Clinics in Surgery

Case Report
Published: 18 Jul, 2019

Primary Intradural Extramedullary Solitary Fibrous Tumor/Hemangiopericytoma of the Thoracic Spine: Case Report

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Abstract

Solitary Fibrous Tumor/Hemangiopericytomas (SFT/HPC) is a rare tumor that arises from pericapillary cells. It is commonly found in musculoskeletal system and skin, but rarely found in the central nerve system, extremely rare in the spinal canal. Because of its rarity of this tumor, its clinical and radiographic characteristics of features and clinical manifestations have not been extensively. We report a rare case of Solitary Fibrous Tumor/Hemangiopericytoma (SFT/HPC) that developed in the thoracic vertebral intradural extramedullary region. A 50-year-old man experienced numbness in the left lower limb and weakness in the right lower limb from 2 months prior to initial visit to our hospital. MRI revealed that an intradural extramedullary tumor with strongly enhancement compressed the spinal cord toward the left on 7th thoracic vertebral level. Due to exhibit dural tail sign, clinically a diagnosis of meningioma was possible and removal of the tumor was performed. The histopathological diagnosis was SFT/HPC. The gait disturbance was improved postoperatively. SFT/HPC tumors arising in central nerve system mainly develop in intracranial region and SFT/HPC tumors occurring in the spinal canal, especially intradural region, are extremely rare. It is difficult to differentiate from meningioma clinically. Surgical excision is recommended as the first choice for treatment of this tumor, but SFT/HPC tumors are more likely to recur locally and to metastasize even over an extended time, so long-term observation is necessary.

Keywords: Solitary fibrous tumor; Hemangiopericytoma; Spinal tumor; Thoracic spine; MRI

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Received Date: *05 Jun 2019*
Accepted Date: *11 Jul 2019*
Published Date: *18 Jul 2019*

Citation:
Fujita T, Nakashima H, Sakai Y, Takatsu T. Primary Intradural Extramedullary Solitary Fibrous Tumor/Hemangiopericytoma of the Thoracic Spine: Case Report. Clin Surg. 2019; 4: 2515.

Introduction

Hemangiopericytoma (HPC) and Solitary Fibrous Tumors (SFTs) are vascular-rich mesenchymal tumors. They are known to develop primarily in the skin and musculoskeletal system; hence, intracranial development is rare, and development in the spinal cord is even rarer [1-7]. In the newly revised 2016 World Health Organization (WHO) Classification of Tumors of the Central Nervous System, SFTs and HPC were placed in the same tumor category (SFT/HPC) on the grounds that they share identical genetic mutations. Conventional low grade (WHO I) tumors with low cellular density and pattern less architecture are classified as SFTs. In contrast, WHO grade II tumors are categorized as HPC, and highly malignant (WHO III) tumors with high cellular density and numerous nuclear division images are referred to as anaplastic HPC [8]. This study reports a case of SFT/HPC that developed in the thoracic vertebral intradural extramedullary region.

Case Presentation

A 50-year-old man with no noteworthy previous medical history experienced pain in the right anterolateral thoracic region approximately 2 months prior to initial examination. As the left lower limb gradually became numb and the right lower limb felt weak, the patient presented to our clinic. The physical findings at the initial examination revealed that his manual muscle testing score was generally low, at approximately 2 to 4 in the right lower limb. Additionally, the patient exhibited hypoesthesia in the left lower limb and enhanced deep tendon reflex in both lower limbs; however, no bladder and rectal dysfunction was noted. Plain sagittal Computed Tomography (CT) revealed a pale high-density area in the spinal canal at the 7th thoracic vertebra. No calcification was observed (Figure 1). On Magnetic Resonance Imaging (MRI), both T1- and T2-weighted images showed a mass lesion of almost equal signal intensity to the spinal cord that was compressing the spinal cord toward the left within the dura mater (Figure 2). Contrast-enhanced MRI revealed that the tumor was homogeneously strongly enhanced, exhibiting a dural tail sign (Figure 3).

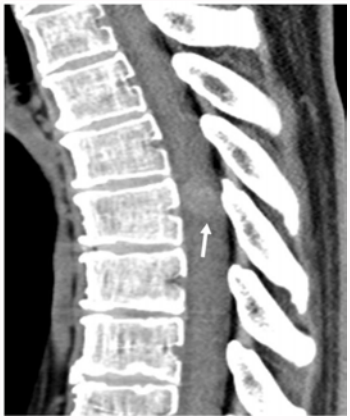

Figure 1: Initial plain sagittal computed tomography scans reveals a pale high-density area in the spinal canal at the 7th thoracic vertebra. There is no calcification in the area.

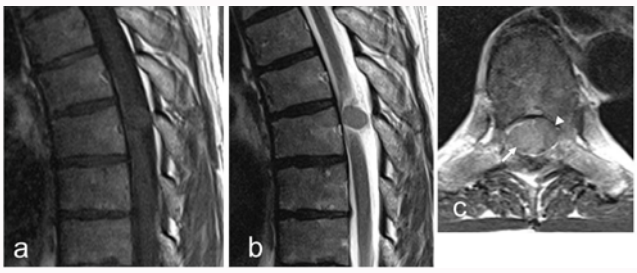

Figure 2: Preoperative MRI images are shown a lesion of Th7. The sagittal imaging reveals an intradural mass with isointense on sagittal T1-weighted imaging and T2-weighted imaging (a,b). The intradural mass (arrow) is compressing the spinal cord (arrow head) toward the left on axial T2-weighted imaging.

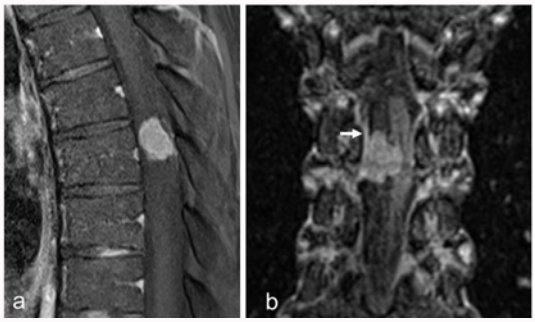

Figure 3: The tumor Marked and homogenous enhancement is observed on contrast-enhanced sagittal T1-weighted images. On coronal view dural tail sign is indicated (arrow).

Clinically, a diagnosis of meningioma was possible. The paralysis progressed, resulting in walking difficulty. Therefore, laminectomy and tumorectomy under general anesthesia were scheduled. Surgery was performed under general anesthesia. Using a posterior median approach, the caudal part of the 6th vertebral arch and the 7th thoracic arch were resected, the dura was dissected under a microscope, and the tumor was confirmed. The tumor was not adhered to the dura mater, and there was no infiltration into the spinal cord. The tumor was hemorrhagic, and the entire tumor was resected piecemeal. Intraoperative monitoring of motor evoked potential showed no decrease in amplitude. The histopathological findings of the excised specimens revealed stag horn-shaped capillaries surrounded by short spindle shaped cell proliferation (Figure 4). Immunohistochemical

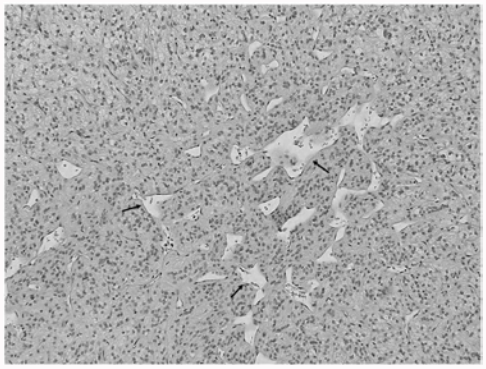

Figure 4: Histological examination of the intradural extramedullary tumor with hematoxylin and eosin staining (original magnification 200x) shows staghorn-shaped capillaries (arrow) surrounded by proliferation of short spindle shaped cells.

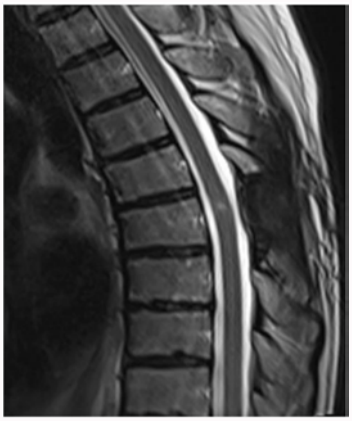

Figure 5: Sagittal T2-weighted image at 12 months after surgery showing high intensity area in the spinal cord but no evidence of recurrent disease.

staining was positive for cluster of differentiation 34 (CD34), slightly positive for vimentin, and negative for epithelial membrane antigen, chromogranin, glial fibrillary acidic protein, synaptophysin, S-100, and factor-VIII. Based on these results, the patient was diagnosed with SFT/HPC (WHO classification grade II). The MIB-1 Labeling Index (LI) was less than 5%. On the day following surgery, the patient was able to walk using a walker. Eight days after surgery, he could walk independently and ascend/descend stairs. As of 12 months after surgery, although MRI shows high signal areas in T2-weighted images, the patient has experienced no recurrence or hindrance to daily life (Figure 5).

Discussion

SFT/HPC is a rare tumor that accounts for less than 1% of all intracranial tumors [1,2]. In particular, the incidence of SFT/HPC in the spinal cord is extremely rare [1-7]. A majority of cases of SFT/HPC in the spinal cord occur in the cervical and thoracic spines [1,2]. In addition, there are more epidural cases and intradural cases are rare [1-7,9,10]. To the best of our knowledge, only 20 cases of thoracic vertebral intradural extramedullary SFT/HPC have been reported, including the present case [1-7,9-11,12]. Overall, men appear to be more predisposed than women, with 13 men and 7 women being documented. The average age of these patients was 39 years (12 to 65 years). In intra-cranial SFT/HPC, tumors are typically adhered to the dura mater however, of the four spinal SFT/HPC

cases in which the tumor adhesion site was reported three had no adhesion to the dura mater, and the remaining case was unspecific [2,4,6,7,13]. In our case, as the intraoperative findings did not reveal any adhesion to the dura mater, and the tumor was underneath the pia mater, the tumor was presumed to have originated from the pia mater. Kashiwazaki et al. [7] and Moscovici et al. [6] also concluded that tumors originate from micro capillaries within the pia mater. The image findings of spinal SFT/HPC are nonspecific; therefore, differentiating spinal SFT/HPC from other spinal tumors, such as meningioma and neurinoma, is difficult [2,3,11]. On plain CT, tumors appear as high-density areas adhering to the dura mater. On MRI images, tumors are either oval-shaped or ovulated, and both T1- and T2-weighted images show similar intensity signals; with gadolinium contrast, lesions are homogeneously and intensely enhanced. Another characteristic of SFT/HPC is the presence of a flow-void sign due to blood flow [2,11]. Chiechi et al., [13] stated that intracranial SFT/HPC can be differentiated from meningioma on the grounds that on CT, thickening and calcification of adjacent bones are observed in meningioma, but calcification is not observed in SFT/HPC. Instead, erosion of adjacent bones is observed in SFT/HPC. Although contrast-enhanced MRI detects dural tail signs in both tumor types, meningioma is typically widely adhered to the dura mater. Therefore, HPC should be considered when adhesion to the dura mater is narrow. Yi et al. [3] reported that a dural tail sign is uncommon in spinal HPC. In our case, the tumor developed in the thoracic vertebral region, compressing the spinal cord toward the left, and the frontal section of contrast MRI showed a dural tail sign. Therefore, a diagnosis of meningioma was considered most probable, which frequently occur in the higher thoracic vertebral region, particularly in posterior and postero-lateral areas making it difficult to list SFT/HPC for differential diagnosis. Although surgical excision is recommended as the first choice for treatment method of SFT/HPC the total excision rate is 50% to 70%, with high risks of recurrence and distal metastasis [1,2,4,5,7,10,11,14]. This is because the tumor is often adhered to the dura mater [15]. The effectiveness of adjuvant radiotherapy for spinal SFT/HPC remains unclear; hence, most research suggests that adjuvant radiotherapy is not recommended. Nevertheless, some reports suggest that radiotherapy should be considered for subtotal excision, highly malignant tumors, and recurring cases [10,11]. Chemotherapy is rarely performed and is considered ineffective [1,2]. In the present case, considering that complete excision was achieved by piecemeal resection and that MIB-1 LI was less than 5%, neither adjuvant radiotherapy nor chemotherapy was administered. However, the prognostic significance of molecular biological markers such as MIB-1 (Ki-67) LI remains to be clarified [16]. There is no established method available to effectively predict the prognosis of spinal SFT/HPC, and the 5 and 15 year recurrence rates are high (60% and 87%, respectively) [2]. The likelihood of metastasis to the central nervous system and/or distant metastasis is 4% to 50% over an extended period [6,11]. In this case, no apparent recurrence has been observed on images as of 12 months after surgery; however, long-term observation is necessary.

Conclusion

We reported an extremely rare case of intradural extramedullary SFT/HPC in the thoracic spine, which caused compression of the spinal cord. It was difficult to diagnose preoperatively and to differentiate from meningioma. The patient recovered soon from incomplete paralysis after surgery.

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