Case Report

Spinal Cord Intramedullary Angiolipoma: Case Report and Literature Review

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The spinal intramedullary angiolipoma is extremely rare when compared to other intramedullary spinal tumors. The spinal intramedullary or extramedullary angiolipoma are conventionally divided into non-infiltrating and infiltrating subtype. The authors reported a case of intramedullary spinal cord angiolipoma in adult patient that presented with significant diagnostic challenges to pathologists because of its rarity. The clinical presentation, neurodiagnostic studies, surgical management, and histopathological findings of biopsy are documented in the present report.

Keywords: Spinal cord tumor; Intramedullary; Angiolipoma

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Angiolipoma is a rare benign soft tissue tumor with its most common location being in the subcutaneous tissue of the forearm. However, it can also occur in the spinal axis involving the vertebral body, epidural, intradural extramedullary, intracranial, and intramedullary regions. Most angiolipomas of spinal axis are located in the epidural space or the vertebral body[1]. Intramedullary spinal cord angiolipoma is an extremely rare benign tumor[1,2], which is composed of mature lipomatous and angiomatous components[3,4], and accounts for 0.1% to 0.5% of all spinal cord tumor[5].

Case Report

A 47-years old woman presented with numbness and heaviness of the left arm, loss of sensation of the right leg, and difficulties in raising her shoulder[6]. Patient had no evidence of spinal dysraphism[3]. The magnetic resonance imaging (MRI) showed a small well-defined intense enhancing intramedullary nodule in the left sided posterior cord at the C4 level, associated with a syringomyelia extending from the medulla oblongata down to the C4-C5 level (Figure 1)[7,8].

The pre-operative differential diagnoses included astrocytoma, ependymoma, and hemangioblastoma. However, the tumor should have been fat in the components, but the MRI showed that the tumors was not enhancing after fat suppression (Figure 1D)[3]. The tumor had the same density as the subcutaneous fat. The patient underwent surgery with total resection of the lesion. The gross section during intraoperative consultation showed a well-defined yellowish nodule (Figure 2).

Clinical presentation data are summarized in 37 patients[3] (Table 1). The clinical course of the patients with intramedullary angiolipomas is brief, or less than one year after onset. Most patients show paraparesis with hyperreflexia and bilateral plantar extensor response. There is a loss of pain, temperature, vibration, and position sensation.

Histopathologic examination of the mass revealed mature adipocytes with a network of atypical, thin-wall capillary-like vessels (Figure 3A)[2,4,9]. The immunohistochemical staining revealed negative expression for alpha-inhibin, CD10, S100, EMA, AE1/AE3, and GFAP. The vascular component such as the capillary-like vessels network, showed positive CD34, CD31 (Figure 3B), and ERG expression. The Ki67 was labeled from 0 to 1% of tumor nuclei. There was no evidence of malignancy. The histopathological diagnosis was that of a spinal intramedullary angiolipoma. The patient's clinical

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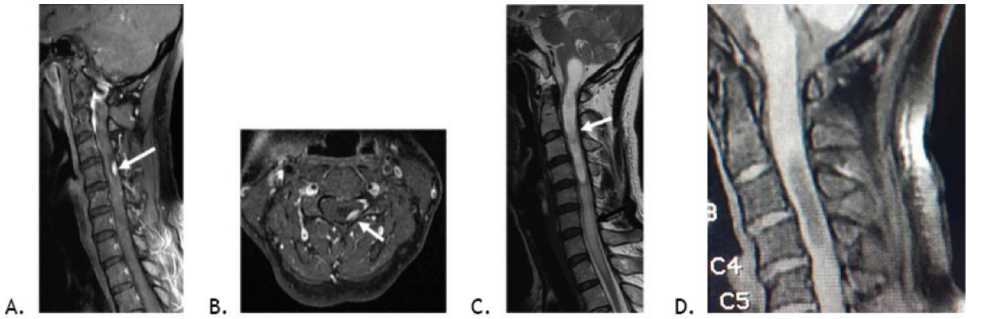

Figure 1. (A) Small well-defined intense enhancing intramedullary nodule in the left-sided posterior cord at C4-5 level with syringohydromyelia[7,9]. (B) Post-contrast T1W shows a small well-defined intense enhancing intramedullary nodule in the left-sided posterior cord at C4 level. (C) Syringohydromyelia extended from medulla oblongata down to C4-C5 level[7,8]. (D) Sagittal T1 FSE shows the tumor at C3-C4 level is not enhancing after fat suppression[3].

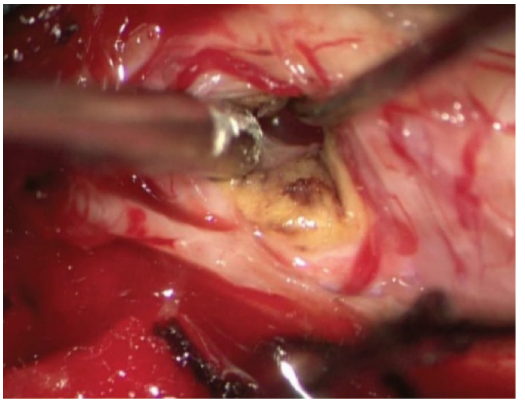

Figure 2. Intraoperative shows a well-defined yellowish nodule.

Table 1. Presentation in 37 patients with angiolipoma[3]

Presentation	Affected/studied; n (%)
Presenting system	
Paraparesis*	35/36 (97)
Sensory dysfunction*	30/32 (94)
Hyperreflexia & bilat Babinski's sign*	26/31 (84)
Sphincter dysfunction	19/37 (51)
Back pain	15/37 (41)
Initial complaint	
Back pain	14/37 (38)
Numbness or paresthesias	13/37 (35)
Leg weakness	12/37 (32)

* Not all data were available for all 37 patients

Two patients in this category were paraplegic

In several reports, there was simultaneous onset of more than one initial complaint

symptoms subsequently improved after surgery to the grade II of clinical and functional classification scheme, which was adapted from McCormick and Stein (Table 2)[10].

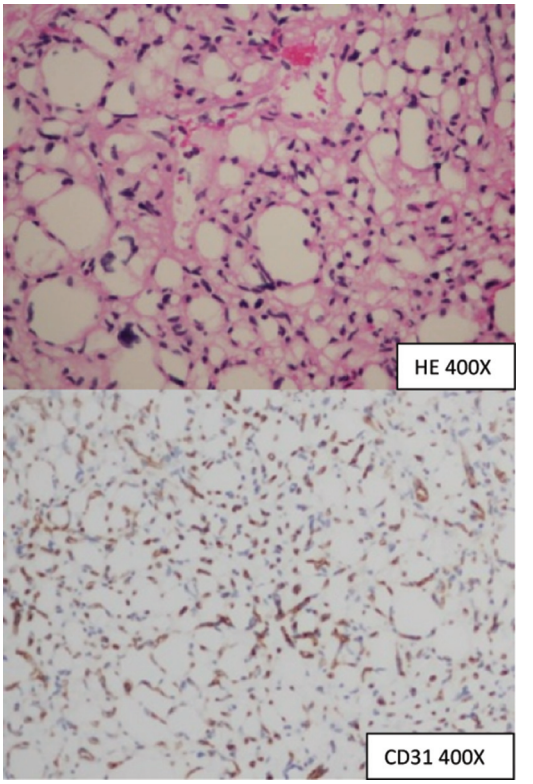

Figure 3. (A) H&E slide revealed mature adipocytes with a network of atypical, thin-wall capillary-like vessels[2,4,9]. (B) The immunohistochemical staining revealed positive expression of endothelial marker (CD31).

Surgical outcome[10]

All patients underwent subtotal resections with a goal of decompressing the spinal cord without inflicting damage to the nerve tissue. Because of intimate relationship between intramedullary lipoma and normal spinal cord, gross total resection was never attempted. The outcome was assessed using the

Table 2. Clinical and functional classification scheme[10]

Grade	Symptoms
I	Neurologically normal; mild focal deficit not significantly affecting function of involved limb; mild spasticity; normal gait
II	Sensorimotor deficit affecting function of involved limb; mild to moderate gait difficulty, severe pain or dysesthetic syndrome impairing patient's quality of life; walks independently
III	More severe neurological deficit; requires cane for ambulation; significant bilateral upper-extremity impairment; may or may not function independently
IV	Severe deficit; requires wheelchair, bilateral upper-extremity impairment; not independent

* Adapted from McCormick and Stein[10]

functional status grading scheme immediately at the post operative examination (Table 2)[10].

Discussion

Spinal angiolipoma is a rare lesion and accounts for 1% of all spinal cord tumor[2]. The usual location is in the cervico-thoracic region, mostly extradural. In the present case report, the authors document an adult female with a spinal intramedullary angiolipoma[10] at cervical region without clinical evidence of spinal dysraphism[10]. Maggi et al reviewed the literature on 40 cases of spinal angiolipoma, and all these angiolipomas were located in the epidural space, except for two cases, which were intramedullary[5]. Leiebscher first described the entity of intramedullary location of spinal angiolipoma in 1901[11]. Marbet et al reviewed 99 cases of cervical or dorsal intramedullary lipomas without spinal dysraphism, which have been reported since 1884. There is poor correlation between lipoma's length and age of onset and the finding of subcutaneous lipoma at the level of the lesion helps the diagnosis. The spinal intramedullary angiolipoma is exceeding rare. Mark et al reviewed the intradural lipomas. Only seven of the 51 cases in the review of all intradural lipomas were reported before 1957 by Caram et al that were true intramedullary lipoma. Additionally, Mark et al presented a series of six patients with intramedullary spinal cord lipoma that included three cervical, two cervicothoracic, and one thoracic. The usual clinical presentations included backpain and progressive numbness and weakness of the limbs due to spinal cord compression. The spinal cord intramedullary lipomas that unassociated with the spinal dysraphism is rare[10].

Spinal angiolipomas account for up to 1.2% of all spinal axis tumor[4]. Unlike spinal lipoma, spinal angiolipoma is not usually associated spinal dysraphism. Spinal cord intramedullary angiolipoma is extremely rare and accounts for 0.1% to 0.5% of all spinal tumors. The main complaints consist of sensory dysfunction, hyperreflexia, and spasticity. The tethered cord is another symptom that is known to be associated with spinal cord lipomas[6]. Angiolipomas are conventionally divided into non-infiltrating and infiltrating. The non-infiltrating type is encapsulated and well-demarcated. This type is more common than the infiltrating type. The infiltrating type is less common and can infiltrate the adjacent tissue even causing bone destruction[8,11]. The surgical approach to the case and the result are not different. The prognosis of infiltrating and non-infiltrating is the same.

There are hypotheses[1,3,5,6,12] proposed for the intramedullary location of angiolipomas including developmental error theory, metaplasia theory, and hamartomatous origin theory[2,11]. The intramedullary adipocytes are postulated to originate from progenitor cells of spinal vessels or due to the failure of defective neural crest cells to inhibit the formation of adipocytes from primitive mesenchymal cells[9]. Other hypotheses attempt to explain the presence of adipocytes within the spinal cord by raising the possibilities of that the adipocytes could derive developmentally from meningocytes, defect of the so-called meninx promotiva as a maldevelopment theory, somatic mesoderm, or even astrocytes[2,13].

In conclusion, from the practical point of view, spinal intramedullary angiolipoma is a diagnostic challenge for both clinicians and pathologists, unless physicians bear in mind the possibility of its existence. It is also a diagnosis of exclusion, which is usually considered after ruling out the more often encountered intramedullary spinal lesions such as astrocytoma, ependymoma, hemangioblastoma, and metastases.

Eighty percent of the angiolipomas display mutations in the protein kinase D2 (PRKD2) gene. The hypothesis is that altered PRKD2 signals the adipocytes formation, which is characterized of angiolipoma. Otherwise, activating PIK3CA mutations happens in sporadic angiolipoma.

What is already known on this topic?

The spinal intramedullary angiolipoma is

extremely rare and challenging for pathologists and surgeons to diagnose. The intramedullary angiolipoma can be detected earlier than the extradural lesion by about 9.5 months and the early surgery will decrease the neurodeficit(3). The MRI without gadolinium and the fat suppression will help to rule out differential diagnoses from the angiolipoma.

What this study adds?

The present case is a rare intramedullary angiolipoma module in spinal cord at C4-C5 and the mass was totally removed. The authors also reviewed literature and pathogenesis.

Conflicts of interest

The authors declare no conflict of interest.

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