**PATHOLOGICA 2019**;111:67-69.; doi: 10.32074/1591-951X-9-19

Case Report

# Peripheral nerve mucoid degeneration involving the sciatic nerve

V.P. Fabbri[1], A. Valluzzi[2], N. Acciarri[2], M.P. Foschini[1]

*[1] Unit of Anatomic Pathology, Department of Biomedical and Neuromotor Sciences, University of Bologna at Bellaria Hospital, Bologna, Italy; [2] Department of Neurosurgery, IRCCS, Bellaria Hospital, Bologna, Italy*

**Summary**

Peripheral nerve mucoid degeneration (PNMD) is a rare non-neoplastic degenerative condition characterized by endoneural deposit of mucoid matrix. Herein, we report a case of PNMD involving the sciatic nerve with preoperative features, surgical treatment and pathological findings.

**Key words**

Peripheral nerve mucoid degeneration • Sciatic nerve

## Introduction

Peripheral nerve mucoid degeneration (PNMD) is a rare, non-neoplastic degenerative condition, characterized by localized axonal damage and Schwann cell/myelin loss associated with intra-neural mucoid changes and fibrosis. PNMD is usually due to chronic nerve injury and entrapment syndrome [1], and it usually affects the upper extremities, mainly in young adult male patients [2]. Differential diagnosis should be carried out with focal mucoid degeneration, which can be observed as progressive neuro-muscular atrophy (Charcot-Marie-Tooth) [3] or peripheral polineuropathy related to hypothyroidism [4]. In daily practice, PNMD is encountered in small cutaneous nerves entrapped in scars after surgical procedures. Local pain, paresthesias, nerve paralysis and possible palpable swelling are the commonest symptoms. Magnetic resonance imaging (MRI) may help in visualizing alteration of the affected nerve, but MRI features are often non-specific. Therapeutically, surgical neurolysis may improve the clinical picture, avoiding recurrences, but the functional result depends on the integrity of neural fibers.
Herein, we report a case of PNMD involving the sciatic nerve, describing preoperative features, surgical treatment and pathological findings.

## Case report

A 52-years-old man presented in August 2016 complaining of pain and paresthesia localized in the left ankle, not referred to any injuries or previous trauma. Lumbar radiography did not reveal any pathological features.
After a few weeks, the symptoms dramatically increased leading to a complete paralysis of the left leg and foot.
The patient was submitted to a MRI of the lumbar spine, which did not show any disk disease or other spinal pathology that could be causing the clinical symptoms.
On the other hand, electromiography and neurography studies showed complete mono-neuropathy of the left sciatic nerve, mainly in the distal part of peroneal and tibial compartments, with both sensory and motor impairment.
Therefore, MRI was repeated with contrast medium,

**How to cite this article:** Fabbri VP, Valluzzi A, Acciarri N, et al. *Peripheral nerve mucoid degeneration involving the sciatic nerve.* Pathologica 2019;111:67-9. https://doi.org/10.32074/1591-951X-9-19.

**Correspondence:** Viscardo Paolo Fabbri, Unit of Anatomic Pathology, Department of Biomedical and Neuromotor Sciences, University of Bologna at Bellaria Hospital, via Altura 3, 40139 Bologna, Italy - E-mail: viscardopaolo.fabbri@studio.unibo.it

focusing on the pelvis. This new MRI showed a diffuse and regular enhancement, measuring 8 cm in length, along the left sciatic nerve, involving the region below the piriform muscle (Fig. 1).
The sciatic nerve did not show features of neoplastic swelling, although the signal intensity suggested the presence of an intra-neural lesion. The MRI features could not rule out benign or malignant neoplasms and therefore a surgical exploration was delivered.
On admission, the patient exhibited a complete left sciatic nerve paralysis in the left leg and foot, with evoked pain by palpation in the rear side of thigh and sensory alteration in the anterior-lateral side of leg and the dorsum of the foot.
The patient was operated on in the prone position, with a surgical approach to the left sciatic nerve through the gluteus. On surgical exploration, the sciatic nerve under the piriform muscle appeared without neoplastic deformations. However, the nerve showed a roughly and soft, pale constitution, with the inferior branch appearing flabby and yellowish. Since a real mass or tumor was not encountered, a complete neurolysis of the sciatic nerve was performed. A 2 cm length biopsy was taken, on the yellowish sciatic nerve branch.
On frozen section, peripheral nerve tissue with intraneural myxoid changes were noted, but a diagnosis could not be reached. Therefore the tissue was formalin-fixed and paraffin-embedded according to routine procedures. Blockes were serially cut and sections were stained with haematoxylin-eosin, Alcian blu pH 1.00. Immunohistochemistry was performed in an automated stainer, (Ventana, Tucson, AZ, using Ventana purchased pre-diluted antibodies).

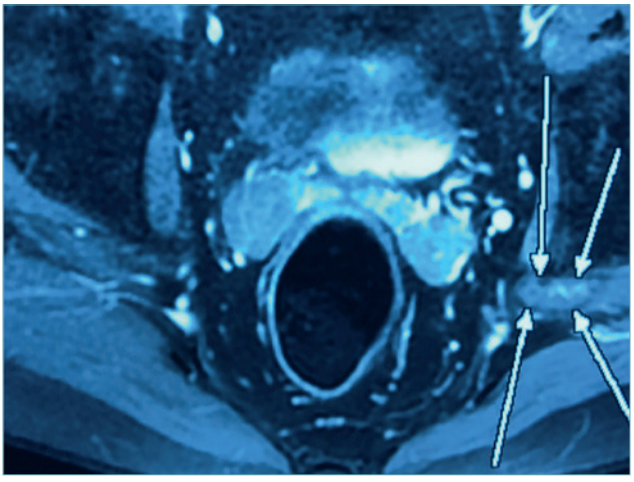

**Fig. 1.** MRI showed a diffuse and regular enhancement along the left sciatic nerve.

## Microscopic findings

On histological examination (Fig. 2), the nerve appeared enlarged due to endoneural deposits of mucoid matrix, Alcian bleu pH 1.00 positive, that displaced and compressed the swelled neural fibers.
Epineurial tissue appeared slightly fibrotic, while perineurium and endoneurium were thickened. Inflammatory or atypical cells were not observed either in nerve

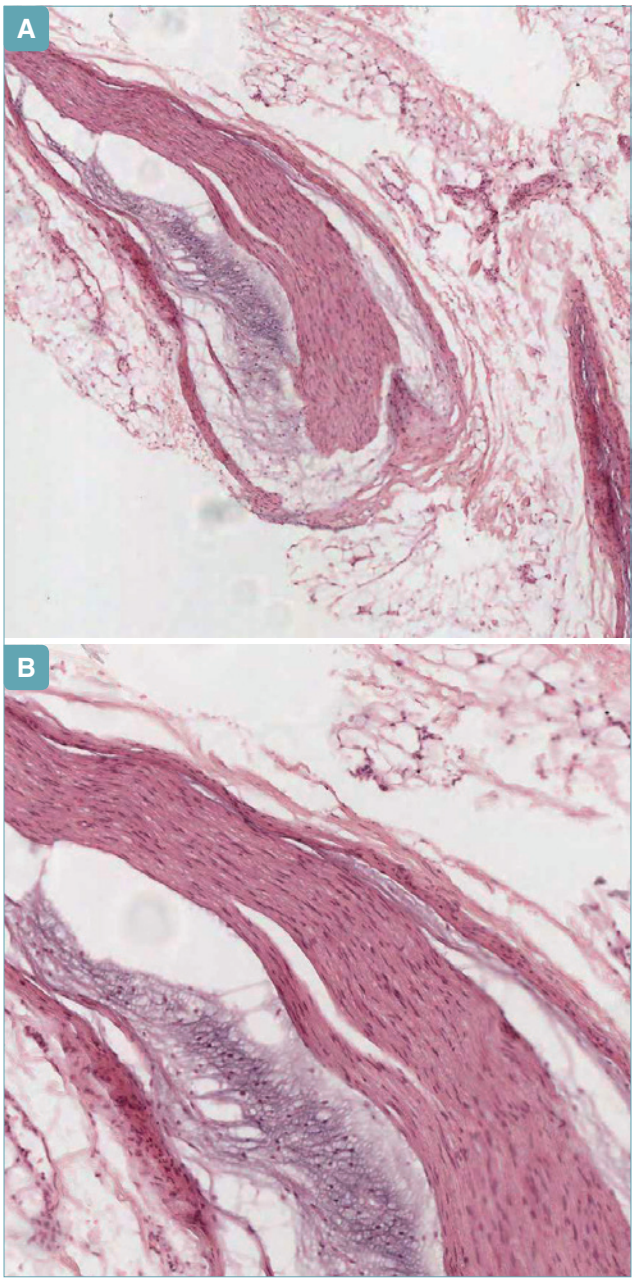


**Fig. 2.** Nerve enlargement due to endoneural deposit of mucoid matrix.

fibers or in the surrounding fat tissue; surrounding blood vessels presented a minimally thickened wall.
Immunohistochemistry confirmed the presence of displaced and compressed swelled axons, positive for neurofilament protein. Schwann cells, positive for S-100 protein, were rare. A perineural cellular line positive for epithelial membrane antigen, EMA was present. No floating histiocytes were detected by CD68 in the myxoid matrix. Ki-67 immunostaining did not reveal any cell proliferation.
The overall histological features, together with the clinical and radiological profile were consistent with the diagnosis of "peripheral nerve mucoid degeneration".

## Follow-up

After surgical neurolysis, the painful symptoms decreased, although the left leg impairment remained. At the clinical examination, eight months after surgery, the patient resulted unchanged in signs of sciatic nerve paresis, while MRI showed a slight increase in the extension of the radiological altered signal of sciatic nerve. Two years after surgery, the patient presented with an almost normal neurological picture.

## Discussion

PNMD is a degenerative lesion usually appearing as a response to chronic nerve injury, caused by repeated traumatic stimulation.
PNMD can be etiologically related to other reactive lesions such as *traumatic neuroma, localized interdigital neuritis* (or Morton neuroma), *ganglion cyst*, *pacinian neuroma*, *intraneural injury neuroma*.
Some authors suggested a relation between PNMD, *Morton neuroma* and *traumatic neuroma* [5]: several events following experimental nerve transaction, occurring in axons during the wallerian degeneration or other chronic trauma, are at the basis of those three entities [6]. In addition PNMD and nerve lesion after ischemia-reperfusion injury can share similar pathogenesis [7].
Neural damage, caused by a near joint position, similarly to what happens in *intraneural ganglion cyst*, has been ruled out in the genesis of PNMD [8]. Nevertheless, the mucin deposits that characterize PNMD are most probably the result of an injury.
PNMD usually involves small peripheral nerves, while major nerves, such as the sciatic nerve, are usually affected by different inflammatory or neoplastic conditions.
Histologically, several reactive, inflammatory or neoplastic, lesions can show a myxoid matrix, miming PNMD. In the present case, inflammatory and reactive neoplastic lesions were excluded since PNMD usually shows no signs of inflammation or reactive proliferation changes. Benign and malignant peripheral nerve sheath tumors were easily excluded since no cellular proliferation was seen in PNMD.
In conclusion, the present case is, to the best of our knowledge, the first report of PNMD involving the sciatic nerve. Although uncommon, diagnosis of PNMD should be always kept in mind when dealing with symptomatic swelling of the major nerves.

### Conflict of interest statement

None declared.

## References


1 Allieu PY, Dersarkissian Y, Teissier J, et al. *9 case of mucoid nerve degeneration. Complication of a tunnel syndrome?* Rev Chir Orthop Reparatrice Appar Mot 1987;73:243-8.

2 Allieu PY, Cenac PE. *Peripheral nerve mucoid degeneration of the upper extremity*. J Hand Surg Am 1989;14:189-94.

3 Meier C, Bischoff A. *Focal mucoid degeneration of peripheral nerve. Light- and electronmicroscopic observation in a sural nerve biopsy of a case of progressive neural muscular atrophy (Charcot-Marie-Tooth)*. Acta Neuropathol 1977;31;37:69-72.

4 Nebuchennykh M, Løseth S, Mellgren SI. *Aspects of peripheral nerve involvement in patients with treated hypothyroidism*. Eur J Neurol 2010;17:67-72.

5 Scheithauer DBW, Antonescu CR, Woodruff JM. *Tumors of the peripheral nervous system, AFIP*. In: *Atlas of Tumor Pathology*. Arlington, VA: American Registry of Pathology, Series IV, Vol. 19, 2013.

6 Faweett JW, Keynes RJ. *Peripheral nerve regeneration*. Annu Rev Neurosci 1990;13:43-60.

7 Hautz T, Hickethier T, Blumer MJ, et al. *Histomorphometric evaluation of ischemia-reperfusion injury and the effect of preservation solutions histidine-tryptophan-ketoglutarate and University of Wisconsin in limb transplantation*. Transplantation 2014;98:713-20.

8 Rezzouk J, Durandeau A. *Nerve compression by mucoid pseudocysts: arguments favoring an articular cause in 23 patients*. Rev Chir Orthop Reparatrice Appar Mot 2004;90:143-6.


Received: March 6, 2019 - Accepted: May 20, 2019