Lipomatosis of nerve (LN) is a spectrum of adipose lesions of nerves[1], wherein the nerve is enlarged by fibrofatty proliferation within the epineurium[2, 3]. This has been variously called lipofibromatous hamartoma, fibrolipomatous hamartoma, intraneural fibrolipoma, fibrofatty tumor, among other names, and was recently redefined by the World Health Organization as lipomatosis of nerve[4].

LN is a histologically benign condition which may be variably associated with overgrowth phenomena within the nerve territory, such as macrodactyly[5]. The combination of massive enlargement of nerve, adipose tissue and skeletal overgrowth is routinely referred to as macrodactylosis lipomatosa (MDL). LN is unique in that it is not part of any other overgrowth syndrome like Proteus or congenital lipomatous overgrowth, vascular malformations, and epidermal nevus (CLOVE) syndrome. LN is most commonly found in the median nerve as well as digital nerves of the hands or feet. We sought to characterize the clinical manifestations of this disorder through review of cases at a single institution.

Methods

Electronic records from our institution’s clinical patient database from 1992 to 2012 were searched for candidates for study inclusion. Patients were found by using the diagnosis keywords: “lipomatosis of hamartoma,” “fibrolipomatous hamartoma,” “macrodactyly,” “hyperplasia,” and “fibroproliferative,” as many practitioners may misdiagnose LN as a different hypertrophy syndrome. All records containing some or all of these terms were reviewed to determine patients to be included in this study. Additionally, patients known or suspected to have LN from paper records or other materials predating 1992 were included for possible inclusion. Exclusion criteria included patients that were found to have dermatologic findings including cafe-au-lait spots, axillary freckling, cerebriform connective tissue nevi, vascular or lymphatic nevi; a history of arteriovenous fistulae or malformations; or pathology, genetic mutation analysis or pedigree consistent with neurofibromatosis or other heritable disorder.

Medical records were reviewed and coded for nerve territory involvement; presence or absence of bone overgrowth, soft tissue overgrowth or other deformities; location of macrodactyly; and, if macrodactyly was present, which digital nerves were afflicted. Patient histories regarding date(s), indication(s), type(s) of surgery, outcome(s) and complication(s) and pathology report(s) were collected.

Results and conclusions

Review of medical records discovered 52 cases of LN, based on either pathologic review or pathognomonic appearance on MRI.

Detailed investigation into several clinical questions revealed the following conclusions:

- Surgical injury of LN appears to be strongly associated with the development of fibroproliferative neumomas. This was not seen in decompression surgeries for LN.
- Serial MR imaging reveals progressive enlargement of LN. The rate of growth was more profound in youth but also occurred in early adulthood.
- LN is not merely a finding in the distal extremities, but may be present in axial nerves, with resultant pleotrophic effects.
- LN may be associated with various forms of osseous overgrowth, ranging from involvement of joint related extostoses to osteochondromas of appendicular long bones.

References

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