Case Report

Intradural extramedullary cavernous hemangioma of the cervicothoracic junction: A case report and review of the literature

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ABSTRACT

Background: Intradural extramedullary cavernous hemangiomas of the spine are rare, benign lesions with only 40 published cases to date.

Case Description: The authors report a rare case of a histologically diagnosed intradural extramedullary cavernous hemangioma of the spine involving the cervicothoracic junction and causing sudden gait disturbances and urinary retention in a 24-year-old male. Gross total tumor removal allowed complete spinal decompression and sensible improvement of the clinical condition with no evidence of tumor relapse at 12-month follow-up examination.

Conclusion: More frequently found in the lower thoracic and lumbar spine, these tumors often cause subtle clinical manifestations including sensory and motor dysfunction secondary to nerve root compression; nonetheless, occasional cases of rapidly progressive worsening of the neurological condition with evidence of myelopathy and autonomic dysfunction have been described. In such cases, urgent surgical resection is crucial since the degree of neurological impairment and the time spanned from the onset of the symptoms are paramount for a good recovery.

Keywords: Case report, Cavernous hemangioma, Intradural spinal tumor, Vascular tumor

INTRODUCTION

Cavernous hemangiomas are uncommon, vascular tumors forming in virtually any area of the neuraxis with only 3% of cases localized intradurally. Of all spinal cavernous hemangiomas, intradural extramedullary tumors represent the rarest ones, with only 40 cases described to date, mostly as case reports, frequently located in the lower thoracic and lumbar level. Microscopic examination reveals unorganized, densely packed dilated blood vessels lined by a single layer of endothelium without interposed neural or glial elements, which exhibit frequent microhemorrhages and thrombotic obliterations that influence the clinical phenotype. Among the most common clinical manifestations, back pain is constantly reported accompanied...
by progressive nerve root compression; however, the presence of a sudden worsening of the general condition with the onset of signs of myelopathy and autonomic dysfunction has also been described.\(^1,5\)

In the present report, the authors describe a rare case of intradural extramedullary cavernous hemangioma located at the cervicothoracic junction causing rapidly progressive gait disturbances and acute urinary retention. Surgical posterior decompression and gross total tumor resection were followed by marked improvement of the clinical condition without radiological evidence of tumor recurrence at 12-month follow-up.

**CASE REPORT**

A 24-year-old male presented to the emergency department in March 2020 with acute urinary retention and fever without any other relevant medical history. The patient referred diffuse myalgia in the past 10 days and subsequent onset of painful dysesthesia in the left arm that was treated with corticosteroids. Physical examination revealed marked hyposthenia of the upper extremities, more pronounced in the left arm, and pain to deep manual palpation of the abdomen in the hypogastric region. Further examination disclosed a light spastic paraparesis and a decreased sensation of pain, position, and touch below T1 level, with negative Babinski’s signs bilaterally. Weak deep tendon reflexes (DTR) with slight difficulty in deambulation were also noticed. A urine sample was collected after the insertion of a Foley catheter, and microscopic examination revealed normal numbers of leukocytes and the absence of erythrocytes. Following the progressive worsening of the neurological state with sudden onset of gait instability, the patient was admitted to the neurological department where an emergency magnetic resonance imaging of the head and spine was performed. Spinal scans revealed an intradural, extramedullary tumor measuring 27 × 8 mm, located posterolateral to the spinal cord at C7-T1 level causing compression and dislocation without radiological evidence of myelopathy. The lesion was homogeneously hyperintense both in T1- and T2-weighted images, with dishomogeneous aspects in STIR sequences, suggesting a highly vascularized nature [Figure 1]. Therefore, the patient was transferred to the neurosurgical department and treated with posterior laminectomy without fusion of the C7-T1 segment followed by surgical monopolar cauterization of the tumor’s feeding artery and gross total tumor resection under constant neurophysiological monitoring [Figure 2]. During surgery, the lesion appeared as a dark red, solid tumor surrounded by a highly vascularized pseudocapsule. A cavernous hemangioma was diagnosed histopathologically. The surgical specimen consisted of a tumor with a loosely packed collection of hyalinized vessels exhibiting multiple thrombotic obliterations [Figure 3].

The postoperative course was uneventful and marked by the progressive improvement of the muscular strength as well as the absence of residual urinary retention. The patient...
was discharged 5 days after admission and referred to a rehabilitation center. At 12-month follow-up examination, the patient had good recovery of both motor and sensory functions, with no signs of residual hyposthenia of the upper extremities. Gait instability resolved completely and normal DTRs were observed. Contextual cervical spine magnetic resonance imaging (MRI) revealed no recurrence of the lesion and documented complete decompression of the spinal cord [Figure 4].

**DISCUSSION**

Intradural extramedullary cavernous hemangiomas of the spine are extremely rare tumors, with only 40 cases described to date. Between 3% and 16% of all vascular tumors of the spine are hemangiomas that can occur in the vertebral body with or without evidence of epidural extension, in the epidural space alone, and as intradural extramedullary as well as intramedullary lesions. Primarily located in the lumbar and lower thoracic spine, with only four cases of tumors described at the cervicothoracic junction, cavernous hemangiomas exhibit benign behavior, although hypertrophy and reorganization of aberrant cells can lead to progressive growth with subsequent mass effect on the surrounding structures. Secondary edema formation occurs most likely due to the interplay of thrombotic obliterations and recurrent microhemorrhages, which may be the cause of the progressive neurological decline. Frequently observed clinical manifestations include gradually progressive nerve root compression, and, in some reported cases, including the present, progressive myelopathy, leading to motor and sensory deficits. Furthermore, the pathological blood vessels that form and feed the lesion are markedly prone to bleeding, leading to the subarachnoid collection of fluid and hemosiderin, that is considered a crucial factor in the presentation of neurological symptoms as it exhibits neurotoxic effects.

In the intradural extramedullary space, hemangiomas can arise from either root nerves, blood vessels, the inner layer of the dura mater, or the pial surface of the neuraxis. Histologically, a cavernous hemangioma must be distinguished from a capillary hemangioma, which consists of a dense cluster of capillary size abnormal vessels coated by flat and hardened endothelium. On the other hand, cavernous tumors are formed by closely packed, large, and dilated blood vessels without interposed neural tissue, typically arranged in diffuse patterns. Microscopic examination of these lesions frequently demonstrates thrombosis, perivascular hemosiderin deposition, and calcification. However, some capillary hemangiomas can exhibit partial cavernous features, hence are referred to as transitional forms. The pathogenesis of this rare tumor remains unknown. Some authors have suggested that the origin may be secondary to the impaired migration and differentiation of fetal mesoderm from the embryonic mesodermal plate at the time of angioblastic differentiation (21–24 days of embryogenesis). Nonetheless, the presence of cases of familiar inheritance through several generations has led to speculating over a possible congenital origin of this malformation following an autosomal dominant pattern of transmission.

MRI is the investigation of choice for spinal lesions and thus also for intradural extramedullary tumors. Cavernous hemangiomas of the spine show clear distinctive features in imaging findings that differ from these observed in the two most common spinal tumors: meningiomas and schwannomas, as well as from capillary hemangiomas. Signal in T1- and T2-weighted images is variable, generally mixed, depending on the intensity of blood flow, the content of hemosiderin, and the presence of calcifications. Gradient echo sequences (T2) result more sensitive in revealing small hemorrhages and calcium deposits and thus are considered the gold standard to describe highly vascularized lesions. Contrast enhancement is variable, but generally poor, and peripheral in most of the reported cases, whereas the capillary subtype exhibits strong similarities with meningiomas as it even presents with a dural tail, posing problems of differential diagnosis. While capillary hemangiomas are normally associated with slowly progressive deterioration of the neurological state, cavernous hemangiomas may present with sudden onset associated with bleeding and spinal cord compression; therefore, urgent surgical resection is crucial since the degree of neurological impairment and the time spanned from the onset of the symptoms are a paramount for a good recovery. Despite the recurrence of this tumor is rare, occasional cases of relapse have been reported following partial resection; therefore, a combination of radiotherapy and subtotal tumor resection or gross total resection alone is proposed as goals of treatment for spinal cavernous hemangiomas.

**Figure 4:** Postoperative sagittal T2-weighted image demonstrating gross total tumor resection and laminectomy of the C7–T1 segment.
In the present report, the authors describe a rare case of intradural, extramedullary cavernous hemangioma located at the cervicothoracic junction in a 24-year-old patient. The patient presented with a short history of diffuse myalgia and painful dysesthesia of the left arm followed by sudden urinary retention and gait disturbances. MRI of the spine revealed a 27 × 8 mm tumor exhibiting dishomogeneous signal in T1- and T2-weighted images and peculiar features on STIR sequences. Posterior C7-T1 laminectomy followed by gross total resection of the lesion was performed and histological examination confirmed the diagnosis of cavernous hemangioma with thrombotic obliteration. Clinical and radiological follow-up showed complete decompression of the spinal cord without evidence of recurrence and full recovery of the motor and sensory function.

**CONCLUSION**

Despite the rareness, intradural extramedullary cavernous hemangiomas of the spine have to be taken into consideration as a potential cause in patients presenting with progressive spinal nerve compression syndromes or sudden signs of myelopathy. Although the interpretation of MRI may be complex, notably after an episode of microhemorrhage, it remains the investigation of choice for spinal lesions, especially for its ability to discern among several tumors, including meningiomas and schwannomas. When feasible, total surgical removal yields an excellent result and is the therapy of choice for symptomatic intradural extramedullary hemangiomas; nevertheless, a combination of subtotal or partial surgical excision and radiotherapy regimens has shown similar results.

**Declaration of patient consent**

Patient’s consent not required as patients identity is not disclosed or compromised.

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**Conflicts of interest**

There are no conflicts of interest.

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