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A Twist in The Diagnosis of Acute Compressive Myelopathy

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ABSTRACT

Benign hemangiomas are often innocuous, intra-osseous and are incidentally found. Rarely, they become aggressive with extra-osseous expansion causing compressing on the spinal cord. However, early surgical intervention has proven benefits with neurological improvement and less reoccurrence. This is a case report of a 65-year-old gentleman who presented with acute on chronic progressive myelopathy after a recent infective episode. He underwent a decompression surgery, initially that was thought to be a spinal abscess but turned out to be a hemangioma. After surgery, the patient had good neurological and functional recovery. We present this case for its rare clinical presentation.

Keywords

Benign spinal tumors, Incidental, Hemangiomas, Vascular malformation.

Background

Benign hemangiomas are a part of a wide spectrum of hamartomas that have an overlapping clinical and imaging presentations but are clearly differentiated from vascular neoplasm. Though MRI scans are the gold standard for diagnosis of such spinal lesions, sometimes accurate preoperative diagnosis can be a great challenge. Hemangiomas account for less than 4% of all spinal tumors, being incidentally found in 27% of all autopsies and spinal imaging [1]. Only 1% of these become symptomatic of which 55% have pain alone and rest 45% have developed neurological compromise due to extra-vertebral extension [2], thus symptomatic neurological presentations are rare.

Case Report

A 65-year-old gentleman was referred to the spinal team after an MRI scan showed an abnormal lesion in the mid thoracic spine. He presented with a 7-week history of global numbness and tingling sensation of both legs leading to balance problems and frequent falls. He had no prior precipitating symptoms of illness, though had insect bites to his lower legs in the previous months. He had similar symptoms in 2012, which had almost fully resolved though his legs remained subjectively weak since then, necessitating the use of a stick to walk. He was otherwise in good health, with a resolved presumed episode of giant cell arteritis (temporal biopsy was negative) in 2009.

On examination he was clinically well, with neither sign of systemic compromise nor signs of infection. The patient demonstrated no tenderness along the entire spine with a good range of cervical and lumbar spine movements. He had altered sensations to light touch in all dermatomes distal to T10 bilaterally with sacral sparing. There were no alterations to his bowel and bladder control. He had some weakness (MRC scale 4/5) affecting his L2 and L3 myotomes but otherwise had full power. Reflexes were symmetrical and intact in his knees and ankles, though he demonstrated bilateral up-going plantar reflexes. He walked with a broad-based gait and was Romberg's sign positive.

Investigation

On presentation he was haemodynamically stable and apyrexial. His initial inflammatory markers showed a WCC of 7.32x109 and CRP of 46. His blood film was normal, with no signs of myeloproliferative disorders. An urgent MRI spine showed a 35mm highly dense focus at the level of T5-T6 spine with spinal cord compression.

Treatment

After discussion with the patient, consent was obtained for emergency decompression of the spinal cord at T5-T6 level. Intraoperatively, the appearance of the lesion was of a tumour

and not an abscess. Intraoperative tissue samples were sent to histopathology and microbiology. No organisms were grown, and histopathology confirmed the diagnosis of a hemangioma; sub-typing was inconclusive with a mixture of capillary sized and large vessels amidst adipose tissues.

Outcome

After surgery, the sensory and motor symptoms gradually improved over two weeks in hospital. Whilst awaiting histology, a screening CT scan of the whole body was performed looking for a potential primary source should the pathology show a metastatic deposit. This CT showed no evidence of a primary source of tumour but incidentally found a pulmonary embolus in his right lung. An opinion from the haematologist was sought in view of the asymptomatic pulmonary embolus, who suggested that it could have developed due to reduced mobility prior to admission for which anticoagulation regime was started.

Discussion

Hemangiomas are primarily a vascular tissue malformation found anywhere along the neuro-axis. They are often congenital, with a prevalence of 10-12% among the population [3,4]. These benign tumours often preclude early medical attention because they seldom have the malignant potential. Often, they are confined within the vertebral body (commonly thoracic spine) and grow slowly through repeated cell destruction and reorganisation [1] to a significant size at the fifth or sixth decade of life. Rarely, they expand extra-osseous into the epidural space commonly by a subtype called cavernous hemangioma. They constitute 4% of all spinal tumours [2] and can cause both progressive compressive myelopathy and sudden compressive features due to acute bleeds. In this case, the patient had a 35mm hemangioma compressing on T5-T6 level with an acute on chronic spastic myelopathy since 2012. His acute symptoms were consistent with anterior cord compression with loss of sensation and pain.

Factors such as compression fractures at the level of lesion, spinal stenosis and pregnancy are thought to be potential causes for precipitating the neurological symptoms in patients with previously asymptomatic vertebral hemangiomas [3,5,6]. Pure epidural hemangiomas are very rare [6].

Histological examination of a hemangioma has the appearance of a disorganised collection of blood vessels interspaced with fibrous adipose tissue and/or smooth muscle. Of the subtypes, cavernous and capillary hemangiomas are commonly found in the bones clearly differentiated from other types by the calibre of the blood vessels. Hwang PM and Rovira et al. [6,7] highlighted that capillary hemangiomas are associated with symptoms mostly because of their propensity to bleed easily whereas hemangiomas with more adipose tissue are generally clinically inactive.

Bhawan et al argued that immunological stains positive for CD31 and CD34 antibodies were suggestive of a hemangioma[8]. However, according to Jennings et al, CD34 antibodies had a broad expression in both vascular and non-vascular neoplasms which limits the diagnosing capability for hemangioma [9].

MRI is the gold standard for diagnosing a hemangioma. Characteristic appearance on other imaging modalities should raise suspicion and timely assessments needs to be done to avoid detrimental neurological effects. Simple imaging such as radiographs showing evidence of vertebral compression with erosion of spinal body appearing like vertical striae or honeycomb should raise a suspicion. CT scans may show a polka dot appearance when viewed in axial plane [10]. However, high resolution MRI clearly demarcates the tumour boundaries. Intraosseous hemangiomas have a fat signal intensity on T1 and T2 weighted images whereas extra osseous hemangiomas demonstrate iso-intensity on T1 and hyper-intensity on T2 weighted images [5]. This correlates with the fact that hemangiomas with less fat cells are more likely to invade posteriorly, thus having a higher propensity to bleed and cause neurological symptoms.

The treatment for acute spinal cord compression as in this case is either a total or partial excision of the hemangioma to relieve the symptoms. Since they are vascular structures, preoperative measures should be taken to avoid excessive bleeding in the limited time available. Performing an arteriography is useful to determine need for preoperative embolization of any feeding or draining vessels; the need for this is greater when managing capillary hemangiomas subtypes [6]. For cavernous subtypes, the treatment is excision because the extent of tumour cannot be clearly demarcated and the aim should be to dissect it carefully away from the dura [2]. Studies have showed that adjuvant radiotherapy is useful when complete resection is not possible and, in those patients,, who cannot undergo surgery [5]. A large multi-centre cohort study conducted in 2015 identified a recurrence rate of 3% in patients who underwent surgical excision, and no mortality was observed during a 4-year period follow up [11].

Summary

Hemangiomas account for a small percentage of spinal tumours, most of which are incidentally detected. Symptoms of chronic neurological compression or acute epidural bleeding are more frequently seen with extra osseous hemangiomas due to their capillary architecture, MRI should prove diagnostic and helps to plan the surgical excision and decompression that frequently results in a good outcome.

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