

*Aktuelne teme /
Current topics*

Correspondence to:

Nemanja Rančić,
Vojnomedicinska akademija Beograd
nece84@hotmail.com
Tel: 063-8524443

MASSON TUMOR IN THE SPINAL CANAL -
RADIOLOGICAL DIAGNOSTIC CRITERIA
FROM SUSPICION TO TIMELY DIAGNOSIS
MASSONOV TUMOR U KIČMENOM
KANALU - RADIOLOŠKI DIJAGNOSTIČKI
KRITERIJUMI OD SUMNJE DO
PRAVOVREMENE DIJAGNOZE

Bojan Labus^{1*}, Milica Pantić^{1,2*}, Andrija Kostić^{2,4},
Jelena Golubović⁵, Miroslav Mišović^{6,7},
Nemanja Rančić^{6,7,8}

*Bojan Labus and Milica Pantić shared first authors.

¹ Clinic for Orthopedic Surgery and Traumatology, Military Medical Academy, Belgrade, Serbia

² Faculty of Medical Sciences University of Kragujevac, Serbia

³ Clinic of Psychiatry, University Clinical Centre Kragujevac, Serbia

⁴ Clinic of Neurology, KBC „Dr Dragiša Mišović-Dedinje“, Belgrade, Serbia

⁵ Department of Radiology, KBC Bežanijska Kosa, Belgrade, Serbia

⁶ Institute of Radiology, Military Medical Academy, Belgrade, Serbia

⁷ Faculty of Medicine of the Military Medical Academy, University of Defense, Belgrade, Serbia

⁸ Centre for Clinical Pharmacology, Military Medical Academy, Belgrade, Serbia

Key words

Masson tumor, radiological diagnosis, criteria, neurology

Ključne reči

Massonov tumor, radiološka dijagnoza, kriterijumi, neurologija

Abstract

Introduction: Intravascular papillary endothelial hyperplasia (IPEH) represents a rare reactive proliferation of endothelial cells around an organized thrombus, localized in a dilated blood vessel, hematoma, or vascular lesion. The symptoms that arise as a result of this lesion depend on the localization of the pathological process itself. Localization of this lesion in the spinal canal is extremely rare, so until now there are only a few published clinical cases. The aim of this work is to present the radiological diagnostic criteria of this localization. **Material and method:** A systematic search of the PubMed, Medline, Radiopedia databases was performed. **Results:** Magnetic resonance imaging shows an iso to slightly hypointense signal on the T1, a hyperintense signal on the T2, and inhomogeneous postcontrast amplification on the T1 C+ sequence. **Conclusion:** Neurological symptoms are often the result of other, primarily somatic, diseases and pathological processes of different localisations. On magnetic resonance the main characteristics of IPEH are isointense to hypointense appearance on T1 and heterogeneous hyperintense appearance on T2 weighted magnetic resonance images, and also shows amplification after intravenous administration of gadolinium. It is very important to keep in mind that intravascular papillary endothelial hyperplasia often resembles a secondary deposit which can confuse radiologists. The importance of this article is in the domain of timely and adequate diagnosis and differential diagnosis, because the initially presented symptoms can resemble numerous neurological or systemic diseases.

INTRODUCTION

Intravascular papillary endothelial hyperplasia (Masson's vegetative hemangioendothelioma) is a sporadic neuropathological disease with pleiotropic clinical presentation (1). Pierre Mason described it for the first time in 1923, but later, his description was criticized by Henschen. Although there are still controversies regarding the histological structure of this tumor, most authors believe it is a type of organized thrombus surrounded by the proliferation of endothelial cells (2). In 1975, Klerkin and Enzigen named this lesion with intravascular papillary endothelial hyperplasia (IPEH). Today, in the literature, these two names are still equated in professional and scientific use (3). The symptoms that appear as a result of this tumor depend on its localization. Considering this, the symptoms can often mislead clinicians, which speaks in favor of the importance of a multidisciplinary approach in diagnosis, although radiological diagnosis is crucial. Localization of the IPEH in the spinal canal is highly unusual, so in the literature, there are only a few clinical cases of this localization (4-6). In the following work, we presented the radiological diagnostic criteria for the thoracic localization of IPEH.

Radiological diagnostic criteria through literature review

IPEH is a slow-growth vascular neoplasm with histologically benign characteristics. For the same entity, the following keywords were used such as: „intravascular papillary endothelial hyperplasia” OR „IPEH” OR „Masson hemangioma” OR „Masson pseudoangiosarcoma” OR „Masson tumor” (7). IPEH is a rare disease that many radiologists and other specialists might be unfamiliar with. It can mimic malignancy from many points of view, such as clinical, endoscopic, and especially radiological (8). The radiological assessment often suggests malignancy, although the detailed description of its radiological characteristics is not accessible (7, 8). Despite this, preoperative radiological evaluation is helpful in surgical planning, suggesting a non-invasive approach more compatible with a benign tumor, preventing unnecessary aggressive surgical approaches (7). Magnetic resonance imaging (MRI) adds a preoperative value in cases of unilateral non-transparency seen on multi-detector computed tomography (MDCT) (8, 9).

IPEH of the spinal canal is an infrequent entity described in only a few cases in the available literature; it is often first seen on an MDCT and additionally diagnosed on an MRI (9). IPEH manifests as solitary or multiple rounds, clearly circumscribed, focal amplifying lesions throughout the spinal canal, primarily involving the posterior vertebral bodies (10). MDCT examination revealed a well-defined and well-circumscribed solid ovoid mass with soft tissue density and calcification without significant contrast amplification. Bone erosion and fatty infiltration have been described in some cases (7). MR imaging lesion shows iso to hypointense appearance on T1 and hyperintense appearance on T2 weighted, and amplification after intravenous administration of gadolinium (9). It may extend from the posterior aspect of the vertebral body to the anterior epidural space, causing compression of the ventral thecal sac. Spinal involvement is rare, with few cases described in the literature (10).

Functional MR imaging, such as diffusion-weighted (DW) imaging, has a role in differentiating the histological characteristics of tumors, as tumor apparent diffusion coefficient (ADC) values tend to vary depending on the stage of tumor cell differentiation, the degree of cellularity, the presence of necrotic tissue, and degenerative changes in the interstitium. Malignant soft tissue tumors usually have low ADC values, which are described by low ADC signal intensity, while benign tumors show higher ADC values. Regardless of the stated benefits, DW imaging has never been described in IPEH lesions of the spinal canal (7).

Most bone tumors share some MRI features, so there are no specific radiological features of IPEH, as similar radiological findings can be seen in other lesions such as hemangioma, eosinophilic granuloma, aneurysmal bone cyst, medullary fibrosarcoma, hemangioma and metastatic tumors of the thyroid gland, kidney, rectum, breast, prostate and lungs. The most common differential diagnosis is metastatic processes. The description of metastatic processes in MDCT will depend on the degree of mineralization. Lytic metastases appear as regions of soft tissue attenuation with irregular margins. If the mass penetrates the cortex, it can cause spinal canal compression. Sclerotic, hyperdense and irregular lesions are less likely to extend beyond the vertebrae (11). Metastases of the neoplasm more often involve the pedicles and the adjacent vertebral body (10).

IPEH is divided into three subtypes: the pure form, where it occurs in dilated vessels; the mixed form in already existing vascular lesions; and the extravascular form, in which it is inside the hematoma (10). Histologically, it is essential to distinguish IPEH from angiosarcoma. Pathohistological analysis indicated an organized hematoma associated with intraluminal endothelial proliferation. A precise diagnosis will enable adequate treatment consisting of complete resection. Since IPEH is often considered a malignant disease, a definitive diagnosis requires careful pathohistological examination using immunohistochemistry, which is the key method for determining IPEH (8, 9).

DISCUSSION

IPEH is an infrequent pathological change that manifests as a reactive vascular lesion with expansion and formation of a compressive mass. Pierre Mason first described it in 1923. It was then called Mason's tumor, and later, the name suffered numerous criticisms. However, considering the localization and the symptoms it causes, it still has the characteristics of a tumor, so the name itself was changed by introducing a term corresponding to this pathological process's pathohistological structure. Although the predominant age at which IPEH occurs has not been demonstrated, data in the literature suggest gender differences. Intracranial localization is significantly more common in females, with a ratio of 4:1. Localization in the spinal canal, although extremely rare, occurs more often in males (3). Usually, IPEH is found in numerous tissues such as the skin and subcutaneous tissue of the extremities, neck or head, then in the oral mucosa, lips, sinus cavities, parotid gland, thyroid gland, lungs, vena cava superior, glands, veins and intracranially. Only a few cases with localization in the spinal canal have been published so far (12-16).

The pathogenesis of IPEH is still insufficiently elucidated. Different authors explain this pathological lesion, from an overreaction to the thromb reorganization, to the proliferation of endothelial cells with secondary thrombosis and fibrin deposition (3). It is significant to note that the diagnosis of IPEH is very demanding and challenging due to non-specific MRI features. IPEH of the spinal column is an infrequent entity described in only a few cases in the available literature. It is described as solitary or multiple rounds, clearly circumscribed, focal enhancing lesions throughout the backbone infiltrating the posterior vertebral bodies (10). On MR imaging, the main features of IPEH are a hypointense appearance on T1, a hyperintense appearance on T2, and amplification after intravenous administering gadolinium (9). MDCT examination revealed a well-defined and well-circumscribed solid ovoid mass with soft tissue density and calcification without significant postcontrast enhancement. Bone erosion and fatty infiltration have been described in some cases (7), although we did not find any bone erosion in the attached images. It is essential to remember that IPEH often resembles a secondary deposit, which can confuse clinicians, and in the case of an inadequate assessment, it can mask the primary diagnosis. Therefore, it is always necessary to differentially consider other pathological processes such as cavernous/capillary hemangioma, Kaposi's sarcoma, endovascular papilloma and angiothelioma, schwannoma, neurofibromas and arteriovenous malformations (16).

The symptoms that occur due to IPEH depend on the localization of the pathological process. Localization in the spinal canal leads to pain in the chest and back, numbness of the lower extremities, paresis or paralysis, and bladder dysfunction due to spinal cord compression or cauda equine (17). Treatment is considered in a situation where continuous, severe pain appears, as well as compressive symptomatology. If diagnosed in time, which is imperative, complete surgical resection is possible. This is the most clinically pre-

ferred treatment modality and has been shown to have the most favorable outcome (18).

CONCLUSION

Neurological symptoms are often the result of other, primarily somatic, diseases and pathological processes of different localisations. Although it is rarely localized in the spinal canal, IPEH is a curable condition if it is diagnosed and treated promptly. MRI is a non-invasive and relatively accessible diagnostic method with reliable sensitivity and specificity. This is an irreplaceable diagnostic method for preoperative orientation and for making an operative plan. On MR imaging, the main features of IPEH are iso to hypointense appearance on T1 and hyperintense appearance on T2, and it also shows amplification after intravenous administration of gadolinium. It is important to remember that IPEH often resembles a secondary deposit, which can confuse radiologists and other clinicians. The significance of this article is in the domain of timely and adequate diagnosis and differential diagnosis because the initially presented symptoms can resemble numerous neurological or systemic diseases, which requires the clinician to be constantly aware of such rare pathological processes as well as a good knowledge of the curriculum of neuroradiological diagnostics.

Abbreviations: *intravascular papillary endothelial hyperplasia (IPEH); Masson's tumor (MT); multi-detector computed tomography (MDCT); Magnetic resonance imaging (MRI); diffusion-weighted (DW); Apparent Diffusion Coefficient (ADC).*

Sažetak

Uvod: Intravaskularna papilarna endotelna hiperplazija predstavlja retku reaktivnu proliferaciju endotelnih ćelija oko organizovanog tromba, lokalizovana u proširenom krvnom sudu, hematoma ili vaskularnoj leziji. Simptomi koji nastaju kao rezultat ove lezije zavise od lokalizacije samog patološkog procesa. Lokalizacija u kičmenom kanalu je izuzetno retka, pa je do sada objavljeno nekoliko kliničkih slučajeva. Cilj ovog rada je predstavljanje radiološko-dijagnostičkih kriterijuma ove lokalizacije. **Materijal i metode:** Izvršeno je sistematsko pretraživanje baza podataka PubMed, Medline, Radiopedia. **Rezultati:** Magnetna rezonanca pokazuje izointenzan do blago hipointenzan signal na T1 sekvenci, hiperintenzan signal na T2 sekvenci i nehomogeno postkontrastno prebojavanje na T1 C+ sekvenci. **Zaključak:** Neurološki simptomi su često posledica drugih, pre svega somatskih, bolesti i patoloških procesa različite lokalizacije. Na magnetnoj rezonanciji glavne karakteristike intravaskularne papilarne endotelne hiperplazije su izointenzna do hipointenzna pojava na T1 i hiperintenzna pojava na T2 ponderisanim slikama magnetne rezonance, a takođe pokazuje poboljšanje nakon intravenske primene gadolinijuma. Veoma je važno imati na umu da intravaskularna papilarna endotelna hiperplazija često podseća na sekundarni depozit koji može zbuniti radiologe i druge kliničare. Značaj ovog članka je u domenu pravovremene i adekvatne dijagnoze i diferencijalne dijagnoze, jer prvobitno ispoljeni simptomi mogu ličiti na brojne neurološke ili sistemske bolesti.

LITERATURE

1. Haluk NO, Ozer M. Spinal intravascular papillary endothelial hyperplasia. Case report and review of the literature, *British Journal of Neurosurgery*. 2023;37:4:738-40.
2. Behera BR, Panda RN. Masson Hemangioma-An Unusual Cause of Thoracic Compressive Myelopathy. *World Neurosurg*. 2017;98(876):e9-876.e13.
3. Gu HL, Zheng XQ. Intravascular papillary endothelial hyperplasia as a rare cause of cervicothoracic spinal cord compression: A case report. *World J Clin Cases*. 2021;9(34):10681-8.
4. Porter DG, Martin AJ. Spinal cord compression due to Masson's vegetant intravascular hemangioendothelioma. Case report. *J Neurosurg*. 1995;82(1):125-7.
5. Ali SZ, Farmer PM. Masson's hemangioma of spinal meninges causing cord compression with paraplegia. *Ann Clin Lab Sci*. 1994;24(4):371-5.
6. Mozhdhipanah H, Samiei F. Masson's hemangioma: A very rare cause of spinal cord compression. *Neurol India*. 2013;61(1):89-90.
7. Giannitto C, Mercante G, Spriano G, Natoli R, Gaino F, Lofino L, Esposito AA, Giannitto N, Vatteroni G, Fiamengo B, Vidiri A, Politi LS, Balzarini L. CT and MRI findings of head and neck Masson's tumor: a rare case report and systematic review of the literature. *Reports in Medical Imaging*. 202;14:53-64.
8. Voruz F, Arnoux G. Intravascular papillary endothelial hyperplasia (Masson's tumor) of maxillary sinus. *Braz J Otorhinolaryngol*. 2022;88(1):141-5.
9. Park KK, Won YS. Intravascular papillary endothelial hyperplasia (Masson tumor) of the skull: Case report and literature review. *J Korean Neurosurg Soc*. 2012;52(1):52-4.
10. Petry M, Brown MA. Multifocal intravascular papillary endothelial hyperplasia in the retroperitoneum and spine: a case report and review of the literature. *J Magn Reson Imaging*. 2009;29(4):957-61.
11. Guillevin R, Vallee JN. Spine metastasis imaging: review of the literature. *J Neuroradiol*. 2007;34(5):311-21.
12. Mahapatra QS, Sahai K. Intravascular papillary endothelial hyperplasia: An unusual histopathological entity. *Indian Dermatol Online J*, 2015;6:277-9.
13. Barritt AW, Merve A. Intracranial papillary endothelial hyperplasia (Masson's tumour) following gamma knife radiosurgery for temporal lobe epilepsy. *Pract Neurol*. 2017;17:214-7.
14. Narwal A, Sen R. Masson's hemangioma: A rare intraoral presentation. *Contemp Clin Dent*. 2013;4:397-401.
15. Clifford PD, Temple HT. Intravascular papillary endothelial hyperplasia (Masson's tumor) presenting as a triceps mass. *Skeletal Radiol*. 2004;33:421-5.
16. Mardani P, Askari A, Shahriarirad R, Ranjbar K, Erfani A, Anbardar MH, Moradmans S. Masson's tumor of the hand: An uncommon histopathological entity. *Case Rep Pathol*. 2020;2020:4348629.
17. Oktar N, Ozer H. Spinal intravascular papillary endothelial hyperplasia. Case report and review of the literature. *Br J Neurosurg*. 2019;1-3.
18. Bhalla N, Husband DJ. Radiotherapy for a benign cause of cauda equina compression in a known case of breast carcinoma. *BMJ Case Rep*. 2013;bcr2013009549.