Case Report

Spinal intradural subpial angiolipoma: Case report and review of literature

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Abstract

Background: Spinal angiolipomas are rare tumors consisting of mature adipose tissue and abnormal vascular elements. Intradural location is very rare, and till now, only seven cases have been reported in literature. Authors report a case of an intradural intramedullary (subpial) angiolipoma located in the thoracic cord.

Case Description: A 26-year-old patient presented with features of progressive myelopathy of relatively short duration. Imaging showed a heterogeneous fat-containing intradural lesion at D5-D9 level, which enhanced on contrast enhanced fat saturation sequences. Subtotal excision was performed and patient had partial recovery of his neurological deficits. Histopathology was suggestive of angiolipoma.

Conclusions: Intradural angiolipomas are very rare. Complete excision often leads to neurological deficits. Hence, safe maximal decompression would suffice leading to long-term recurrence-free periods.

Key Words: Intradural, spinal angiolipoma, subpial

INTRODUCTION

Spinal angiolipomas (SAL) are rare tumors constituting around 1% of all spinal tumors.1,4,8,9 They are most commonly located in the thoracic spine in the extradural compartment. Only seven intradural cases have been reported till date.1,4,5,9 Authors report a case of thoracic intradural subpial tumor in a middle-aged male patient presenting with progressive myelopathy. Near total excision was performed and histopathology features were compatible with angiolipoma.

CASE REPORT

A 26-year-old male patient presented with progressive spastic paraparesis with autonomic involvement of 6 months duration. There was bilateral lower limb spasticity with power of around 3/5 (Medical Research Council grading) and a sensory level at D3 dermatome. Magnetic resonance imaging (MRI) showed features of a heterogeneous intradural intramedullary mass lesion extending from D5-D9 level. The posterior part was hyperintense on both T1 and T2 sequences while the anterior part was isointense on T1 and hypointense on T2-weighted sequences. The hyperintense signal of posterior part showed suppression on fat saturation images reflecting the presence of fat and the anterior part showed irregular enhancement on contrast administration [Figure 1a-c]. A D5-D9 en bloc laminoplasty was performed. A subpially located lesion consisting of both fat and blood vessels was present and a near total excision was performed. On histopathology,
features were compatible with angiolipoma, consisting of mature fat cells mixed with vascular channels. Postoperatively, patient had initial deterioration of power in both lower limbs but made a gradual recovery.

**DISCUSSION**

Angiolipomas are benign mesenchymal tumors consisting of both mature adipose tissue and abnormal blood vessels.\(^6\) Primary SAL was first reported in 1901 and Lin et al. categorized angiolipomas into noninfiltrating and infiltrating types.\(^{4,8}\) SAL are usually noninfiltrating types but the infiltrating angiolipomas can be either vertebral or intramedullary. They constitute around 0.4–1.2% of all spinal tumors and 20% of spinal lipomas are not associated with spinal dysraphism.\(^{8,9}\) They are almost always extradural tumors in the thoracic location. Intradural location is very rare with only seven cases reported in the literature till now [Table 1].\(^{1-5,9}\) Exact etiopathogenesis of these lesions remains unknown. They might probably represent an intermediate stage between lipomas and hemangiomas as they are believed to originate from the same progenitor cells similar to these two lesions.\(^1\) A more recent theory postulates that early inclusion of pluripotent stem cells into the neural arch causes SAL while late inclusion of mature fat cells causes lipoma.\(^7\) SAL are most commonly seen in women in their fourth and fifth decades, which is slightly longer than that of intradural subtypes (mostly seen in females in third decade).\(^{5,8}\) They are extremely rare in children.\(^3\) Extradural tumors mostly present with progressive neurological deficits, usually spanning several months to years but the presenting duration of intradural subtypes is typically short.\(^{8}\) As noted in Table 1, the average duration of presentation is 6.12 months (range 2–12 months). Symptoms are also reported to be exacerbated by obesity, pregnancy, and steroid treatment.\(^{1,8}\)

MRI is the investigation of choice for these lesions. Intradural angiolipomas usually span 4–6 vertebral levels in length, average being 4.5 levels [Table 1]. Generally, they are heterogeneously hyperintense on T1 sequences and possess variable signal intensities on T2 sequences.\(^{1,6,8}\) The proportion of fat in the lesion probably accounts for this variability. Conventionally, vascular flow voids are not seen, but, in cases with thick walled vessels, they may be identifiable on MRI. Contrast enhancement on fat saturation sequences is the characteristic hallmark. The most important differential diagnoses include lipoma and lipomatosis, liposarcoma, metastases, lymphoma, meningioma, and nerve sheath tumor.\(^{1,6}\) On histopathology, the tumor consists of varying amounts of mature fat cells admixed with abnormal vascular elements ranging from capillaries, sinusoidal vessels to venules and arteries. They do not exhibit mitosis and pleomorphism.\(^{1,8}\) Treatment of SAL is surgical removal.\(^{1,8}\) Complete removal can be achieved in almost all cases of noninfiltrative extradural subtypes. Intramedullary angiolipomas are usually subpial

### Table 1: Literature review of spinal intradural angiolipoma

| Author    | Age/sex | Duration       | Location | Surgery    | Result                        |
|-----------|---------|----------------|----------|------------|-------------------------------|
| Palkovic\(^4\) | 27/M | 5 months       | C6-D2    | Partial    | C8 nerve root palsy; Follow up NA |
| Weill\(^9\)    | 27/F  | Several weeks (exact NA) | D5-D8   | Biopsy    | Unchanged                     |
| Preul\(^8\)   | 36/F  | 1 year         | D7-D11   | STE        | Improved                      |
| Maggi\(^3\)   | 8/F   | 3 months       | D11-L2   | CE         | Improved                      |
| Klisch\(^2\)  | 34/F  | 8 weeks        | C6-D4    | Partial    | Improved                      |
| Garg\(^1\)    | 26/M  | 3 months       | D3-D7    | STE        | Partial improvement           |
| Prasad\(^8\)  | 28/M  | 1 year         | C6-D2    | STE        | Partial improvement           |
| Prasad\(^8\)  | 26/M  | 6 months       | D5-D9    | NTE        | Partial improvement           |

NA: Not available, STE: Subtotal excision, CE: Complete excision, NTE: Near total excision, F: Female, M: Male

Figure 1: (a) Sagittal T1-weighted MRI showing heterogeneous intradural subpial lesion from D5-D9 level. The anterior part is isointense (arrowhead) while posterior part is hyperintense (bold arrow); (b) Sagittal T2-weighted MRI showing the anterior part to be isointense with internal hypointensities (bold arrow) while posterior part remains hyperintense; (c) Sagittal postcontrast T1-weighted fat saturation images showing the signal suppression of the posterior part, while irregular enhancement can be noted in the anterior part (bold arrow)
in location and differentiation from normal tissue is often difficult. As noted in Table 1, only one case had a complete excision.\(^3\) No attempt should be made to define the tumor–normal tissue interface. The recurrence risk is very low; hence, a subtotal excision would probably suffice, achieving long-term control and progression-free periods.\(^5\) No adjuvant treatment is required.\(^6\) Prognosis is usually excellent in the extradural variety and good in intradural subtypes.

**CONCLUSIONS**

Spinal intradural angiolipomas are benign, extremely rare tumors. Contrast enhancement on fat saturated sequences along with heterogeneous signal on T1- and T2 weighted sequences are often the diagnostic features. Complete excision often leads to neurological deficits. Hence, safe maximal decompression would suffice leading to long-term recurrence-free periods. No adjuvant therapy is required.

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