1.
 Sireeten, D. H. P. (1949). *S. Afr. med. J.*, 23, 777, 850.

ANGIOMA VENOSUM RACEMOSUM WITH ANGIOMATOUS LESIONS OF SKIN AND OMENTUM

BY

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Vascular malformations and tumours of the central nervous system are sometimes associated with similar lesions of the skin and viscera. Such associations are known by the names of Sturge-Kalischer-Weber, von Hippel, and Lindau when the nervous tissue involved is either the cerebrum or cerebellum.

Wyburn-Mason (1943) quotes numerous instances of skin naevi coexisting with a variety of vascular lesions of the cord. However, he notes only seven instances in which angioma venosum racemosum is associated with a skin naevus, and quotes the cases of Cobb, Alexander, Rand, Sterling, and Jackinowicz, adding three cases of his own. In only one of these (his Case 12) was the skin lesion non-segmental. All cases described had the cord lesion in either the lower dorsal or the lumbar areas.

Subsequently, Cross (1947) published the first case of extensive subarachnoid blood vessels involving the whole of the cervical region associated with an angioma of the left side of the neck and prominent vascular markings of the upper part of the chest and over the seventh cervical vertebra. The present case appears to be the second one on record. Of additional interest is the occurrence of numerous small angiomata of the omentum and a further large skin naevus of the flank.

Description of the Case

The patient was a well-built boy aged 15. Three years before admission he jumped off a 5-ft. (1.5-m.) wall and landed on his feet. He sustained no obvious injury, but 30 seconds later, while walking, he developed occipital headache. He was laid up for two weeks at home, complaining of pain at the back of his head, across both shoulders, and radiating down the left arm as far as the elbow. He stated that for several weeks his left leg tended to drag, but that ultimately recovery was complete. He was well until 10 days before admission, when he made a racing dive, swam two lengths of the swimming-bath, and then suddenly developed pain across the forehead and at the back of the neck. He got out of the water and almost lost consciousness on account of the severity of the pain. He was taken home, where his headaches continued for two days, accompanied by vomiting.

Five days before admission, while lying in bed, the pain in the nape of his neck suddenly became worse and again

FIG. 1.—Large skin naevus over the left flank.

spread across the back of his shoulders. This lasted for three days. On admission he complained only of stiffness at the backs of both legs.

Examination showed him to be a tall, well-built boy. There was general weakness of the left upper limb only, while neck rigidity and Kernig's sign were both present. A large naevus was observed over his left flank and several small ones elsewhere (Figs. 1 and 2), including one within the distribution of the fifth cervical neuronal segment.

Lumbar puncture produced a xanthochromic fluid with raised protein and cells. Radiographs of the skull and neck were negative.

The patient improved gradually, and on discharge three weeks later had recovered completely and was symptom-free. In view of the history suggesting recurrent subarachnoid haemorrhage, the lumbar puncture findings, and the presence of skin naevi, a tentative diagnosis of angioma of the cervical spinal cord was made. Other systems were normal, blood pressure was not raised, and urine examination, E.S.R., and chest radiograph showed no gross abnormality. He was sent to a neurosurgical unit for further investigation, but while waiting for admission to this unit he had a recurrence of symptoms after a friendly tussle with a school friend, and was readmitted to hospital 10 weeks later. Symptoms consisted mainly of pain across the right shoulder which gradually spread down the right side of his body. Examination revealed gross weakness and hypotonus of the right arm and leg, with diminution of tendon-jerks on that side. Neck rigidity was marked, Kernig's sign was positive, whilst the right plantar response was extensor. No sensory loss was detected at this stage. Lumbar punc-

FIG. 2.—The left-flank naevus and other small naevi.

ture produced a clear fluid with evidence of spinal block.

There was a gradual increase of motor weakness, which spread to the left side of the body, so that five days after admission he presented a picture of quadriplegia. There was sensory loss to pin-prick up to the level of C 5 and both plantar reflexes were extensor. Respiratory embarrassment developed, cough reflex was weak, and dysphagia was complained of. His general condition remained poor and gave great cause for anxiety, as it was obvious that the paralysis was gradually spreading.

A neurosurgical team was called in and laminectomy was performed. Unfortunately the patient did not survive the operation, and in view of the extent of the lesion it was doubtful whether it was amenable to surgical treatment. Post-mortem examination revealed a large venous angioma overlying the whole of the cervical cord (Fig. 3); numerous small angiomas of the omentum (Fig. 4); and the skin naevi previously mentioned, which of course were observed *ante mortem*.

FIG. 3.—Large venous angioma of cervical cord.

Discussion

Reference to the symptomatology of recorded cases (Wyburn-Mason, 1943) indicates a fairly constant pattern. There is sudden onset of pain of root type associated with subjective sensory phenomena of tingling and numbness. Weakness or paralysis supervenes sooner or later, and is followed by a variable degree of recovery. Relapse occurs, with general worsening of the clinical state. Although sudden stress of exercise may precipitate symptoms there is no real evidence that trauma plays a part in basic aetiology.

Wyburn-Mason suggests that vascular naevi of the skin are not associated with any special type of blood-vessel abnormality of the cord, but may coexist with any variety. Furthermore, such naevi may or may not occupy segmental skin areas corresponding to the state of the cord lesion.

However, it is worth noting the words of Stanley Cobb (1915), who says that skin naevi are at times of diagnostic value when segmental phenomena referable to the central nervous system are present, and that congenital vascular

FIG. 4.—Small angiomas of the omentum.

tumours arise from a developmental fault of the central nervous system, so these lesions may occur in any of the organs innervated by filaments from that neuromere. Appreciation of the skin naevi in this case enabled a correct pre-operative diagnosis to be made.

Summary

A case is described of angioma venosum racemosum of the cervical spinal cord associated with angiomata of the omentum and skin naevi of the same metameres. The diagnostic value of the cutaneous lesions is stressed.

I wish to thank Dr. Eliot Mackworth and Dr. Philip Buckley for permission to publish this case, and to express my appreciation of the efforts of Dr. L. Potaschmacher for his painstaking post-mortem examination.

REFERENCES

- Cobb, S. (1915). *Ann. Surg.*, 62, 641.
Cross, G. O. (1947). *Arch. Neurol. Psychiat.*, Chicago, 58, 359.
Wyburn-Mason, R. (1943). *Vascular Abnormalities and Tumours of the Spinal Cord and its Membranes*. London.

Medical Memoranda

Two Cases of Tetanus Neonatorum Treated with Penicillin and Aureomycin

Umbilical tetanus is fortunately rare in the British Isles; possibly for this reason the literature on the subject is relatively scanty, and standard textbooks do not deal fully with the use of modern drugs in this condition. W. Sheldon, for instance, in his *Diseases of Infancy and Childhood* (5th edition, 1946, p. 24), recommends 20,000 units of antitetanic serum (A.T.S.) and sedation with chloral and magnesium sulphate. He does not mention the use of the sulphonamides or penicillin. The following cases may therefore be of interest. Both occurred before the end of the second week of life, and made a complete recovery.

CASE 1

An African boy aged 7 days had a history of constant crying. On examination some tendency to rigidity of limbs but no definite spasm was seen. The umbilical wound had not healed and was dirty and plastered with leaves. His temperature was 99° F. (37.2° C.) and pulse 130. Heart and chest were normal. He was given A.T.S., 100,000 units immediately, and penicillin "G," 200,000 units four-hourly, and the cord was dressed with sulphanilamide powder. By the afternoon of admission muscular spasm and trismus of the jaws was obvious. Sedation with chloral hydrate and potassium bromide, 15 gr. (1 g.) of each four-hourly per rectum, was begun. Any reduction of this dose at once resulted in a return of the spasms. For the first 24 hours no attempt was made to pass a nasal catheter, and fluid was given per rectum—2½ oz. (70 ml.) of water two-hourly.

On the second day four-hourly nasal feeds of expressed breast milk were given, but the water per rectum was continued, to guard against dehydration. For five days the whole of this quantity was retained. On the third day penicillin was reduced to 100,000 units four-hourly, and sedation continued. It was now found that the child could swallow, and part of the feeds was given by syringe. As soon as the child tired of swallowing, the tube was passed to complete the feed. On the fifth day he was sucking the syringe strongly; part breast-feeding was begun; phenobarbitone, 1 gr. (65 mg.) four-hourly, was substituted for bromide and chloral; and rectal fluids were discontinued. On the seventh day he was fully breast-fed and phenobarbitone was discontinued. He was discharged in good condition on the ninth day. Two days later he was seen at the clinic. He was then perfectly well and there was no residual stiffness.

CASE 2

An African girl aged 9 days was brought to hospital because she was unable to suck. The child was crying continuously, with risus sardonicus, clenched fists, stiffly extended legs, and taut abdomen. Handling produced convulsive arching of the spine. The umbilical wound was 1½ in. (3.1 cm.) across, moist, and filthy. Her temperature was 100° F. (37.8° C.) and pulse 134. Her heart and chest were normal. She was admitted at once (February 13, 1951). A.T.S. was not obtained till February 17, when 100,000 units was given. Treatment was carried out on exactly the same lines as in Case 1, but the violent reactive spasms remained unabated for seven days, although the child was quiet unless touched. It was necessary to aspirate four-hourly, or more frequently, with a rubber mucus catheter, the nurse summoning medical aid for this purpose each time the child became cyanosed or showed difficulty in breathing. Without these ministrations she would undoubtedly have died. Passing the catheter to the back of the mouth and directing it to the epiglottis was made difficult by the jaw trismus.

On February 19, as the temperature had not responded to penicillin, "aureomycin," 250 mg. four-hourly, was substituted. From this time onwards the improvement was marked and steady, except for one setback. During a nasal feed on the 21st the child suddenly collapsed and ceased to breathe. She was restored by artificial respiration and nikethamide. No feed had been inhaled, and the collapse may have been due to vagal inhibition coupled with respiratory spasm. Curiously enough, the child appeared much better the day after this episode. The aureomycin was now reduced to 120 mg. four-hourly. On February 22 all reactive spasms had ceased and the child could be handled gently without arching of the spine resulting. Nevertheless the whole body remained stiff, especially the legs, which were all the time held stiffly extended except in sleep, when they relaxed slightly. The fists remained clenched and the head retracted. Phenobarbitone, 1 gr. (65 mg.) four-hourly, was substituted for the other sedatives. Massage and movements were started, and in eight days the limbs were back to normal. The aureomycin was discontinued on February 22 (total 3,000 mg.).

COMMENT

Only one dose of A.T.S. was given owing to local shortage. The successful outcome in Case 2, which was in all respects extremely severe, suggests that delay in giving A.T.S. need not prove fatal. It seems logical to give large doses of penicillin or of a sulphonamide, because septicaemia is a strong probability. It is interesting to note the response to aureomycin in Case 2. The very high fluid intake would seem to be necessitated by the muscle spasm, which no doubt increases the body's needs. It may possibly help to prevent serious consequences from the heavy sedation. It is certain that in Case 2 the patient would have died from pneumonia, if not from asphyxia, had she not been aspirated frequently.

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A Combination of Congenital Anomalies

It is well known that the presence of one congenital defect in a person should make one examine for the presence of other anomalies. The following case illustrates this fact.

CASE REPORT

A female Bantu child weighing 4 lb. 5½ oz. (2 kg.) was born in the maternity section of Baragwanath Hospital on September 26, 1950. She was premature, of approximately 36 weeks, and delivery was very rapid. She was noticed to be blue from birth. She did not cry or breathe after birth, but the heart was beating strongly. Lobeline was given by injection, the air passages were cleared, and oxygen was administered. She soon started to breathe, but remained