but which may also progress to mild systemic sclerosis.

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Localized Polyarteritis Nodosa in the Lower Limb with New Bone Formation

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Abstract. A 61-year-old male patient, with a burned-out ankylosing spondylitis, developed polyarteritis in the right lower limb, associated with periosteal new bone formation in tibia and fibula. An acute exacerbation may have been precipitated by an infection with yersinia. HLA type: A 28, 26, B 5, 27, Cwl.

Key words: Polyarteritis nodosa; Vasculitis; New bone formation; Yersiniosis

Localized polyarteritis nodosa is a vasculitis in small and medium-sized muscular arteries in cutis, subcutis and occasionally skeletal muscles, particularly in the lower limb (1, 2). The clinical picture is characterized by recurring pain in the involved extremity, livedo reticularis, subcutaneous nodules, ulcerations, and peripheral neuritis. The disease runs a benign course, in contrast to the systemic form.

A few cases of localized polyarteritis nodosa with simultaneous periosteal new bone formation have been published (5, 8). In 1974, 3 cases with new bone formation were reported from the Mayo Clinic (8) and a further 7 cases were collected from the literature. We report here one additional case on this rare pathological entity.

CASE REPORT

A 61-year-old male was admitted to hospital on suspicion of a tumour in the right lower limb.

At the age of 45 he had low back pain from an ankylosing spondylitis. The pains disappeared and the spondylitis burned out, with typical roentgenological sequelae.

Two years prior to hospitalization, scaly, red plaques, bluish discoloration and periodic ulceration appeared on the upper part of the right calf. After nearly 2 years the patient developed increasing swelling, pains, and weakness in the right calf. During this exacerbation there was fatigue and a slightly elevated temperature.

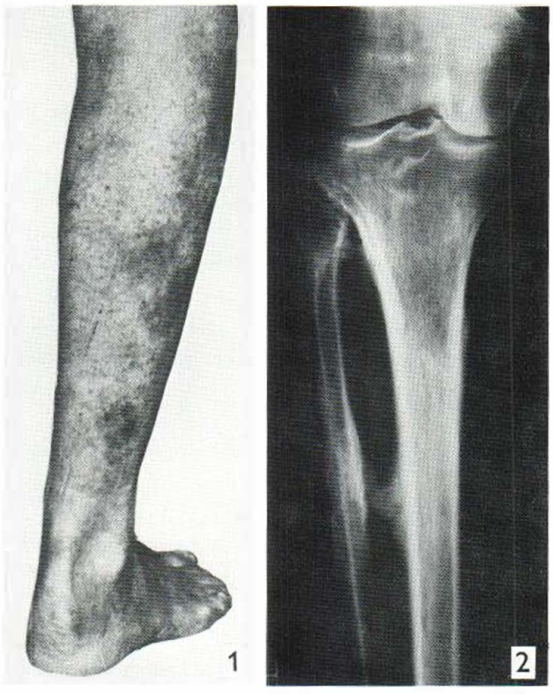

Fig. 1. Blotchy, bluish discoloration on and firm infiltration of the right calf.

Fig. 2. Radiograph of right calf. Periosteal new bone formation and sclerosis of the endosteal osseous structure are revealed.

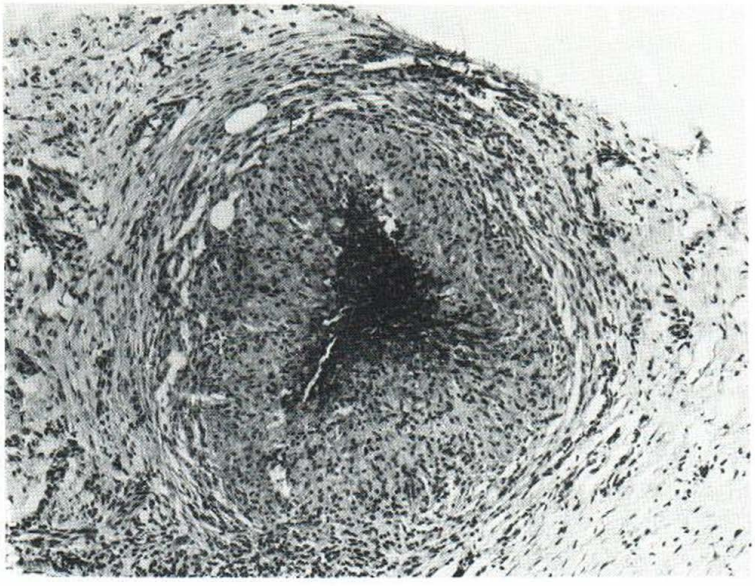

Fig. 3. Muscular artery from corium, with infiltration of leukocytes and proliferation of intima, resulting in severe stenosis (hematoxylin and eosin, ×85).

At examination 2 months after the exacerbation there was moderate swelling of the right calf and on the inner aspect, blotchy, bluish discoloration of the skin, and firm infiltration in the subcutis. On the upper 2/3 of the calf slightly infiltrated, scaly plaques and three small ulcerations were seen (Fig. 1).

The blood pressure was 180/100 mmHg. The erythrocyte sedimentation rate was 47 mm/h, and a leukocytosis (12000/μl) with normal differential count was found. The agglutination titre for *Yersinia enterocolitica* serotype 3 was elevated: 320 (normal <80 (4)) at examination 4 months after the exacerbation. The latex-fixation test was negative, and no LE cells or antinuclear antibodies were found. Other laboratory tests were normal and no signs of systemic involvement were found. HLA antigen typing showed: HLA, A 28, 26, B 5, 27, Cwl.

A radiograph of the right lower leg (Fig. 2) showed marked periosteal new bone formation and sclerosis of the endosteal osseous structure from 5 cm below the knee. No osteolytic foci were found.

Right femoral arteriography showed normal arteries with only slight alteration in calibre centrally as well as peripherally.

Scintigraphy showed marked activity around the right knee and the proximal 1/3 of tibia and fibula.

After these investigations an osteogenic tumour was suspected, and skin-, muscle- and bone-biopsies were taken from the inner aspect of the upper part of the right calf.

Microscopical examination revealed severe arteritis in the muscular arteries of the deep parts of the corium, subcutis, and in striated muscle. Perivascularly and throughout the thickened, oedematous vessel wall, severe infiltration with leukocytes was seen. The infiltrate consisted of both lymphocytes and polymorphonuclear granulocytes, of which many were eosinophils. In severely affected vessels the internal elastic lamella was destroyed and the lumen almost obliterated (Fig. 3). No giant cells were seen. The periosteum was partly hyalinized, and the arteries were here inconspicuous. The underlying bone was normal.

After the exacerbation previously mentioned the temperature normalized during the following week and the pain abated gradually after treatment with caps. indomethacine (Confortid®) 25 mg×3. Subsequently, transient paralysis of the right peroneal nerve with drop foot was observed.

The patient has now been under observation for 12 months. The swelling has disappeared and the ulcers healed by local treatment and continuous therapy with indomethacine. There is no pain in the leg or signs of ischaemia apart from the persistent bluish discoloration and scaly red plaques. The sedimentation rate normalized and the *Yersinia enterocolitica* titre fell spontaneously to 80 after 10 months.

DISCUSSION

Woodward & Andreini (8) published 3 cases of localized periosteal new bone formation in polyarteritis nodosa and 7 cases collected from the literature. The findings were quite uniform and in many respects similar to our case. All of the patients were men, 35 to 63 years old, with pain and swelling in the lower extremity. Increased erythrocyte sedimentation rate was noted in all and a consistent finding was a periosteal reaction in the lower leg and sometimes the metatarsals, and in one case simultaneous new bone formation was seen in the distal radius and ulna. The new bone formation is possibly due to stimulation of the periosteum by the arteritis in the surrounding striated muscles (5).

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.