CASE REPORT

Acute sensorimotor polyneuropathy as an early sign of polyarteritis nodosa.
A case report

Valery M. Kazakov12, Dmitry I. Rudenko12, Tima R. Stuchevskaya12, Oxana V. Posokhina12, Alexander A. Skoromets1 and Semen V. Perfilyev1

1 Department of Neurology, First Pavlov State Medical University and 2 Neurological Department of City Hospital 2 of St. Petersburg, St. Petersburg, Russia

We examined a patient aged 31 who had a sudden burning paraesthesia, pain and numbness in the lower legs together with an increased temperature of 39°C. Clinical examination showed asymmetrical sensory polyneuropathy more clearly seen in the lower legs and the left wrist, with high ESR (up to 44 mm/h), leukocytosis, slight anaemia and proteinuria. CSF was normal. After three weeks the temperature suddenly increased again up to 39°C and severe flaccid distal tetraparesis was seen more clearly with foot drop in the left lower leg and dense oedema in the left wrist, purple cyanosis and haemorrhagic foci appeared on the skin of the toes, feet, lower legs and left wrist. ESP increased up to 65 mm/h, CK was 200 IU (normal ≤ 190 IU) and hypergammaglobulinaemia developed. An EMG study showed sensorimotor, mainly axonal, polyneuropathy with different degrees of involvement of some nerves and with conduction block in the left ulnar nerve. Muscle biopsy revealed findings of inflammatory vasculitis that resembled polyarteritis nodosa with secondary denervation atrophy and non-specific myositis. The patient was treated with high doses of prednisolone, dexamethasone and cyclophosphamide with plasmapheresis. Motor disturbances and pain decreased and the patient began walking with a stick. However, the necrosis of the toes gradually progressed to dry gangrene and amputations of the toes were carried out three months after the disease began. At that time the patient had the clinical features of multisystem disease with progressive heart, lung, liver and kidney failure. The patient died suddenly of pulmonary artery thrombo-embolism a year after the onset of the disease. An autopsy confirmed the diagnosis of polyarteritis nodosa (PN). Thus, in this patient the asymmetrical sensory polyneuropathy progressed rapidly in symmetrical sensorimotor peripheral polyneuropathy which preceded the clinical multisystem involvement in polyarteritis nodosa.

Key words: asymmetrical sensory polyneuropathy, burning paraesthesia, necrosis of the toes, progressive heart, lung, liver and kidney, polyarteritis nodosa

Note. This article was presented as Abstract under name “Neurological complication on periarteritis nodosa” 1999, pp.117-119 in Russian medical Collection “Questions of clinical neurology” ed. By Prof. N.M. Zulev (Moscow) and as Abstract under the name “Acute sensorimotor polyneuropathy as an early sign of polyarteritis nodosa" in the Collection valled “XXIVTH Oxford Symposium on Muscle Disease”, Oxford 1999, p. 24.

Introduction

Polyarteritis nodosa (PN) is one of the systemic necrotic vasculitis. The characteristic pathological feature is inflammation and necrosis of vascular walls of small and middle sizes arteries. These changes are the basis for the infarction and ischemia which can be seen in different tissues and organs. Lesion of the nervous system occurs in 50% of cases with PN (1) The classical example of a peripheral nervous system lesion in PN is mononeuritis multiplex or asymmetrical peripheral motorsensory polyneuropathy. Often neurological signs appear after polyorganic pathology in PN patients, but in some cases lesions of peripheral or central nervous systems may be the first sign of the disease (2-4).

Case report

A male patient aged 31 was admitted to the Neurological Department complaining of burning pains and numbness of the distal parts of the legs, and difficulty in walking due to severe pain. His temperature rose to 39°C and symptoms of nasal congestion appeared. Four days after the onset he was hospitalized with the diagnosis of...
Guillain-Barre Syndrome. However two weeks after admission his general conditions deteriorated with increasing neurological abnormalities. His temperature suddenly increased again to 39°C. Pain burning and numbness increased in the legs and the same symptoms appeared in the hands. He complained of asymmetrical weakness of the distal parts of the legs and small hand muscles, worse on the left. Marked oedema of feet and toes appeared which spread over the lower legs. There was a change of skin of feet toes and lower legs.

The pulse was 96, the blood pressure increased to 160/90 mmHg. The patient was normally orientated in place and time. Cranial nerves were normal. Chest and abdomen were normal. The flexors of the neck were spared. All the small muscles of the hand were severely atrophied and a moderate atrophy of the distal part of forearms was seen. Muscular strength was measured by MRC scale (grade 1-5): shoulder girdle, deltoid, upper arms and extensors of the wrists, grade 5; extensors digitorum communis and flexors of the wrists, grade 3; interosseus volaris and dorsalis and lumbricales, grade 1-2, with both side involved, more clearly on the left. Muscle tone in the upper limbs was decreased. Deep reflexes of the arms were reduced. Abdominal reflexes were absent. There was a moderate atrophy of the thigh and a severe atrophy of shin muscles. Proximal muscle power including hip and knee flexion and extension was normal. There was no movement of the ankle and toes on the left, and muscle power was reduced to 1-2 grades on the right. Muscle tone and knee reflexes were reduced. The ankle jerks and plantar responses were absent on both sides.

There was dysesthesia, hyperpathia and hyperesthesia of all kinds of sensation in the toes, feet and lower third of the lower legs. Lasègue sign was positive. Palpation of ulnar, radial and especially peroneal communis, tibial and dorsal pedis nerves was painful. Palpation of arm and leg muscles was painless. Dense oedema of the lower legs and feet worsen on the left. Purple cyanosis and hemorrhagic foci were seen on the skin of the feet, lower legs, back of the hands and distal part of the left arm. There were necrotic changes of the distal phalanges of the first and third toes of the left foot.

Blood analysis showed a decrease of Hb (to 117 g/L) and erythrocytes (to 3.5 x 10^11) and moderate leukocytosis up to 9.3 x 10^9/L. ESV increased to 69 mm/h. The blood levels of sugar, bilirubin, creatine, urea, cholesterol, serum protein, potassium, calcium and phosphorus were within the normal limits; alpha-1 and alpha-2 globulins were increased up the normal levels. Blood levels of ALT and AST were increased up to 97 U/I (normal, less than 43 U/I) and 156 U/I (normal, less than 34 U); CK was greatly increased up to 2190 U/I (normal below 190 U/I). There was proteinuria 0.39 g/L.

ECG showed a sinus tachycardia of 110 per min. Fiborgastroscopy was normal. Repeated chest X-ray radiographies showed signs of slight oedema of the lungs and possible pericarditis. Ultrasonography showed slight hepatosplenomegaly.

Needle EMG (gastrocnemius, tibialis anterior, extensor digitorum communis and abductor pollicis brevis muscles) showed many fibrillation and single/moderate fasciculation potentials, with positive sharp waves at rest. Many long duration and increased amplitude of MUPs appeared on minimal voluntary contraction. On maximum contraction a discrete pattern was evident. 25-33% of the action potentials were polyphasic, suggesting a neurogenic process.

On electric stimulation of the saphenous and afferent fibres of median nerves the motor responses were absent. Sensory right ulnar nerve conduction velocities were greatly decreased in distal and mildly slowed in proximal parts (14 and 44 m/sec, respectively). Terminal latency was markedly increased up to 7.9 msec.

Motor nerve conduction velocities in the distal parts of right and left ulnar nerves were mildly slowed (44 and 38 m/sec, respectively), but in the proximal parts of ulnar and in the distal and proximal parts of the median nerves were within normal limits. The terminal latencies were increased (in median nerves 6.6 and 4.3 m/sec, and in the ulnar nerves 3/7 and 4 m/sec). The compound muscle action potentials (CMAPs) were markedly decreased in amplitude on stimulation of the ulnar (1 and 0.1 mV) and median (0.5 and 1.2 mV) nerves. On stimulation of the right peroneal nerve the very lower amplitude (0.7 mV) response was seen only in the tibial anterior muscle. Motor conduction velocity was greatly decreased to 24 m/sec (below fibular head-popliteal fossa).

Left motor unlar nerve conduction showed partial conduction block between the elbow and wrist. The amplitude of the CMAPs was reduced by 79% and the area by 88%. The criterion for partial conduction block was a 50% or greater reduction in amplitude and a 40% or greater reduction in negative peak area of surface-recorded CMAPs obtained with proximal compared with distal stimulation of ulnar nerve, in the absence of increased duration of CMAPs more than 20%, according (3).

These ENMG findings showed mainly axonal sensorimotor polyneuropathy with different degrees of affection of some nerves, more clearly seen in the lower limbs with partial motor conduction block in the left ulnar nerve (see Tables 1-2).

The biopsy (stain H.E. and Van Gieson) performed at the right gastrocnemius muscle showed various changes of the majority of epimysial, perimysial and endomysial arteries of medium and small sizes. Simultaneous atrophy of almost all muscle fibres with knots and chains of
dark sarcolemmal striations were invisible and sarcolemmal nuclei disoriented. In the transverse section, in some regions uniform small fibers with degenerative changes, dark cluster nuclei and mononuclear infiltration were also seen. In other regions of the sections increased variation of the diameter of muscle fibres, loss of few fibers, focal necrosis of some muscles with phagocytosis, increased of connection tissue in the endomysium and cellular infiltration were evident. The skin biopsy from right lower leg, at the site of muscle biopsy, showed smoothing of the papillary layer and oedema with inflammatory mononuclear infiltration in the arterial and venous walls (Figs. 1-6).

Cross section of biopsy of right gastrocnemius muscle (formol-calcium, Van Gieson stain) (Figs. 1-6).

Table 1. Electrophysiological data in the affected motor nerves.

| Nerve          | Amplitude (mV) | Duration (ms) | CV m/s | Terminal latency (ms) | Area mV ms |
|----------------|----------------|---------------|--------|-----------------------|------------|
| Ulnar R.       | 1.0            | 9.5           | 44/69  | 4.0                   | 5.0        |
| Ulnar L.       | 0.1            | 3.8           | 38/55  | 3.7                   | 0.2        |
| Median R.      | 0.5            | 8.4           | 56/68  | 4.3                   | 1.5        |
| Median L.      | 1.2            | 17.6          | 54/64  | 6.6                   | 10.5       |
| Peroneal R.    | 0.7            | 18.6          | 24     | 2.9                   | -          |

(below fibular head-popleteal fossa)

Table 2. Site and value of the conduction block and electrophysiological data in the left ulnar nerve.

| Level          | Amplitude (mV) | R1 Area (mV ms) | R2 Area (mV ms) | Duration (ms) | CV m/s | Terminal latency (ms) |
|----------------|----------------|-----------------|-----------------|---------------|--------|-----------------------|
| Prox. Distal   | Prox. Distal   | Prox. Distal    | Prox. Distal    |               |        |                       |
| Below elbow    | 0.13           | 0.2 (79%)       | 0.23 2.08       | 0.1 (88%)     | 3.7    | 38/55 3.7              |

Figure 1. Thickening in the arterial walls with mononuclear cellular infiltration in the arterial walls and around arteries with stenosis of their lumens. Cellular infiltration of the muscle is seen, as well. X 80

Figure 2. Thickening and inflammatory cellular infiltration in the arterial walls with trombosis of a small muscle arteria. X 200

Figure 3. The same section. Foci of fibrinoid necrosis with inflammatory infiltration and with marked thickening of the arterial walls with marked stenosis of the lumen of arteries. X 400
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The muscle biopsy findings were compatible with productive inflammatory vasculitis that resembled polyarteritis nodosa with secondary neural muscular atrophy and nonspecific myositis.

The patient was treated with intravenous infusions of dexamethasone 150 mg daily for 5 days, then prednisolone – 500 mg daily – for 5 days and later with prednisolone, 90 mg/day per os in association with intravenous infusion of cyclophosphamide, 200 mg/day for 15 days, followed by cyclophosphamide, 150 mg/day per os, and plasmapheresis.

Sensory and motor disturbances decreased and the patient began walking with a cane. The skin swelling decreased, ESR and leucocytosis fell, but a pronounced trophic and vegetative vascular disorders persisted in toes. The necrosis of the toes gradually progressed to dry gangrene. The patient was transferred to a surgical department; further he developed a progressive heart, lung, liver and kidney insufficiency, despite a moderate regression of the neurological symptoms. The patient suddenly died of thromboembolism of the pulmonary artery one year after the onset of the disease. The autopsy confirmed the diagnosis of polyarteritis nodosa.

Discussion

The first symptoms of the disease indicated a peripheral nervous system involvement, namely, an asymmetrical sensory neuropathy, without signs of systemic or polyorganic involvement. Furthermore, positive sensory disorders (pain, paresthesia, hyperpathia, hyperalgesia) in legs were the main clinical signs in the early stages of the disease, before the appearance of motor disorders. The asymmetrical sensory polyneuropathy rapidly progressed to a symmetrical sensorimotor peripheral polyneuropathy which anticipated the multisystemic manifestations.

The ENMG and needle EMG findings confirmed the typical manifestation of sensorimotor peripheral polyneuropathy showing the characteristic axonal nerve lesions. However, one of the peculiarities of the ischemic neuropathy in our patient was the signs of partial conduction block in the left ulnar nerve (3, 4). At our knowledge, there are only a few descriptions of the appearances of conduction block in ischemic peripheral nerve lesion in medical literature (1).

The unusual feature of the severe trophic disorder of the distal parts of the legs not responding to specific therapy and leading to dry gangrene of the toes, remains unexplained.

Conflict of interest

The Authors declare to have no conflict of interest.
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How to cite this article: Kazakov VM, Rudenko DI, Stuchevskaya TR, et al. Acute sensorimotor polyneuropathy as an early sign of polyarteritis nodosa. A case report. Acta Myol 2019;38:184-8.

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