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Spinal Neural Fibrolipoma: A Case Report and Literature Review of a Rare Pathology in Our Practice

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Abstract
Introduction: *Neural fibrolipoma is a benign tumor characterized by infiltration of epineurium by adipose and fibrous tissues. This tumor is commonly seen in peripheral nerve but occurring and involving the spinal nerves is extremely rare.*
We report a case of spinal neural fibrolipoma in a 35 year old man who presented with paraplegia and bi-sphincteric incontinence of more than a year duration. Examination findings revealed muscle power of 0/5 in both lower limbs and sensory level was T2. Magnetic Resonance Imaging showed an intradural extra-medullary mass from C6 –T3, which severely compressed the spinal cord. Preoperative diagnosis of intradural lipoma was made. He hadtumor excision and histopathological examination showed infiltration of perineural/neural tissues by variable amounts of mature adipose tissue and fibrous tissue and diagnosis of spinal neural fibrolipoma was made. Six months after the excision patient has no functional improvement in neurologicaldeficit.
Conclusion: *Neural fibrolipoma is a benign tumor but the outcome in our patient is not encouraging and this is likely due to delayed presentation. Although spinal case is rare, it should be included in differential diagnosis of extra-medullary intradural lesions.*
Keywords:*neural, fibrolipoma, spine*

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I. Introduction

Fibrolipomatous hamartoma of nerve also known as neural fibrolipoma is a rare and poor delineated lesion. It commonly affects peripheral nerve. Median nerve is the most commonly affected followed by ulnar nerve. (1, 2) The lesion grows within and around the affected nerve bundle thereby causing enlargement. (3) In 1953, Mason first reported in English literature of a patient with neuroma of the median nerve and another patient with a fusiform mass involving the median nerve. (4) Fibrolipoma was histologically identified as intraneural lipoma because the fibers of the median nerve were spread out over the surface of a fatty tumor containing very scant fibrous supporting stroma. (5, 6, 7) Various names have been proposed for this lesion, among which include; neural fibrolipoma, fibrolipomatosis of nerve, fibrolipomatous hamartoma of nerve, lipofibroma of nerve, and neurolipomatosis. The classification published by World health organization, WHO in 2002 referred to this benign tumour as lipomatosis of nerve. (3) Neural fibrolipomatous lesion arising from cervico-thoracic nerves and extending to intradural is extremely. We are reporting a patient with epidural and intradural cervico-thoracic spinal nerves fibrolipoma.

II. Case report

A 35 year old man presented with progressive weakness which culminated into inability to walk foroneyearduration.Examinationfindingsrevealedpowerof0/5inbothlowerlimbsandsensory level was T2.

Magnetic Resonance Imaging showed an intraduaral extramedullary mass from C6–T3, which was severely compressed the spinal cord (Figure 1).

The patient had laminectomy and cervico-thoracic stabilization. The intraoperative findings were anextraduralmassaroundtherightexitnerverootsconnectingtheintradural componentthrough a duralrent.

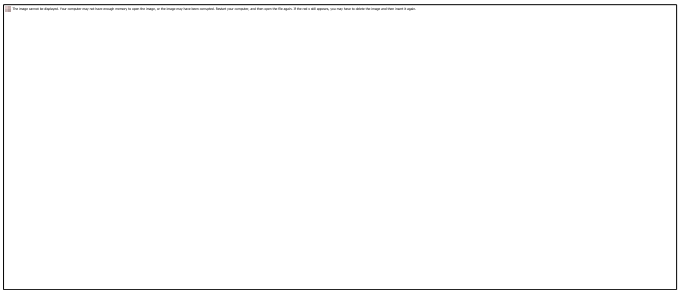

Figure 1:MRI Images. **Left:** Sagittal T2-image demonstrating hyperintense extramedullary intradural mass extending from C6 to T3. **Middle:** T1-image showing a hyperteintense lesion.
Right: AxialT2-imageshowingextramedullarymassoccupyingposteriorandrightantero-lateral region.

Histological evaluation showed an extradural components comprised of relatively circumscribed lesion composed of sheet adipocytes interspersed by fibrovacular tissue and the intradural component shows marked infilteration of an enlarged nerve bundles by fibroadipose tissue with area of pseudo-onion bulb formation and extensive perineural fibrosis. (Figure 3)
One month after the surgery, patient's clinical and neurological examination remain the same.

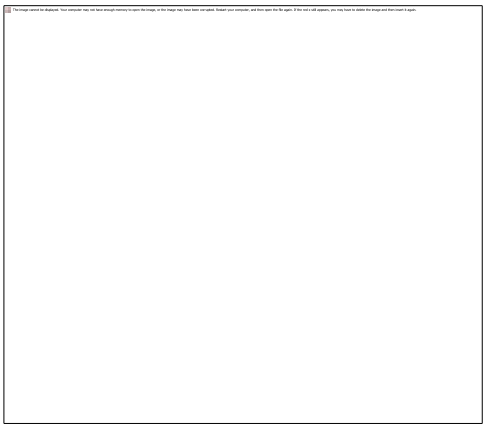

Figure 2: Intraoperative images (pre- excision and post-excision of the tumor

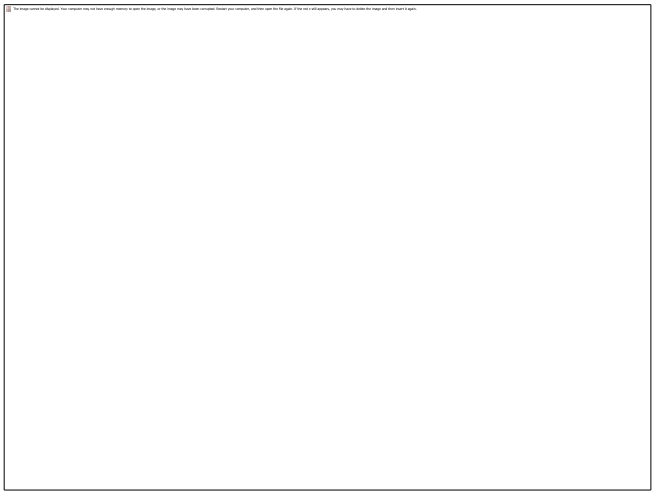

Figure 3. Histologic sections showing infiltration of perineural/neural tissues by variable amounts of mature adipose tissue and fibrous tissue (A-C). Areas showing pseudo-onion bulb formations (arrows) are noted (D). (H&E x100)

III. Discussion

Neural fibrolipoma of the nerve is a benign tumor of peripheral nerves originating from the perineural tissue. It is also variously referred to as fibrolipomatosis of the nerve, fibrolipomatous hamartoma or perineural lipoma, though there are distinct features inherent in each. This tumour is characterized by varying degree of fibrous and fatty tissue that grows between entrapped fascicles that are macroscopically normal. The resultant effect is the fusiform enlargement of the nerve. (8. 9) The lesion is made up of normal components of epineurium and because of this, it is consideredhamartomaofnervesheath.Mostoftheneuralfibrolipomaarefirstnotedatbirthorin early childhood of life. The age range of the patients is between 11 -39 years.(3)

The tumor localizes more frequently in median nerve followed by ulnar nerve and can also occur in deep soft tissue. (1, 2, 10) Unusual sites like brachial plexus and cranial nerves have also been reported to harbor neural fibrolipoma. (11, 12) Very few cases of neural fibrolipoma have been reported in literature; one involved the intradural space of lumbar spine by Lee et al and another case was in the spinal canal by Majchrzak et al. (13, 14)

Ourpatient'spathologicalreportshowedanextradurallesionwithsheetofadipocytesinterspersed by fibrovascular tissue and intradural lesion with marked infilteration of enlarged nerve bundles with fibroadipose tissue, area of pseudo-onion bulb and perineural fibrosis. This report is fairly comparable with other histological reports; massive epineural and perineural fibrosis surrounding and compressing nerve bundles by Silver manetal.(2).The neural tissue and fibrosis were shown to be interspersed in harmatomatous fatty tissue within the affected nerve sheath by Cavallaro et al. (15)

Intraneurallipomaandlipofibromatosisarethemaindifferentialdiagnosesthatareoftenconfused withneuralfibrolipomabuteachhavingitsdistincthistologicalcharacteristics.Intraneurallipoma occurs in $4^{th}$to 5^{th}decade of life. It is well encapsulated and histologically composed of matured fibroadiposetissue.(16)Incontrast,neuralfibrolipomaarediffuselyinfiltrative.Lipofibromatosis is a fibrofatty tumor of childhood, characteristically composed of alternating streaks of mature adipose tissue and a fibrous spindle cell component mainly involving thesepta of adipose tissue. (17) Distinctly, neural fibrolipoma is made up of enlarged nerve bundles infiltrated with fibrous tissue and nerve shows pseudo-onion bulb and peri neuralfibrosis.

IV. Conclusion

We reported a case of neural fibrolipoma with severe spinal cord compression causing paraplegia inayoungman.Neuralfibrolipomaofaspinalnerveisextremelyrare,thoughthelesionisbenign. Complete excision may not be feasible, even at that, good prognosis has been recorded in few cases reported. However, with delayed presentation and prolonged compression of neural tissue, the expected good outcome after excision may not be possible as seen in our indexpatient.

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