Case Report

OPEN

Intravascular Papillary Endothelial Hyperplasia (Masson's Tumor) Within Cauda Equina

Masahiro Tanaka, MD, PhD
Akihiko Hiyama, MD, PhD
Daisuke Sakai, MD, PhD
Hiroyuki Katoh, MD, PhD
Masato Sato, MD, PhD
Masahiko Watanabe, MD, PhD

From the Department of Orthopaedic Surgery, Tokai University School of Medicine, Kanagawa, Japan.

Corresponding to Dr. Tanaka: m.tanaka@is.icc.u-tokai.ac.jp

Dr. Sato or an immediate family member has received research or institutional support from CellSeed. None of the following authors or any immediate family member has received anything of value from or has stock or stock options held in a commercial company or institution related directly or indirectly to the subject of this article: Dr. Tanaka, Dr. Hiyama, Dr. Sakai, Dr. Katoh, and Dr. Watanabe.

JAAOS Glob Res Rev 2018;2:e087

DOI: 10.5435/JAAOSGlobal-D-17-00087

Abstract

Objective: To describe a rare case of intravascular papillary endothelial hyperplasia (IPEH), also called as Masson's tumor, in a patient with the cauda equina syndrome presenting with low back pain and leg pain.

Summary of Background Data: There are no studies that have reported a case of IPEH within the cauda equina.

Methods: A 40-year-old man with no prior medical history presented with low back pain and leg pain beginning approximately 5 years ago and a month ago, respectively. MRI revealed a mass at the L2-L3 level that was isointense on T1-weighted images and hypointense with partial areas of high signal intensity on T2-weighted images. A myelogram demonstrated an intradural lesion without any respiratory fluctuation of tumor position, thereby suggesting a metastatic lesion; however, no abnormalities were found on a full-body CT scan or serum investigations for tumor markers. Therefore, surgery was performed on the lesion, within the cauda equina, for diagnosis as well as treatment.

Results: Histopathological examination of the excised tumor indicated IPEH. The symptoms and quality of life improved postoperatively; there was no recurrence of the lesion.

Conclusion: This is the first report of an IPEH intradural lesion within the cauda equina.

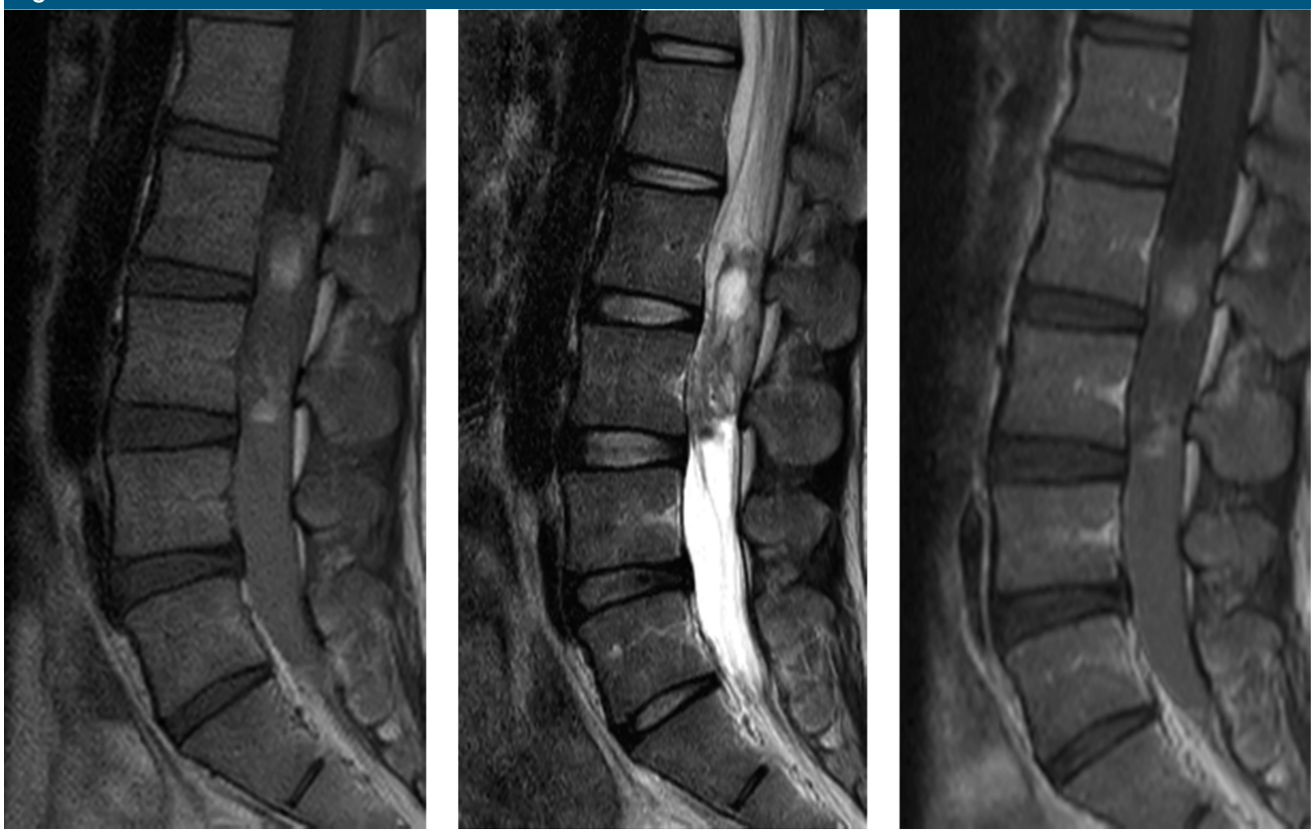
Study Design: Case report

Intravascular papillary endothelial hyperplasia (IPEH) was reported in 1923 by Masson et al. as a benign and non-neoplastic lesion.[1,2] IPEH occurs in the subcutaneous and soft tissues of the body and is commonly seen in the limbs as well as the head and neck region, which are relatively prone to injury.[3] The occurrence of IPEH in the spinal canal is rare; there are only four reported cases of extradural IPEH in the spinal canal[4-7] and none reported in the intradural space. Here, we report a case of intradural IPEH within the cauda equina and present a review of the literature.

Case Report

A 40-year-old man with no prior medical history presented with low back pain and leg pain beginning approximately 5 years ago and a

Figure 1



A **B** **C**

Images of the lesion at the L2-L3 level. T1-weighted image (WI; sagittal view) (**A**), T2-WI (sagittal view) (**B**), and gadolinium-enhanced, fat saturated T1-WI (sagittal view) (**C**).

month ago, respectively. MRI revealed a tumor in the cauda equina at the L2-L3 level, and the patient was referred to our hospital for further tests and surgery.

The patient reported of bilateral pain from the buttocks to the lower legs resulting in an intermittent claudication of approximately 200 m. Physical examination revealed normal muscle strength, reflexes, and bladder function, along with a positive straight leg raise test (70° bilaterally), and a negative femoral nerve stretch test bilaterally. The Japanese Orthopedic Association score was 13 out of 29.[8]

Plain radiographs were normal. MRI revealed a mass at the L2-L3 level that was isointense on T1-weighted images and hypointense with partial areas of high signal intensity on T2-weighted images. Repeat contrast-enhanced MRI did not result in signal amplification (Figure 1). "Signal characteristics" is intended here, but its signal characteristics on MRI imaging are nonspecific and could represent ependyoma, meningioma, or cavernous hemangioma. A myelogram demonstrated an intradural lesion without any respiratory fluctuation of tumor position (Figure 2), and a subsequent CT did not reveal calcification within the tumor. Although the MRI characteristics were not typical of a metastatic tumor, a full-body CT scan and blood tests for serum tumor markers were performed, ruling out the possibility of a malignant metastatic tumor.

Surgery was performed to diagnose the lesion and to relieve the pain. The patient underwent a L2-L3 laminectomy. A dura mater was exposed, but there was no epidural tumor. A durotomy revealed a highly vascularized, soft, elastic tumor measuring 2.5 × 1.5 × 1 cm. Adhesion was minimal, and the tumor was dissected from the surrounding cauda equine and was excised en bloc (Figure 3).

Histopathological examination revealed thrombus and fibrin clots mixed with tissue fragments; numerous dilated papillary structures of various sizes were found with prominently hyalinized stroma, but findings indicating malignancy such as anaplasia, mitosis, and necrosis were not observed (Figure 4). Immunohistologically, the endothelial cells exhibiting papillary proliferation were positive for CD31 and CD34, thus

identifying these cells as vascular endothelial cells. From these findings, the diagnosis of IPEH was reached. Resection of the tumor resolved the patient's symptoms and the Japanese Orthopedic Association score improved from 13 to 29. No recurrence has been observed in the 2-year follow-up period.

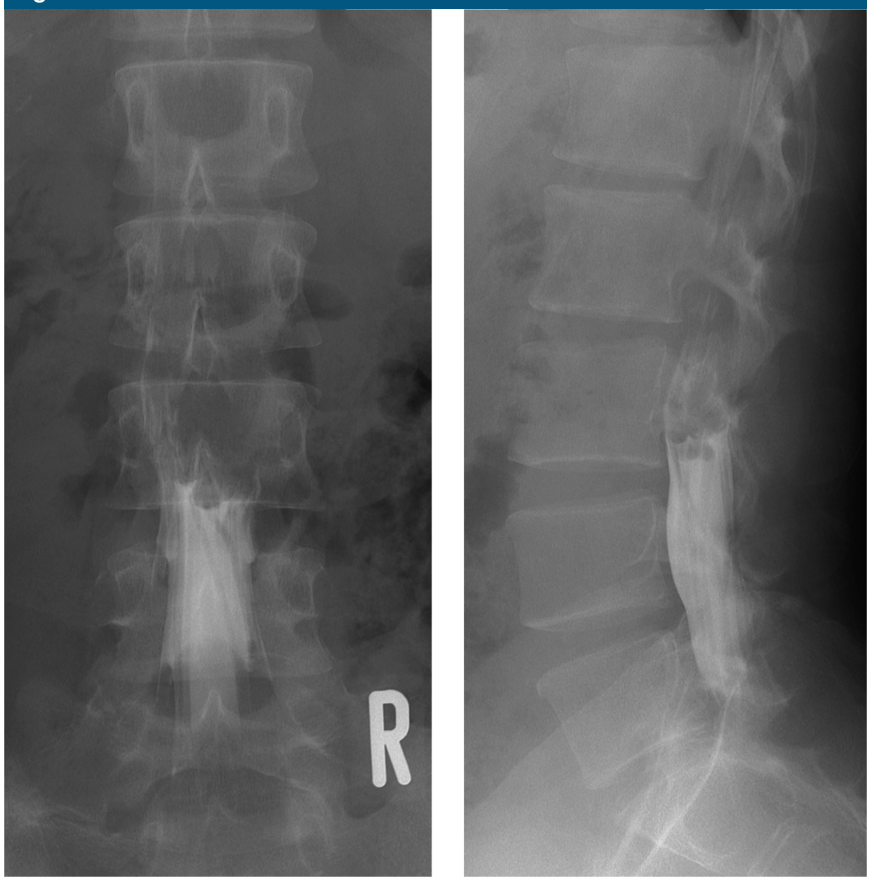
Discussion

IPEH was first reported by Masson[1] in 1923 when he identified proliferation of intravascular endothelial cells in a hemorrhoidal vein, calling it "hemangioendotheliome vegetant intravascular." Subsequently, Clearkin and Enzinger[2] recognized that it was not a true tumor and hence named it "intravascular papillary endothelial hyperplasia" in 1976, and the name has been in use ever since. Masson surmised that IPEH occurs due to thrombosis secondary to papillary proliferation of vascular endothelial cells,[1] whereas Salyer and Salyer[9] postulated that it is a type of reactive lesion in which the vascular endothelial cells proliferate in reaction to thrombus organization.

IPEH is not a true tumor. It occurs in the subcutaneous and soft tissues throughout the body, but is more common in the limbs as well as the head and neck region, which are relatively prone to injury. IPEH in the head and neck region accounts for 10% to 13% of all cases,[10] and there are very few reports on its occurrence in the nervous system, particularly the spinal canal, with only four known cases.[4-7]

IPEH has no age predisposition, with slight female preponderance. It often presents as a slow-growing, tender, violaceous cutaneous mass on the head, neck, fingers, and trunk. Deeply situated lesions occur in intramuscular hemangiomas. Although IPEH is benign lesion, it forms expansile and compressive masses.[5]

Figure 2



A B

Plain myelogram of the lumbar spine showing hypodensity at the L2-L3 level. AP view (**A**), lateromedial view (**B**).

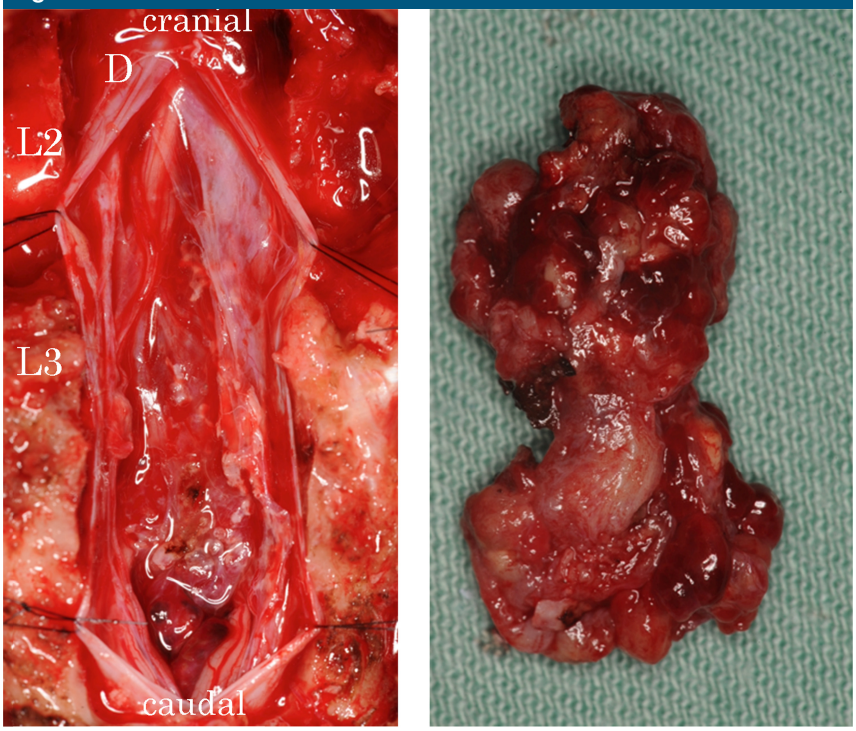
IPEH is generally hypervascular and is enhanced by gonadalium. T2-weighted MR imaging generally shows IPEH as hyperintense, whereas T1-weighted imaging typically demonstrates the lesion as hypointense to isointense.[11]

The histopathological characteristics of IPEH are: (1) papillary proliferation of vascular endothelial cells is localized intravascularly, (2) normal thrombus formation is observed and almost the entire papillary tissue is associated with the thrombus, (3) the vascular endothelial cells are in one or two layers and are not multilayered, (4) almost no abnormalities or mitotic changes are observed in the vascular endothelial cells, and (5) there is no accompanying tissue necrosis.[3] This case was diagnosed as IPEH by satisfying four of the above criteria, with the exception that the papillary proliferation was extravascular.

Hashimoto et al[3] divided IPEH into three categories based on the differences in occurrence: a primary form that occurs without existing vascular lesions, a secondary form that arises within existing vascular lesions such as varicose veins, hemangioma, or arteriovenous malformation, and an extravascular form that arises extravascularly and is derived from a hematoma. This case was an extravascular form because no preexisting vascular lesions were identified.

Although the pathogenesis of IPEH is still unclear, it is postulated that the

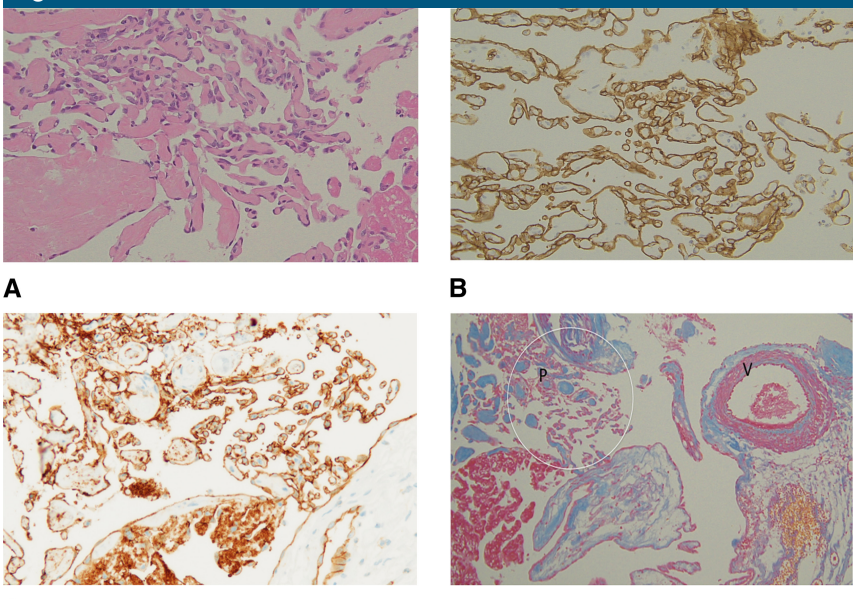
Figure 3



A B

Intraoperative and postoperative findings. The intradural mass during surgery (**A**). The 2.5 × 1.5 × 1.0 cm mass (**B**) was soft and elastic. L = lamina, D = dura mater

Figure 4



A B C D

Photomicrograph showing papillary structures lined by the endothelium in the extravascular space; Hematoxylin-eosin staining at ×20 (**A**), CD34 immunohistochemical staining at ×20 (**B**), CD31 immunohistochemical staining at ×20 (**C**), and Azan staining at ×4 (**D**). P = papillary structures, V = vessel wall

trigger may be the formation of a thrombus due to trauma, followed by reactive proliferation of the vascular endothelial cells. IPEH does not exhibit malignant characteristics such as association with malignant neoplasms, metastasis, or invasion. Complete resection is considered the best approach to avoid future local recurrence as long as its location is accessible.[12] There have only been four reported cases of IPEH within the spinal canal: Porter et al[4] reported an extradural tumor at the level of T6 compressing the spinal cord and Taricco et al[5] reported an extradural lesion at the level of T12-L1 compressing the cauda equina. Ali et al[6] reported a mass in the posterior epidural space at the T8 level. In three cases, neurological symptoms resolved completely after resection of the lesions and the diagnosis of IPEH was carried out by histopathological examination of the resected tissue. Petry et al[7] reported of IPEH involving both the retroperitoneum and the cervical, thoracic, and lumbar spine. In particular at the T10, T11, and T12 levels, spread to the epidural space was associated with mild ventral thecal sac compression demonstrated by MRI. No abnormal cord signal was identified. This is the first report of an IPEH intradural lesion. Although the etiology is not clear, we postulate that an intradural thrombus formed for reasons unknown such as trauma or signs of meningismus or other source of intradural hematoma from which patients are usually symptomatic and resulted in the reactive proliferation of the vascular endothelial cells during thrombus formation. In particular, this lesion extended from the meninges and it may be possible that the tumor in this case was attached to the meninges. Resection of the tumor led to complete resolution of the symptoms and we have not observed any recurrence. Hence, complete resection is recommended when an intradural tumor is suspected to be a vascular lesion.

Conclusion

Here, we reported a case of an intradural IPEH within the cauda equina that was treated with complete resection.

Acknowledgement

The authors would like to thank Chie Inomoto (Department of Pathology, Tokai University School of Medicine) for supporting our studies.

References

1. Masson P: Hemangio-endotheliome vegetant intravasculaire. *Bull Mem Soc Anat (Paris)* 1923;93:517-523.
2. Clearkin KP, Enzinger FM: Intravascular papillary endothelial hyperplasia. *Arch Pathol Lab Med* 1976;100:441-444.
3. Hashimoto H, Daimaru Y, Enjoji M, et al: Intravascular papillary endothelial hyperplasia: A clinicopathologic study of 91 cases. *Am J Dermatopathol* 1983;5:539-546.
4. Porter DG, Martin AJ, Mallucci CL, et al: Spinal cord compression due to Masson's vegetant intravascular hemangioendothelioma. *J Neurosurg* 1995;82:125-127.
5. Taricco MA, Vieira JO Jr, Machado AG, et al: Intravascular papillary endothelial hyperplasia causing cauda equina compression: Case report. *Neurosurgery* 1999;45:1478-1480.
6. Ali SZ, Farmer PM, Black K, et al: Masson's hemangioma of spinal meninges causing cord compression with paraplegia. *Ann Clin Lab Sci* 1994;24:371-375.
7. Petry M, Brown MA, Hasselink JR, et al: Multifocal intravascular papillary endothelial hyperplasia in the retroperitoneum and spine: A case report and review of the literature. *J Magn Reson Imaging* 2009;29:957-961.
8. Izumida S, Inoue S: Assessment of treatment for low back pain: Japanese Orthopaedic Association [Japanese]. *J Jpn Orthop Assoc* 1986:60:391-394.
9. Salyer WR, Salyer DC: Intravascular angiomatosis: Development and distinction from angiosarcoma. *Cancer* 1975;36: 995-1001.
10. Luce EB, Montgomery MT, Redding SW, et al: Intravascular angiomatosis (Masson's lesion). *J Oral Maxillofac Surg* 1988;46: 736-741.
11. Sedat C, Nezih O, Tayfun D, et al: Intravascular papillary endothelial hyperplasia of the central nervous system—Four case reports. *Neutrol Med Chir (Tokyo)* 2004;44:302-310.
12. Avellino AM, Grant GA, Harris AB, et al: Recurrent intracranial Masson's vegetant intravascular hemangioendothelioma: Case report and review of the literature. *J Neurosurg* 1999;91:308-312.