

CASE REPORT

Spinal neural fibrolipoma: A case report and review of the literature

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ABSTRACT

Neural fibrolipoma is characterized by infiltration of the epineurium by adipose and fibrous tissue. Intradural spinal cases are extremely rare. We report an unusual case of spinal neural fibrolipoma. A 31-year-old pregnant woman presented due to weakness of right leg (muscle strength 2/5). Magnetic resonance (MR) evaluation of the spine revealed an extramedullary intradural mass at the T1-T4 level. MR findings were suggesting a dermoid cyst or a lipoma. Subtotal surgical excision of the tumor was done. Histopathological examination showed enlarged nerve bundles infiltrated by fibroadipose tissue. Thus, the diagnosis of neural fibrolipoma was established. One month after surgery, lumbar MR showed residual tumor tissue, but successful decompression of the spinal cord. Six months after surgery, the neurological examination, muscle strength 4/5 evaluated. Neural fibrolipoma is characterized by infiltration of the epineurium by adipose and fibrous tissue. The tissue grows between and around nerve bundles thereby causing enlargement of the affected nerve. Neural fibrolipoma is a benign lesion with no effective therapy. Surgical excision usually causes severe damage of the involved nerve. Although spinal cases are extremely rare, it should be included in the differential diagnosis of extramedullary intradural masses.

Key words: Hamartoma, pathology, spine

Introduction

Neural fibrolipoma (fibrolipomatous hamartoma, lipomatosis of a nerve) is characterized by infiltration of the epineurium by adipose and fibrous tissue. The tissue grows between and around nerve bundles thereby causing enlargement of the affected nerve.^[1] Mason initially described neural fibrolipoma in the English literature in 1953.^[2] The median nerve and its digital branches are most commonly affected, followed by the ulnar nerve.^[3,4] Intradural spinal cases are extremely rare.^[5]

Case Report

A 31-year-old pregnant woman presented due to the weakness of the right leg (muscle strength 2/5). Magnetic resonance imaging (MRI) of the spine revealed an extramedullary intradural mass at the T1-T4 level measuring 1.5 cm × 6.5 cm [Figure 1]. MR findings were suggestive of a dermoid cyst or a lipoma. Subtotal surgical excision of the tumor was done [Figure 2].

Histopathological examination showed enlarged nerve bundles infiltrated by fibroadipose tissue [Figure 3]. The nerves showed pseudoonion bulb formation and perineural fibrosis [Figure 4]. The nerve bundles immunostained with S-100 protein [Figure 5]. Thus, the diagnosis of neural fibrolipoma was established. One month after surgery, lumbar MRI showed residual tumor tissue but successful decompression of the spinal cord. Six months after surgery, the neurological examination, muscle strength 4/5 evaluated.

Discussion

Neural fibrolipoma is frequently first noted at birth or in early childhood, but patients may not present for treatment until early or mid-adulthood. The patients range in age from

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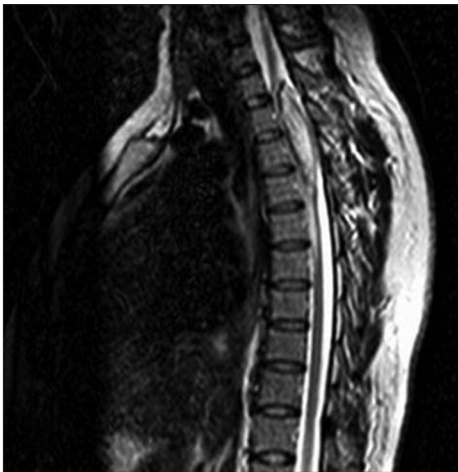


Figure 1: Extradural intradural mass. Sagittal T2-weighted magnetic resonance image of the thoracic spine demonstrates a heterogenous hyperintense intradural extramedullary mass at the T1-T4 level

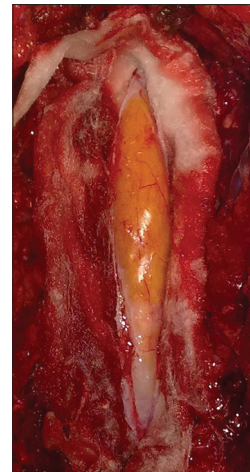


Figure 2: The mass as revealed at surgery

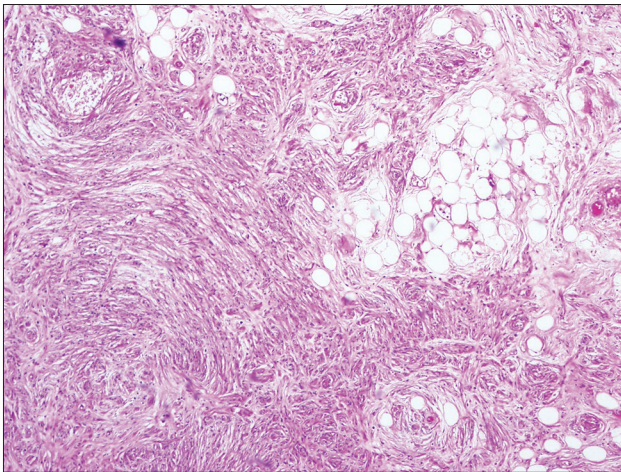


Figure 3: Enlarged nerve bundles infiltrated by fibroadipose tissue (H and E, x40)

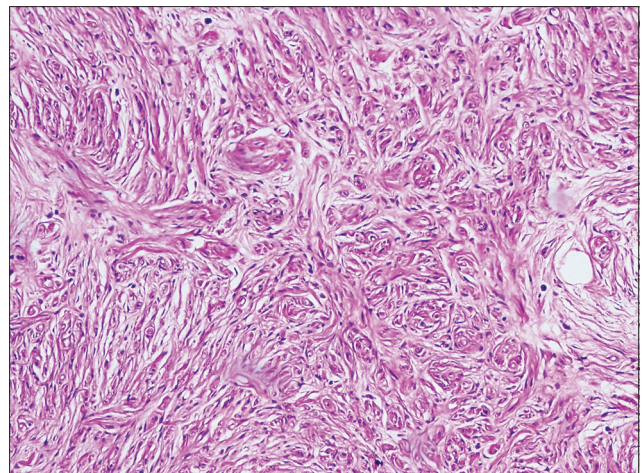


Figure 4: Nerves showing pseudoonion bulb formation and perineural fibrosis (H and E, x100)

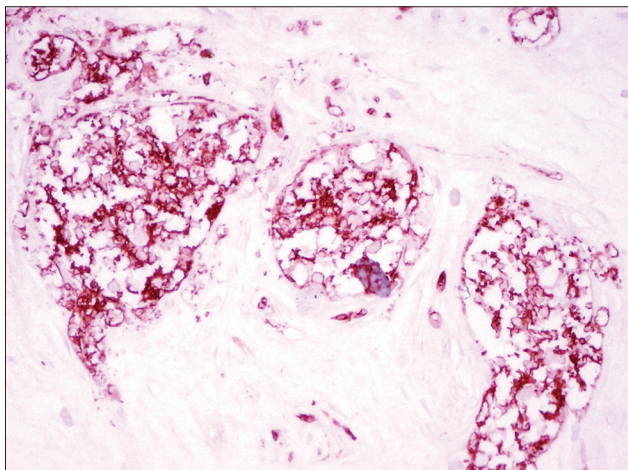


Figure 5: Immunostaining of nerve bundles with S-100 protein (x200)

11 to 39 years. Because the constituent tissues are normal components of the epineurium, some have considered this

lesion to be a hamartoma of the nerve sheath. In some cases it is associated with macrodactyly of the digits innervated by the affected nerve. Females predominate when lipofibroma is accompanied by macrodactyly, whereas males are more commonly affected when macrodactyly is absent.^[1]

The median nerve and its digital branches are most commonly affected followed by the ulnar nerve.^[3,4] The process has also been reported to involve unusual sites such as the cranial nerves and the brachial plexus.^[6,7] Very rarely, the lesion is found in other nerves such as the nerves of toes.^[8] Only one case of fibrolipomatous hamartoma involving the intradural space of lumbar spine was described in the relevant literature.^[5]

Magnetic resonance imaging characteristics may reflect the histology of the tumor. The individual nerve fascicles and surrounding fibrosis result in long cylindrical bands of low T1- and T2-weighted signal.^[9] The differential diagnosis of this tumor includes intraneural lipoma, ganglionic cyst, traumatic neuroma, and vascular malformations.^[10]

Pathologically, the tumor is characterized by fibrofatty enlargement of the nerve, usually confined by the nerve sheath. There is massive epineural and perineural fibrosis surrounding and compressing individual nerve bundles.^[4] The nerves and surrounding fibrosis are interspersed in hamartomatous fatty tissue, usually confined to the nerve sheath.^[10,11]

Differential diagnosis includes intraneural lipoma and lipofibromatosis. Intraneural lipomas are well encapsulated. They tend to occur in the fourth and fifth decade, with female predominance. Intraneural lipomas are usually well encapsulated, and complete excision can be achieved without damage to the adjoining nerve.^[12] Histopathologically they are composed of mature fibroadipose tissue.^[13] Neural fibrolipomas are diffusely infiltrative. They arise in a younger age group.^[12] Lipofibromatosis is a histologically distinctive fibrofatty tumor of childhood. Microscopically the tumor is composed of alternating streaks of mature adipose tissue and a fibrous spindle cell component mainly involving the septa of adipose tissue.^[14] Neural fibrolipoma is composed of enlarged nerve bundles infiltrated by fibroadipose tissue. The nerves show pseudoonion bulb formation and perineural fibrosis.

The etiology of neural fibrolipoma is unknown. It is not associated with any syndrome nor is there any known hereditary predisposition.^[1] Although neurofibromas may have rapid episodes of growth associated with pregnancy,^[15] there is no information about such an association for neural fibrolipomas in the literature.

Conclusion

We presented here a very rare case of fibrolipomatous hamartoma located in the spinal cord. It caused weakness of the right leg during pregnancy. Successful decompression of the spinal cord was achieved by subtotal excision.

Neural fibrolipoma is a benign lesion with no effective therapy.^[1] Complete excision is not feasible as it usually leads to a neurological deficit.^[16] Although spinal cases are extremely

rare, it should be included in the differential diagnosis of extramedullary intradural masses.

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