A Rare Case of Primary Epidural Cavernous Hemangioma Mimicking Lobulated Upper Lobe Lung Mass

Sindhusha Veeraballi MD; Noreen Mirza MD; Brooke Kania DO; Leena Bondili MD; Vinod Kumar MD; Michael Maroules MD

1Saint Michael’s Medical Center, 111 Central Ave, Newark, NJ 07102, USA.
2Saint Joseph’s University Medical Center, 703 Main St, Paterson, NJ 07503, USA.

Abstract

Isolated primary epidural hemangioma is extremely rare and constitutes approximately four percent of epidural lesions, with the cavernous subtype being the most common. Hemangiomas are benign proliferative vascular lesions that are classified based on the predominant type of vascular channels involved into capillary, cavernous, arteriovenous, or venous. These vascular lesions can also be classified into malformations that are high flow (arteriovenous) and low flow (capillary, cavernous, venous). Herein, we present a rare case of epidural cavernous hemangioma presenting as lobulated right upper lobe lung mass extending to the spine demonstrated on Computed Tomography (CT) scan. Diagnosis of epidural cavernous hemangioma was made with the histopathological report after resection. In the case report, we discussed the clinical presentation, radiological findings, and morphology of different hemangiomas. We emphasize on the fact that there are several radiological mimics of rare epidural cavernous hemangioma, and this phenomenon can be easily missed pre-operatively. By our case report, we propose that the diagnostic suspicion of hemangiomas in epidural lesions is highly important to reduce the complications of bleeding during surgical resection.

Keywords: Cavernous hemangioma; Spinal cord compression.

Introduction

Hemangiomas are thought to be malformations of microcirculation rather than benign vascular neoplasms or hamartomas, and have multiple synonyms including cavernoma, cavernous hemangiomas, or cavernous malformations [1,2]. They are classified based on the predominant type of vascular channel (capillary, cavernous, arteriovenous, or venous). These hemangiomas may affect any part of the neuroaxis - most commonly the brain, cerebellum and brainstem, with the spine being the least commonly affected [3]. Most spinal epidural hemangiomas, which have been reported, are of the cavernous subtype [4]. It is extremely rare for cavernous hemangiomas to present as an epidural lesion with minimal cases reported [5]. Herein, we illustrate a rare case of a 61-year-old female with pure epidural cavernous hemangioma with signs and symptoms of radiculopathy and myelopathy radiologically presented as lung mass.

Case presentation

A 61-year-old Hispanic female with a past medical history of arthritis, osteoporosis with a history of multiple falls, presented with worsening back pain radiating to bilateral legs and difficulty walking for the past 1 year. She also complained of headaches with no associated changes in vision, dizziness, urinary or fecal incontinence. Her vitals were stable with a heart rate of 78 beats per minute, respiratory rate of 16 breaths per minute, blood pressure of 128/66 mmHg, and a temperature of 36 degrees Celsius. Physical examination was significant for midline spinal tenderness in the thoracic and lumbar regions. Motor strength of the right lower extremity was 3/5 and left lower extremity was 4/5. Sensation was intact throughout. The straight leg test on the right leg was positive at 45 degrees and the left leg was positive at 60 degrees. Bilateral knee examination was notable for limited range of motion. Examination of the neck revealed nuchal rigidity. However, Kernig’s and Brudzinski’s signs were negative.

On admission, lab results were significant for a hemoglobin level of 12.7 (normal: 13.5-17.5 g/dl), a white cell count of 9.5 (normal: 4,500-11,000 cells/microliter), platelet count of 243 (normal: 150 k - 450 k), erythrocyte sedimentation rate of 66 (normal: 1-13 mm/hr) (Table 1). CT spine was significant for a lobulated right upper lobe mass extending into thecal space at T3-T4 level (Figure 1), for which MRI was recommended for further evaluation. MRI cervical and thoracic spine without contrast revealed a right paraspinal soft tissue mass by the right apex; hyperintense on T2 weighted and isointense to muscle on T1 weighted images. The mass extended into the right T3 neural foramen and spinal canal, displacing and compressing the spinal cord. The mass measured 5.5 x 2.1 x 3.7 cm, occupying most of the small spinal canal at the level of T3 on the right. It extended below as well as above T3, especially posteriorly rated stents from T2-T4, and measured 4.1 cm in length and 8 mm in width on the sagittal T2 weighted projection. The anterior component also compressed the spinal cord and measured 1.5 cm in length (Figure 2). CT of the chest revealed a lobulated lung mass in the right upper lobe medially measuring 3.3 x 2.3 cm extending into the thecal space at T3-T4 level. CT chest also revealed dependent atelectatic changes of the lungs with mild scattered nonspecific ground-glass and interstitial opacities. A bone scan was performed, which was negative for metastatic skeletal disease.

The patient underwent emergency T2-4 bilateral laminectomies, T3 laminectomy and partial resection of epidural T3 tumor by neurosurgery. Histopathology report was suggestive for epidural cavernous hemangioma (Figure 3) with normal cartilage, bone, and skeletal muscle tissue. The patient had significant improvement in muscle strength after the resection and was initiated on steroid therapy.

![Figure 1](CT spine significant for lobulated right upper lobe mass extending into thecal space at T3-T4.)

![Figure 2](Magnetic resonance imaging of cervical and thoracic spine results (left picture STIR SAG view, right picture T1 AXIAL view) significant for mass extending into the right T3 neural foramen and spinal canal.)

![Figure 3](Pathology consistent hemangioma, with lobules of vascular channels with single-layer endothelial cell lining.)

Table 1: Initial laboratory parameters.

<table>
<thead>
<tr>
<th>Complete Blood Count</th>
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<tbody>
<tr>
<td>Hemoglobin</td>
<td>12.7 g/dl</td>
<td>(normal: 13.5-17.5)</td>
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<tr>
<td>Hematocrit</td>
<td>39.5 %</td>
<td>(normal: 41-50)</td>
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<tr>
<td>white cell count</td>
<td>10.5 cells/microliter</td>
<td>(normal: 4,500-11,000)</td>
</tr>
<tr>
<td>Platelets</td>
<td>249 K cells/microlitre</td>
<td>(normal: 150k-450k)</td>
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<tr>
<td>lymphocytes</td>
<td>17</td>
<td>(normal 24-44%)</td>
</tr>
<tr>
<td>Neutrophils</td>
<td>75</td>
<td>(normal 36-75%)</td>
</tr>
<tr>
<td>Monocytes</td>
<td>5</td>
<td>(normal 4-10%)</td>
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<thead>
<tr>
<th>Blood Chemistry</th>
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<tbody>
<tr>
<td>Blood urea</td>
<td>13</td>
<td>(normal 7-23 mg/dl)</td>
</tr>
<tr>
<td>Serum creatinine</td>
<td>0.61</td>
<td>(normal: 0.60-1.20 mg/dl)</td>
</tr>
<tr>
<td>Alkaline phosphatase</td>
<td>113</td>
<td>(normal: 9-46 U/L)</td>
</tr>
<tr>
<td>Aspartate transaminase (AST)</td>
<td>15</td>
<td>(normal: &lt;35U/L)</td>
</tr>
<tr>
<td>Alanine transaminase (ALT)</td>
<td>16</td>
<td>(normal: &lt;35U/L)</td>
</tr>
<tr>
<td>Erythrocyte sedimentation rate</td>
<td>66</td>
<td>(normal: 1-13 mm/hr)</td>
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</tbody>
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Discussion

In general, hemangioma represents an umbrella term used to describe benign vascular lesions that share similar histological features [6]. Malignant transformation of these lesions does not occur, and they are known to be common tumors of the head and neck. These benign proliferative vascular lesions are classified based on the predominant type of vascular channel involved into capillary, cavernous, arteriovenous, or venous. These vascular lesions can also be classified into malformations that are high flow (arteriovenous) and low flow (capillary, cavernous, venous) [8]. Arteriovenous malformations are known to have a direct association between arteries and veins without any intervening capillary network. They commonly affect the brain in young adults, manifesting as seizures or hemorrhages [8]. These lesions typically develop concurrently with the brain tissue; neural tissue develops between them, causing them to become atrophic and gliotic. The brain tissue surrounding the AVMs may undergo atrophy, creating a shunt, as it is a low resistance pathway that may steal blood from normal tissue [8].

Capillary hemangiomas of the spine are rare and are often times extramedullary occurring in the extra or intradural region. These vascular lesions are built up with small thin-walled vessels surrounded by pericytes and reticular fibers with the lining of flattened or plump endothelial cells [8]. These lesions tend to present within the cutaneous system and regress spontaneously [13].

Cavernous hemangiomas, as seen in our patient, are known to be “blood sponges” affecting the central nervous system. The mean age of presentation ranges from 30 to 60 years old, with the peak age of 40 years old. It is an uncommon low-flow lesion consisting of intercapillary vascular spaces and sinusoids with no neural tissue existing between the vascular spaces. These hemangiomas have deep and irregular dermal blood-filled channels [6]. Oftentimes, sinusoids and thin-walled cavernous vessels become tangled together to form these lesions [8]. They are separated by connective tissue stroma. These vascular lesions do not typically lead to mass effect or vasogenic edema unless they are associated with hemorrhage. Most of these lesions affect the intracranial area, specifically the supratentorial sites [7]. These lesions can affect the spine and when they do, they tend to affect the vertebral bodies. Intramedullary, intradural and extramedullary lesions of the spine are uncommon. Furthermore, purely epidural lesions are known to be extremely rare [7].

In our case, our patient had a paraspinal soft tissue mass known as an epidural cavernous hemangioma. Epidural cavernous hemangiomas account for less than 4% of all epidural spinal masses [7]. The most common location is in the posterior region of the epidural space. These lesions typically present clinically as an insidiously progressive compressive myelopathy over the course of a year [13]. An acute presentation would only be seen if there are signs of intramedullary hemorrhage. The most common symptoms are back pain that may mimic the presentation of a disk protrusion, slowly leading to signs of progressive paraparesis or paraplegia [7]. A precipitating factor for these symptoms is trauma, which can cause patients with these vascular lesions to present with radiculopathy, including sensory disruptions mimicking acute disc herniation [9]. Overall, the symptoms in our patient were nonspecific. Our patient presented with no radicular signs. This could be explained by the fact that the nerve roots are able to better tolerate the gradual development of soft compression versus the spinal cord [13]. However, she did present with symptoms of myelopathy including progressive worsening weakness as was seen in a case by Goyal et al., 2002 [3].

Differential diagnosis of these spinal epidural hemangiomas typically includes synovial cysts, certain granulomatous infections, lymphomas, meningiomas, neurogenic tumors, nerve sheath tumors, pure epidural hematomas, herniated disks, angiolipoma and epidural extramedullary hematopoiesis [11]. Rarely, as seen in our patient, they may present with extra foraminol extension of cavernous hemangioma presenting as lung mass [14]. In order to narrow down the differential diagnosis, it is important to accurately diagnose these lesions.

Diagnosis of these vascular lesions is achieved by MR imaging, which can provide vital information including location and extension [13]. Since these lesions are highly vascularized, it is imperative that they are identified correctly before surgical intervention. Based on a study by Lee et al., 2007, the most common MRI features that are seen in these spinal epidural hemangiomas are solid vascular masses that demonstrate a lobular contour [10, 11]. Low T2 intensity signal may be present due to the fibrotic capsule, the interface between the mass and the adjacent dura or posterior longitudinal ligament, an artifact produced due to chemical shift created by epidural fat and intratumoral fluid content, and lastly due to the deposition of hemosiderin [11].

Total surgical removal is the mainstay of treatment for these lesions and usually results in a good prognosis. In 2015, Li et al., completed a case series of 14 patients with epidural cavernous hemangiomas. All 14 patients underwent surgical removal with either complete resection, subtotal resection, or partial resection. Ten of these patients were then followed up at 34 months post-operation, and it was seen that 1 patient died, 5 patients showed clinical improvement and 4 patients did not show any neurological improvement [12]. Overall, many patients have a favorable prognosis with gradual resolution of neurological symptoms [13].

The high vascularity of the lesion makes it more prone to bleeding during surgical procedures. Hence, the patient is placed in a lateral position to control intraoperative bleeding and decrease the chest and abdominal compression induced increase in epidural venous pressure [13]. Complete resection in a microsurgical fashion is utilized to decrease the chance of intraoperative bleeding. Bipolar coagulation is used to progressively shrink the hemangioma by starting at the location across the thecal sac [13].

Conclusion

Our case highlights the unique presentation of epidural cavernous hemangioma mimicking a lung mass. Thus, due to the wide differential diagnoses and nonspecific clinical presentation, physicians ought to be vigilant in accurately diagnosing these vascular lesions to prevent the development of intraoperative hemorrhage.

References


