A new pattern of lipomatosis of nerve: case report

Nikhil K. Prasad, MBChB,1 Mark A. Mahan, MD,3 Benjamin M. Howe, MD,2 Kimberly K. Amrami, MD,2 and Robert J. Spinner, MD1

Departments of 1Neurologic Surgery and 2Radiology, Mayo Clinic, Rochester, Minnesota; and 3Department of Neurosurgery, Clinical Neurosciences Center, University of Utah, Salt Lake City, Utah

Lipomatosis of nerve (LN) is a rare disorder of peripheral nerves that produces proliferation of interfascicular adipose tissue. It may be associated with soft-tissue and bony overgrowth within the affected nerve territory. LN has been almost exclusively reported in appendicular peripheral nerves; the median nerve at the wrist and palm is among the most common locations. The authors present a new pattern of LN that shows circumferential proliferation of fat around the epineurium of the nerve. They believe that this case and the two other documented examples in the literature (also affecting cervical and thoracic spinal nerves) share the same new pattern of LN. Defining the full spectrum of adipose lesions of the nerve and establishing a cause-effect relationship with nerve-territory overgrowth disorders may offer options for future management through targeted nerve lesioning.

https://thejns.org/doi/abs/10.3171/2016.2.JNS151051

KEY WORDS lipomatosis of nerve; osseous hypertrophy; segmental overgrowth; peripheral nerve

Lipomatosis of nerve (LN) is a “benign overgrowth of epineurial fibroadipose tissue,”1 previously referred to as fibrolipomatous hamartoma, lipofibromatous hamartoma, and fibro-fatty tumor. Approximately 50% of cases are associated with nerve-territory overgrowth; this hypertrophic tissue variably takes the form of diffuse lipomatous and osseous proliferation,12 including focal benign tumors such as lipomas or osteochondromas.7 LN of the median nerve at the wrist and palm is most frequently reported; the reasons for this frequency are that the soft-tissue and bony overgrowth may produce impressive macrodactyly in the nerve territory and that the median nerve is compressed at the transverse carpal ligament, leading to neurological symptoms. LN has a classic appearance on MRI, with proliferation of interfascicular adipose tissue that produces a sausage-like swelling of the nerve and a splaying of nerve fascicles.12

In this paper, we describe a new pattern of LN in which fat is seen circumferentially around the epineurium of spinal nerves associated with soft-tissue and bony overgrowth in the nerve territory. We critically reviewed the literature and tested our hypothesis on the most recently published case to strengthen the association of circumferential fat around spinal nerves and nerve-territory overgrowth.

Case Report

First Presentation

History and Examination

A 19-year-old woman presented with tingling and numbness in the left hand, with no dermatomal distribution pattern, and an associated soft-tissue mass in the posterior triangle of the neck. She had no history of steroid use, genetic disorders in the family, or physical findings of congenital overgrowth syndromes.

Imaging

On 1.5-T MRI, we observed a slow-growing “paraspinal lipoma” extending from the intervertebral foramen of the C-8 spinal nerve (Fig. 1A) toward the lateral mass of C-7 and between the paraspinous muscles of the neck beneath the trapezius. There was no evidence of osseous hypertrophy on CT (Fig. 2A).

Operation

The patient underwent a debulking procedure at our institution in 1993. A horseshoe-shaped paraspinous incision was made at the level of the T-1 vertebra, carried cephalad 1 cm below the mastoid and then anteriorly over the ster-
necleidomastoid. The trapezius was sectioned partially at the cephalad extent. The histopathologically proven lipoma was almost completely removed after finding the appropriate plane down to the point where it dove around the lateral mass of C-7 into the C-8 intervertebral foramen.

Second Presentation
History and Examination
The patient presented again in 2014 with focal pain at the site of the previous surgery. There was regrowth of a palpable and visible soft-tissue mass, consistent with lipoma, which extended along her lower neck and upper back. Her symptoms of radiating paresthesias into the ulnar digits had persisted over the years, but were described as not troublesome. Findings on neurological examination were normal. There was no percussion tenderness over the brachial plexus, and thoracic outlet maneuvers were negative.

Imaging
High-resolution 3-T MRI (Fig. 1B) showed that the lipomatous mass had regrown and was now associated with striking bony overgrowth (Fig. 2B). There was a circumferential proliferation of fatty tissue around the left-sided spinal nerves (C-7 to T-2) with extradural extension through the intervertebral foramen as far proximally as the spinal cord; distally it extended into the axillary sheath surrounding the brachial plexus trunks and into the pleural space between the first and second ribs. Distal extension was less easily distinguished from native fat in the axillary sheath and chest wall. The C-8 and T-1 spinal nerves were markedly hypertrophic compared with the contralateral side, with the T-1 spinal nerves exhibiting the most striking difference in diameters (9 mm on the affected side vs 2 mm on the contralateral side). This corresponded with significant nerve-territory osseous overgrowth of the T-1 transverse process and first rib and ankylosis of the first costovertebral joint (Fig. 2). Soft-tissue lipomas circumscribed the C7–T1 spinal nerves and expanded beyond the nerve territories along a posterosuperior plane between the paraspinal muscles of the neck (Fig. 1B).

Operation
The recurrent lipoma was resected by a different sur-
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The previous incision was nearly fully reopened from the posterior triangle of the neck to the paraspinal region posteriorly. The bulky dominant lipomatous masses extended from the spinal accessory nerve anteriorly to the spine posteriorly. They were partially encapsulated in the subcutaneous and deep regions and partially infiltrative within the paraspinal muscles. The histopathology was consistent with a benign lipoma.

Literature Review

A literature search was performed using PubMed and Google Scholar with the following key words: lipomatosis, lipoma, fibrolipomatous hamartoma, lipofibromatous hamartoma, and lipofibroma. These terms were combined using Boolean logic with the following key words: intrasacral, paraspinal, spinal nerve root, extradural, and intradural.

Review of the literature revealed 389 abstracts. Of these, 15 noted lipomas, spinal epidural lipomatosis, or lipomatosis affecting spinal nerves. We identified 3 cases, including one by our group demonstrating segmental thoracic lipomatosis with osseous hypertrophy and fatty infiltration of paraspinal muscles. Two cases had MRI evidence of fat encasing thoracic spinal nerves. In one case, MRI was not available, but the segmental pattern of bony overgrowth, together with the infiltration of fat into a subcutaneous/intermuscular compartment within the territory of the T-6 spinal nerve, would be consistent with LN. None of these 3 patients experienced symptoms from thoracic spinal nerve compression.

In an effort to challenge our hypothesis, we retrieved the MR images from the most recent case in the literature. We found several striking similarities between their case (Fig. 3), our current case (Fig. 1), and a previously reported case: the pattern of nerve-territory overgrowth and fatty proliferation surrounding specific nerves circumferentially along their anatomical course.

Discussion

We have described a case of segmental thoracic LN in association with nerve-territory osseous overgrowth. We believe that the “paraspinal lipoma” resected at the first presentation would be consistent with territorial lipomatous proliferation as part of the spectrum of overgrowth in LN. The expansion of soft-tissue lipoma beyond (but still centered around) the anatomical course of the T-1 spinal nerve (Fig. 1) may represent territorial overlap of spinal nerves or extraterritorial expansion of soft-tissue lipomas along planes of least resistance that were created by the first operation. The combination of adipose tissue proliferation (as well as the osseous hypertrophy) and nerve-territory overgrowth suggests that these findings are not coincidental. With significant heterogeneity in the nomenclature of lipomatous nerve lesions, it is necessary to define the full spectrum of clinical and imaging findings to accurately diagnose, treat, and prognosticate patients who present with LN and overgrowth. Establishing a cause-effect relationship between adipose lesions of nerve and nerve-territory overgrowth could provide an option to mitigate the impact of skeletal and soft-tissue hyperplasia by selective nerve lesioning.

The 3 cases of segmental thoracic overgrowth share several common features: 1) confined concentric proliferation of adipose tissue around and along the course of hypertrophic spinal nerves; 2) segmental nerve-territory osseous hypertrophy/ankylosis (affecting the corresponding vertebral bodies, pedicles, transverse processes, and ribs); 3) territorial lipomas/lipomatous overgrowth (in

FIG. 3. A: Sagittal T1-weighted MR image of the thoracic spine showing hypertrophic intercostal nerves (open arrows), hypertrophied ribs (plus sign), and fatty infiltration of the paraspinal muscles (asterisk). B: Axial MR image of the thoracic spine showing unilateral hypertrophy of the vertebral body, pedicle, and posterior elements (plus sign); hypertrophy of the adjacent rib (arrow); and fatty infiltration of the paraspinal muscles (asterisk) without involvement of the trapezius muscle (caret).
4, fatty infiltration of paraspinal muscles. The circumferential proliferation of fat around spinal nerves represents a new finding; this produces an MRI pattern different from classic LN in the distal limbs\(^2,8\) and different from extradural compression of spinal nerves by isolated intraforaminal lipomas.\(^4,5,13\)

Although the typical pattern of LN involves interfascicular fat proliferation, producing a spaghetti-like appearance on MRI,\(^2,8\) there are instances when fat proliferates around the nerve\(^12\) similar to the cases discussed in this report. We have previously highlighted the similarity of this pattern to lipomatosis of the digital nerves in the fingers.\(^6\) While there are insufficient data to determine which anatomical layer of the cervical/thoracic spinal nerves (epineurium or paraneurium\(^9\)) the lipomatosis in the 3 presented cases was confined to, it circumscribed the nerve(s) and proliferated along its anatomical course. We acknowledge a limitation of not having histopathological evidence to show the circumferential proliferation of fat around the spinal nerves. The appearance of fat by itself and fat in the nerve (as in LN) on MRI is pathognomonic.\(^8\) We debulked the lipomas to improve the patient’s cosmetic appearance and lessen her pain. More radical resection of the adipocytic lesion of nerve was not deemed clinically indicated.

The association of LN and the bony/soft-tissue overgrowth seen in these cases in the axial spine is analogous to cases of mild or severe bony and soft-tissue overgrowth in the appendicular skeleton, including, in extreme cases, macrodystrophia lipomatosa (MDL) (Fig. 4). It is not clear what order the epiphenomena of this rare disorder occur in, but it would be logical to assume that bony and soft-tissue overgrowth is a sequela of nerve pathology. Lipomatous tissue excised from MDL patients exhibits the same \(PIK3CA\) mutations seen in other overgrowth syndromes such as Proteus syndrome, hemihyperplasia syndrome, and Klippel-Trenaunay syndrome;\(^10\) however, the pattern of overgrowth in MDL is restricted to specific nerve territories.

We believe that the patterns outlined in the 3 presented cases suggest that LN does not need to exhibit a spaghetti-like appearance, i.e., interfascicular adipose tissue; rather, the association of nerve enlargement with circumferential fat and nerve-territory overgrowth is sufficient for diagnosis. This finding represents an expansion of the spectrum of LN, which may unify other cases of adipose overgrowth.

**Acknowledgments**

We thank David Factor for his illustration.

**References**

3. de Poorter JJ, Kroon HM, Dijkstra SP: Hyperplasia of rib and vertebra, associated with infiltrating lipoma: a rare case

Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Spinner, Mahan. Acquisition of data: Spinner, Amrami. Analysis and interpretation of data: all authors. Drafting the article: Spinner, Prasad, Mahan. Critically revising the article: all authors. Reviewed submitted version of manuscript: Spinner, Prasad.

Correspondence
Robert J. Spinner, Department of Neurologic Surgery, Mayo Clinic, Gonda 8-214, Rochester, MN 55905. email: spinner.robert@mayo.edu.