Neurology, Psychiatry and Brain Research xxx (xxxx) xxx–xxx

Contents lists available at ScienceDirect

Neurology, Psychiatry and Brain Research

journal homepage: www.elsevier.com/locate/npbr

Amyloidoma of the peripheral nerve–First experiences with two cases

Lena Federle[a,*], Gregor Antoniadis[a], Angelika Scheuerle[b], Maria Teresa Pedro[a]

[a] Peripheral Nerve Surgery Unit, Department of Neurosurgery, University of Ulm, Guenzburg, Germany
[b] Institut of Neuropathology, University of Ulm, Guenzburg, Germany

ARTICLE INFO

Keywords:
Amyloidosis
Peripheral nerve amyloidoma
Magnetic resonance neurography
Neurosonography

1. Introduction

Amyloidosis is a very rare disease. The pathogenesis is a disorder in protein folding. This leads to insoluble proteins which deposit intra- or extracellular in vessels, organs or nerves. These insoluble proteins cause dysfunction like bleeding, organ failure or polyneuropathy (Herold, 2011).

The most frequent amyloidogenic proteins are transthyretin (TTR), serum protein A (AA) and light chain (AL) protein. The former is hereditary whereas latter are non-hereditary forms (Herold, 2011).

Clinical manifestations of amyloidosis can be very different, so it often takes years to final diagnosis. Mostly it is a systemic disease, but in some few cases it may occur locally. If peripheral nerves are affected by amyloidosis, besides clinical examination and electrophysiological testing, a biopsy of one suspicious fascicle is essential to confirm the diagnosis. Magnetic resonance neurography (MRN) and neurosonography (NS) are complementary tools but due to the few existing cases no specific morphologic criteria of amyloidoma of the peripheral nerve is available.

In the following we present two cases of isolated amyloidoma of a peripheral nerve with proof of protein fibrils consisting of light chains, so-called AL-amyloidoma.

2. History and physical examination

2.1. Case 1

A 55-year-old lady presented with a foot and toe flexor weakness of the right side for one year in January 2013. Symptoms occurred suddenly and there was no accompanying trauma, hypaesthesia or radicular pain. Initially symptoms had improved by transcutaneous electrical nerve stimulation and physiotherapy. Since September 2012 a deterioration of existing paresis occurred. The physical examination showed a complete paresis of the anterior tibial muscle as well as of the dorsiflexors and the big toe-lift muscle (MRC grade 0) according to the medical research council muscle scale (MRC). There was no hypaesthesia and no Tinel's sign.

2.2. Case 2

A 43-year-old woman suffered of hypaesthesia of the left ulnar side of the hand and the fifth finger for two years in 2011. In addition she complained about pain in the ulnar forearm and partially in the upper arm. These complaints had improved by osteopathic treatment. Further a weakness in the fifth finger occurred, so she presented herself to our department.

The patient showed an atrophy of the hypothenar and the first interosseus dorsalis muscle. Tinel's sign above the left sulcus ulnaris was positive. Muscle power in the ulnar nerve innervated muscles was two to three (MRC grade 2–3) in the flexor digitorum profundus and the first dorsal interosseus muscle up to four in the abductor digiti minimi and lumbricales muscles (MRC grade 4). Two-point discrimination of the fifth finger was ten millimetres.

3. Electrophysiological testing

3.1. Case 1

Several neurological electrophysiological examinations of the peroneal nerve were performed. Initially the motoric nerve conduction velocity was not derivable and there was pathologic spontaneous activity in the anterior tibial muscle. Throughout jumps in the amplitude

* Corresponding author.
E-mail address: lena.federle@gmx.de (L. Federle).

https://doi.org/10.1016/j.npbr.2017.12.001
Received 20 September 2017; Received in revised form 19 November 2017; Accepted 2 December 2017

above the tip of the fibula in the motor neurography were seen. Electromyography of the anterior tibial muscle showed persistent pathologic spontaneous activity without volitional activity.

3.2. Case 2

Electrophysiological examination demonstrated a decreased nerve conduction velocity of the left ulnar nerve (56 m per second on the right side per second vs. 70 m per second on the left side) in sensory electroneurography. By use of fractional derivatives in motoric electroneurography a reduced amplitude of the motor response above the left ulnar sulcus was determined (2,6 mV, 7,0 mV and 7,6 mV on the right side vs. 3,1 mV, 10,3 mV and 12,6 mV on the left side).

4. Preoperative MRN and neurosonography

4.1. Case 1

Within the outpatient presentation nerve ultrasound was performed. It showed a thickened hypoechoic fascicle of the right deep branch of the peroneal nerve about ten centimetres proximal to the fibula. There were inconspicuous fascicles as well. In an MRN of the right knee and lower leg no tumor in the knee joint was detected.

4.2. Case 2

In the preoperative neurosonographic examination thickened hypoechoic fascicles of the ulnar nerve in the range of the ulnar sulcus were seen.

Two MRNs were performed before patient's presentation. Both showed a long-segment partly cystic partly solid soft-tissue proliferation in the course of the ulnar nerve. It looked similar to the morphological appearance of perineurioma with a long distance uptake of contrast medium in the T1 sequence (Figure1 Fig. 1).

5. Intervention

5.1. Case 1

It was decided to perform a neurolysis of the right deep branch of the peroneal nerve and take a biopsy of one thickened fascicle. Intraoperative nerve action potentials (NAP) concerning the deep peroneal branch amplitudes were absent. Intraoperative ultrasound of the nerve showed a number of thickened fascicles proximal the popliteal fossa. A biopsy of one of these fascicles was taken after electrical stimulation showed no contraction of the muscles supplied by the peroneal nerve.

5.2. Case 2

Due to diagnosis of a cubital tunnel syndrome and suspected diagnosis of a perineurioma of the ulnar nerve a decompression of the nerve and a biopsy of one thickened fascicle was planned (Figure3 Fig. 3). Neurolysis and volar transposition of the left ulnar nerve above the ulnar sulcus was performed. Intraoperative neurosonography showed string of pearl-like hypoechoic thickened and hardened fascicles (Figure5 Fig. 5,). A biopsy of a conspicuous fascicle was taken after stimulation showed no motor response.

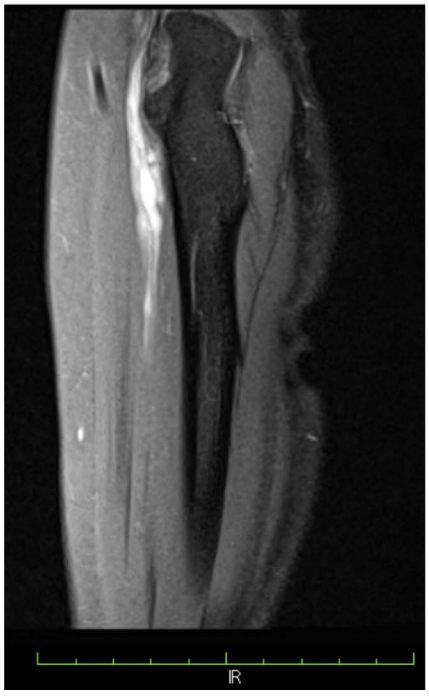
6. Histological examination

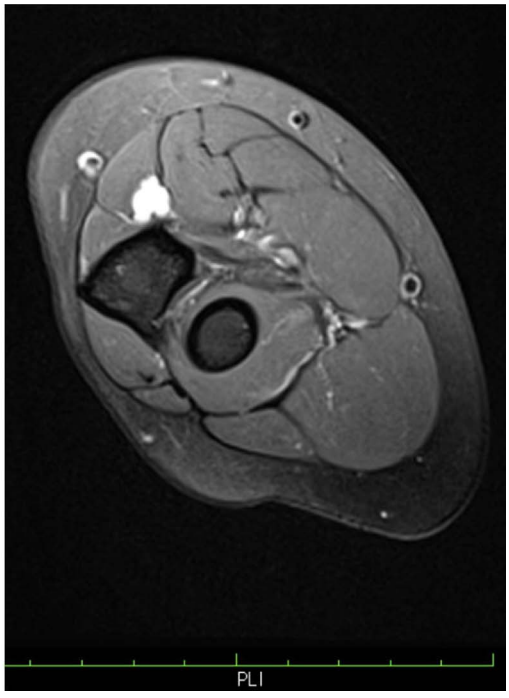
6.1. Case 1

Immunohistochemistry was performed. Congo red staining was positive for the existence of amyloidosis. Immunohistochemistry revealed perineural deposits of amyloid light chains. The subtype of the light chains was lambda.

6.2. Case 2

Immunohistochemistry of the bioptate showed depositions of amyloidoma. In the subtype classification by performing immunohistochemistry a lambda light chain amyloidoma was diagnosed (Figs. 7 and 8).





Figs. 1 and 2. Coronal and transversal T1 sequence with gadolinium contrast.