Cutaneous ulcers and blue mottling of the skin was seen in some patients and peripheral neuropathy in two cases. Some of the patients had joint swelling of ankle or knee, but ankylosing spondylitis was not described.

The prognosis in localized polyarteritis nodosa is good (1, 2, 8) and remission can be induced with systemic corticosteroids, but, due to the rather benign and prolonged course, continuous treatment with high doses of steroids should be avoided (3).

Polyarteritis nodosa is possibly an immune complex disease (7), but the aetiology remains obscure. In our case an elevated agglutination titre for *Yersinia enterocolitica* was found 4 months after the acute exacerbation, although symptoms of yersiniosis—especially gastro-intestinal symptoms—had not been present, and the titre fell to 80 after 10 months. Yersiniosis may be the causative agent in some acute and chronic rheumatic diseases (4), and this infection may possibly have aggravated or even caused the symptoms in our case.

In our patient HLA antigen B 27 was found, confirming the strong association between this antigen and ankylosing spondylitis (6). The presence of this tissue type may have been the cause of an abnormal immune response in our patient. HLA typing should be performed in other cases of localized polyarteritis nodosa in order to assess the possibility of an association with HLA B 27.

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Pachymeningitis Cervicalis Hypertrophica Syphilitica

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Abstract. A rare form of neurosyphilis with hypertrophic cervical pachymeningitis is presented. It is concluded that serological tests for syphilis are still indicated in patients with unexplained neurological or psychiatric symptoms.

The classical forms of neurosyphilis are dementia paralytica and tabes dorsalis. Uncharacteristic forms of the above-mentioned classical forms have been described recently with increasing frequency (1, 6, 9), an event that may be explained by insufficient treatment with penicillin in the early stages of the disease (5).

In this paper, a case of a rare manifestation of neurosyphilis, the hypertrophic cervical pachymeningitis, is presented.

CASE STORY

A 44-year-old woman was admitted to the Department of Neurosurgery under the diagnosis of tumour of the cervical medulla. Six months earlier the woman had irradiating pains in her left upper extremity aggravated by coughing and sneezing and by abdominal contraction. She also complained of paresthesia within the pain area and paralysis of muscles in the same area. For several years, the patient had a tendency to pains in the nape of the neck without radicular symptoms. She was treated with massage, but without benefit. The patient was admitted to the Department of Neurosurgery with a medical history of 2 months. On the basis of clinical evidence of a cervical slipped disc between C5/C6, a cervical spondylodesis 5/6 after Cloward was performed, resulting in a brief relief of pain. No prolapse was revealed by the operation and an EMG performed after the operation was interpreted as a plexus radialis neuritis.

Soon the symptoms from the left upper extremity progressed, resulting in spastic paresis and paresthesias. Similar symptoms were noted from the right upper extremity and, besides, a slight spasticity of the left lower extremity. No disturbance of the spincter function was reported. The patient was admitted to the Department of Neuromedicine where it was concluded that most likely a tumour was situated in the cervical part of the spinal medulla. The patient was then transferred to Neurosurgery 6 months after the debut of her illness.

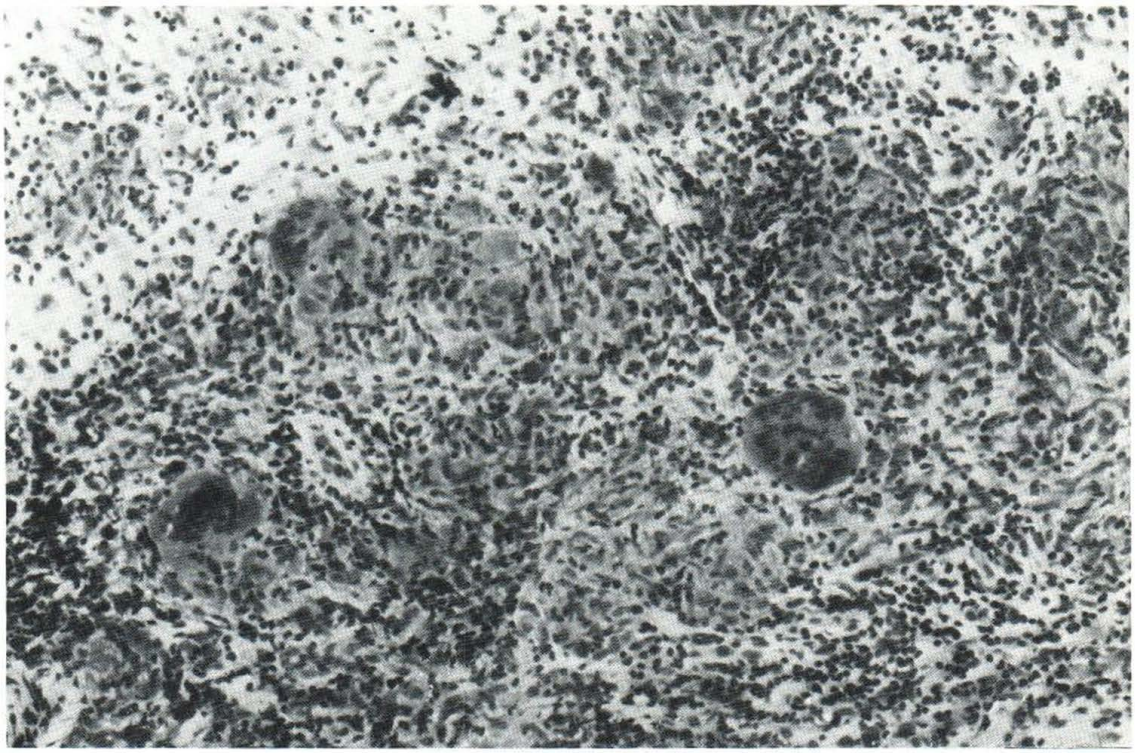

Fig. 1. Photomicrograph of biopsy of the "tumour".

On the suspicion of a tumour, a cervical laminectomy was made. The operation consisted of a decompression of the dura and a biopsy of the "tumour". A large extradural process was found firmly attached to the dura. The tissue was described as hard, scar-like, richly vascular, possibly sarcoma. Freeze microscopy showed a foreign body reaction with giant cells and crystals, but no tumour. The histological diagnosis was: Connective tissue and granulation tissue with subacute, non-specific inflammation and foreign body reaction—suspicion of tuberculosis, suspicion of sarcoidosis (Fig. 1). After the operation, no real improvement was observed.

It was now anticipated that the disease was most likely caused by sarcoidosis of the medulla spinalis, situated extradurally. No other signs of sarcoidosis were present. However, a Wasserman test done at this late stage showed the value of 12, Kahn 8, and Meinicke was strongly positive. TPI was strongly reactive. A lumbar puncture was performed and showed an increased white cell count (13 mill/litre) (<4), a spinal protein of 0.92 g/l (0.20–0.40) and a Wasserman reaction of 1, Meinicke dubious and TPI strongly reactive (3+). Thus, it was concluded that the patient had an active neurosyphilis and the clinical manifestations were consistent with this diagnosis (pachymeningitis hypertrophica syphilitica).

The patient was treated with injections of benzathine penicillin (Penilente®) 2.4 mill IE i.m. four times with intervals of one week. No Jarisch-Herxheimer reaction was seen after the first injection. It is worth mentioning that a Wasserman test performed 10 years before in connection with a delivery was negative.

The patient's husband had a Wasserman titre of 12, Kahn 6, and a strongly positive Meinicke. TPI was strongly reactive (3+). The cerebrospinal fluid showed a white cell count of 65 mill/l and the Wasserman positive with a value of 8. TPI was strongly reactive (3+). The patient was allocated to the same treatment as mentioned above.

DISCUSSION

The hypertrophic cervical pachymeningitis is a rare manifestation of neurosyphilis. The dura mater and the leptomeninges in the cervical part of the spinal medulla are thickened and become adherent because of gummatous lesions, and the blood vessels of the medulla are damaged. The nerve roots and the cervical part of the medulla are compressed, giving radicular pains, muscle atrophy, sensory loss and hyporeflexia. Due to ischemia and compression of the medulla, spastic paraplegia may develop (3).

The patho-anatomic study shows a granulomatous inflammation of the dura, which is considerably thickened (5–10 mm) and often associated with a leptomeningitis. A microscopic examination may

show milliar gummata and panarteritis of the dura but in some instances a hyperplastic fibrous pseudomembranous transformation of the dura is seen, with a non-specific cellular infiltrate of lymphocytes and plasma cells (4, 7, 8).

Tuberculosis and sarcoidosis may also cause hypertrophic pachymeningitis (7).

Other diseases which might give rise to a similar clinical picture are tumours of the medulla spinalis and a cervical slipped disc as in the above-mentioned case. A definite diagnosis can be made only after explorative surgical investigation with a biopsy of the dura, or post mortem. This case underlines the importance of a serological test for syphilis (SRS). It also stresses the need to perform a serological test for syphilis in cases presenting unexplained neurological or psychiatric symptoms. It is also known that the Wasserman test in tertiary syphilis can be negative in both blood and spinal fluid (2).

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