FIG. 3.—A microphotograph of a section, stained with hæmatoxylin and eosin, of the posterior sinus. The sinus is lined by a modified type of stratified squamous epithelium.

aided by probing. The differential diagnosis however includes the primary chancre of syphilis, chan-

croid, granuloma venereum, tuberculous sinuses, infected sebaceous cysts and the very rare pilonidal cyst of the perineum (Woolridge, 1955). The sinuses under discussion have no communication with the urethra and so may be distinguished from hypospadias and urethral fistulæ secondary to some other urethral pathology.

On section, the sinuses are lined by stratified epithelium, while on the other hand cysts may be lined either by stratified squamous or columnar epithelium.

Various methods of treatment have been employed but in this case simple laying open of the sinus was speedily effective.

Summary

A case of ventral peno-scrotal sinus is described. Its embryology, differential diagnosis and treatment are discussed.

I would like to thank Professor H. Ellis for helpful advice and for allowing me to publish this case, Dr. I. Dawson who prepared and reported on the histology, and also the Photographic Department of the Westminster Hospital.

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PNEUMOCOCCAL MENINGO-ENCEPHALITIS

An Unusual Case

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PNEUMOCOCCAL meningitis has always a poor prognosis. This particular case is interesting for two reasons: firstly, although it seemed at one time that recovery would be very limited, in fact it was remarkably complete; and secondly, there were, during the latter part of the illness, some unusual neurological features.

Case Report

The patient, a male aged 40 years, was admitted on January 11, 1962. There was no history of skull trauma. About two weeks prior to admission he had had a mild respiratory illness for which his general practitioner prescribed oxytetracycline. Recovery was satisfactory, but he had not yet returned to work, when on January 11, he became suddenly febrile and confused and complained of intense headache.

On Admission. The patient was confused and drowsy.

Temperature 98.8°F., rising rapidly to 103°F, pulse rate 80/min., B.P. 100/70 mm. Hg. Cardio-vascular system, lung fields and ear drums normal.

CNS: marked meningeal irritation with moderate dysphasia, particularly manifest as spoonerisms. Normal optic fundi and pupils. Left-sided facial weakness of supranuclear type. Tendon reflexes and plantar responses normal.

CSF: increased pressure, positive Pandy reaction, cells 325/cu. mm., mainly polys.; protein 130 mg.; sugar 70 mg., chloride 630 mg./100 ml.; there were abundant encapsulated Gram-positive diplococci, mainly extracellular, which were confirmed on culture as being pneumococci, sensitive to all routine antibiotics.

A diagnosis of pneumococcal meningitis was made and treatment begun with benzylpenicillin, (20,000 U. intrathecally daily, 1 megaunit 4-hourly i.m.) and sulphathiazole (1.5 g. then 1 g. 4-hourly i.m.).

Progress

January 12: Nasal feeding and continuous sedation with intramuscular paraldehyde. General condition unchanged except for the appearance of slight weakness of the left arm. Hb 14.7 g./100 ml., w.b.c. 23, 900/cu. mm.

January 14: Upgoing right plantar response and early bilateral papilloedema.

January 15: Answering simple questions. Flaccid paralysis of left arm.

January 16: Left lateral hemianopia detected by confrontation tests; central fixation of both eyes; pupils normal in all respects. In view of the possibility of there being an intracranial abscess, he was transferred to the Neurosurgical Unit, Killearn Hospital.

January 17: Right carotid angiogram normal apart from minimal atheroma in the carotid siphon.

January 18: Ventriculogram normal apart from a mild degree of cerebral atrophy, more marked on the left side.

January 19: Transferred back to Gateside Hospital. General condition essentially unchanged.

January 22: Afebrile, but very restless and apparently in pain, assuming the foetal position when left undisturbed. Semiconscious but responding to sharp commands. Marked meningeal irritation and intermittent opisthotonos. Abdominal reflexes absent; clonus, right ankle; frankly incontinent of urine for the first time.

CSF: cells 216/cu. mm., 60% polys, 40% lymphs; protein 80 mg., sugar 80 mg./100 ml. Culture: no growth. Blood white cell count normal.

A diagnosis was made of post-basic meningitis and drug therapy changed to chloramphenicol (250 g. 6-hourly) and prednisolone (30 mg. daily).

January 23: Conversing to a minimal extent. Right eye movements present for the first time in the past seven days. Left pupil larger than the right, but both reacting normally to light.

January 24: Answering simple questions lucidly and obeying simple commands, but becoming easily restless and confused and uttering the meningeal cry. Papilloedema regressing and horizontal eye movements normal.

January 29: Sitting up, answering questions in a rather dazed fashion. Retrograde amnesia striking: he did not recognize his wife or other close relatives, but he did instantly recognize and reminisce with a work-mate of his boyhood days, who, by a strange coincidence, was also a patient in the ward at this time.

More formal neurological testing was now possible and this showed that whilst gross motor or sensory loss was absent from all four limbs, and conjugate eye movements were present except in a downward direction, he had apraxia of his left arm and left leg; when asked to move either of these he would move the corresponding limb on the right side. By starting with a simple explanation that he had two arms and two legs, and identifying each in turn by pinprick, it was sometimes but not always possible to make him move the correct left limb to order. After an initial correct try, however, he would select the right-sided limb the next time he was asked to move the left and so the whole procedure of explanation would have to be repeated. There was no demonstrable hemianopia at this time by confrontation tests and he would lean over spontaneously to his left side to pick things up with his right arm. He could feed himself by now and it was noted that he would never use his left arm or hand for this purpose.

By now also his micturition had become automatic as in an infant and he had multiple pressure point damage from friction burns dating from his period of extreme restlessness.

January 30: Abdominal reflexes normal. Identification of left leg nearly normal.

February 3: Cracking jokes and singing 'There's a hole in my bucket'. This folk song has many verses, and he was, most surprisingly, word-perfect. Occasional urinary incontinence.

February 7: More detailed examination now possible, this showed global dysphasia, including alexia with confabulation, and severe acalculia. There was also marked past-pointing in the left finger-nose test.

February 15: Walking with help. Fully continent.

February 20: Betting on horses, and winning, in spite of persistent acalculia on simple formal testing.

February 27: Right plantar response upgoing only on stimulation of the lateral strip of the foot. All drug therapy stopped.

March 7: Detailed examination showed normal visual fields on confrontation, slight left-sided facial weakness of supranuclear type, slight generalized weakness of left arm, slight past-pointing of left finger-nose, drooping and slight supination of the left outstretched arm with the eyes closed, 5 second loss of vibration sense at the left wrist compared with the right and equivocal right plantar response on stimulation of the lateral side of the foot. The heel-shin tests were normal, but he drifted to the right when walking. Dysphasia was minimal: he could count and read aloud fairly well, and could read a novel with some effort. He would lose the place watching a fast television sequence, but could pick up the thread of the story again when the action slowed down.

Orientation remained disturbed in that, although voluntary movements were quite normal and right-left discrimination was normal on command, imitative movements were always made as a mirror image, and he could not stop making this error even when it was pointed out to him.

March 24: He was examined by Mr. McGuire, Senior Clinical Psychologist at the Southern General Hospital, Glasgow, who reported:

'W. M. was cooperative during all the testing, his mood was cheerful and he seemed to have insight into his difficulties.

'I began by giving the patient the Eisenson test for aphasia and found nothing of note. He could name objects, pictures, colours, shapes, letters, words and sentences. He could recognize sounds and also objects placed in either hand. Neither did there appear to be any expressive disturbance or dyspraxia. Writing was poor but the patient maintained it had always been so and spelling was good. The patient tended not to use his left hand and if forced to do so was slower and more clumsy than is usual. Speech was normal. A significant feature that emerged first in this test but recurred frequently was W. M.'s tendency to ignore things appearing in the left visual field. For example, in reading a three or four figure number he would omit the first figure; told to transfer pegs from his left to his right he assumed that he had completed the task although there was still a peg on his left. This defect recurred in other tests; for example in copying a square or similar shape the left side would be left open. On rough testing the patient seemed nevertheless to have peripheral vision on the left side.

'The patient was next given the Wechsler Adult Intelligence Scale. On the verbal tests he obtained an I.Q. of 108, i.e. high average intelligence. The sub-test scores were reasonably consistent, with none below average, although they included a test of abstract thinking and arithmetic. This high average score would take to be the patient's previous intellectual level. On the performance sub-tests, matters were

quite different. Owing to the shortage of time only two sub-tests were done but on each the patient scored at mentally defective level, I.Q. 53. Not only was he extremely slow, but he showed gross perceptual defect, succeeding with only the simplest item on the Block Design test.

'On the Progressive Matrices, which is a pattern completion intelligence test involving no motor activity, the patient again scored at mentally defective level. W. M.'s memory was not greatly impaired. On selected sub-tests from the Wechsler Memory Scale he scored at average level. Orientation was normal apart from his giving the year as 1961 which he corrected when checked.

'Finally the patient was given the Bender Gestalt test of copying simple designs and did very badly. As already stated he tended to ignore or distort the left side of figures and not to use the left side of the paper. He showed, in addition, gross perceptual disturbance at times with parts of designs in completely altered positions, designs running into each other and some perseveration from one design to the next.

'In summary then, before his illness W. M. was probably of high average intelligence. He can still achieve this level on verbal intelligence tests, but on performance tests and test involving perception he can only perform at mentally defective level, and in a manner common among brain-damaged patients. No evidence was found of aphasia or agnosia. Memory was average'.

March 31: Examination showed the following disabilities:

1. Slight weakness of the left arm, except for abduction which was normal, with increased tone and reflexes.
2. Minimal drift to the right when walking, together with a tendency to swing the right arm with the right leg and the left arm with the left leg.
3. Slight right-left confusion on imitative movements only, without a unilateral bias.

April 1: Patient discharged home.

Examinations as an Out-Patient

November 12: At work for the past seven weeks and feeling well apart from being easily distracted when reading. Minimal increase in tendon reflexes in the left arm; minimal weakness of the left arm and left side of face. Slight right-left confusion persisting on vis-a-vis imitative testing.

December 15: Seen by Mr. McGuire. Verbal intelligence tests not repeated because of previous good scores. Performance tests of the Wechsler Adult Intelligence Scale yielded an I.Q. of 69; Progressive Matrices score was an I.Q. of 91, i.e. low average, compared to the previous mentally defective level of I.Q. <70. Continued inattention to objects on the left of his field of vision: copying of drawings was more accurate than before, but there was persistent omission of left-hand digits when copying 3- or 4-figure numbers.

Discussion

The first point that arises is the precise nature of the diagnosis in this patient. The diffuse pattern of abnormal signs is characteristic only of meningo-encephalitis, and the early period of almost decerebrate rigidity would seem to point to particular involvement of the diencephalon at that time. Such is often called post-basic meningitis, and, as much of the damage therein may be due to ischaemia of nerve centres, the apparent efficacy of glucocorticoids in this case is probably highly significant.

W. M.'s defect in orientation was the most interesting disability. While he remained apparently oblivious of his inability to identify and use his left arm and later to distinguish between his right and left sides, he never suffered from the more gross forms of perceptual dysfunction: he might, for example, have been expected to have had difficulty in dressing, getting into bed or sitting down in a chair to face a specific direction. His confusion at watching a fast-moving television film sequence was, however, quite characteristic (Kremer, 1958).

Again, in his neglect of his left visual fields, although this was discovered only latterly and on psychological testing, he resembles the case described by Isaacs (1962) as an instance of neglect of the left half of space, but, whereas Isaac's patient would draw only half an object or read only half a phrase or word, W. M. would not do this on simple testing, and his drawing and reading were normal in this respect, as least by the time formal testing became possible.

Agnosia of the left half of space is usually the result of a right parieto-occipital lesion, and unawareness of the opposite half of the body from such a lesion is termed autotopagnosia.

Anton's Syndrome consists of the rejection of evidence of bodily disease (Brain, 1961), and Isaac's patient showed this as regards his hemiplegia, but W. M. did not. Gerstmann's Syndrome (Gerstmann, 1940) consists of finger agnosia, agraphia, acalculia and failure to discriminate between right and left, and W. M. may be said to have exhibited this syndrome.

Apraxia and visual agnosia frequently co-exist, and the bizarre variety termed prosopagnosia, where the ability is lost to recognize faces, even one's own, has recently been reviewed (*Brit. med. J.*, 1962).

From an overall point of view, however, all these forms and syndromes involving apraxia and agnosia are but facets of one disability, namely memory upset, particularly as regards subjective orientation, or, to put it another way, they represent defects in the subject's body image.

This basic consideration of body image has been discussed in a masterly way by Kremer (1958). Russell (1957) regards the body image as a memory: a circuit pattern established and reinforced by repetition of body movements. In likening the brain in this way to a computer, it is easy to explain the difficulty in localizing functional areas in the cerebral hemispheres. It is likewise easy to explain the importance of the reticular activating system, where such neuronal circuits are narrowly confined to the brain stem (Kremer, 1958).

Again, Kremer (1958) showed that dissociation can exist between a sense of passive movement and a sense of position in space. W. M. had such a dissociation, even latterly, the former being normal in the left arm, the latter being deficient.

Finally, for an explanation of W. M.'s disability in walking, I suggest that the reason was a defect in the flocculo-nodular lobe of the cerebellum. In

such a defect ataxia is present, but formal cerebellar testing is normal because the control of the γ -system is intact (Kremer, 1958).

Summary

A case of pneumococcal meningo-encephalitis is described where initial disability was great, yet recovery surprisingly complete, probably due to glucocorticoid therapy.

Confusion of the body image and ataxia per-

sisted to a limited extent, and the mechanisms of these are discussed.

It gives me great pleasure to thank Dr. Robert Lamb for help and encouragement in the preparation of this paper; Dr. John Goudie and Dr. John Stephens for laboratory investigations; the Staff of the Neurosurgical Unit, Killearn Hospital, Stirlingshire, for their help in the management of the patient; Mr. Ralph McGuire of the Southern General Hospital, Glasgow, for the psychological assessments; Mr. Harvey Cairns for the photographs; and Miss Joyce Grant for secretarial help.

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PSEUDO-RHEUMATIC LEUKAEMIA

A Case Report with Special Attention to the Influence of Treatment

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ACUTE leukaemia, particularly in children, may present in many guises. Skeletal involvement as a cause of pains in the limbs is well recognized. Less commonly polyarthritis occurs as a prominent clinical feature, and the illness can then mimic acute rheumatic fever or juvenile rheumatoid arthritis very closely. In several well-documented reports, emphasis has been laid on the ineffectiveness of salicylates in relieving symptoms, an observation which might lead to consideration of acute leukaemia as an alternative diagnosis in such patients. The case described now illustrates such a mode of presentation; in addition, attention is paid to the influence of treatment, mainly with a steroid preparation, on the subsequent course of the illness.

Case Report

In June 1959, a schoolboy aged 15 was referred to the London Hospital with the provisional diagnosis of juvenile rheumatoid arthritis. During the preceding three weeks he had complained of intermittent aching pain in both knees and mild pain and stiffness in the neck. The left knee had appeared swollen for a few days, some two weeks before his first visit to hospital. During this fortnight he suffered from general malaise, lethargy and anorexia. He had received aspirin and paracetamol without relief of his joint pains.

At the time of his first visit to hospital there were few abnormal features. He was a thin boy of normal

development with mild pallor of mucous membranes. No adenopathy was detected and there was little objective change in the joints, apart from tenderness over the femoral condyles of the left knee. ESR (Westergren) 32 mm./hr., haemoglobin 64%, and radiological appearances of the chest and knee joints normal. Over a period of three weeks, while awaiting admission to hospital, he was given calcium aspirin 1.3 g. q.i.d. and ferrous sulphate 0.2 g. t.i.d. Flitting joint pains continued with increasing intensity and frequency and involved wrists and shoulders as well as knees. Short bouts of intense pain in the knees were particularly distressing at night. Malaise increased, he experienced night sweats and his weight fell.

On examination on entering hospital on July 5, 1959, he was febrile (100°F) and sweating; he looked pale and ill; there was tachycardia and a soft apical systolic murmur was audible. No enlargement of superficial lymph nodes was detectable; liver and spleen were not palpable. There was now objective evidence of abnormality in several joints. Movement was restricted by pain at the left elbow and right shoulder; both knees were warm and contained effusions. Three days after his arrival in hospital the tip of the spleen could be felt and was tender.

A clinical diagnosis of juvenile rheumatoid arthritis was still favoured. ESR was now 80 mm. and haemoglobin 60%. The latex agglutination test was negative. A routine total and differential white cell count revealed acute leukaemia as the true diagnosis. There were 39,400 wbc/cu. mm., of which 'blast cells' constituted 89%, polymorphs. 9%, and lymphocytes 2%. Examining