CASE REPORTS

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Masson's tumor of the thoracic spine: a rare cause of slowly progressive paraplegia

Masonov tumor torakalnog dela kičmenog stuba: redak uzrok sporoprogresivne paraplegije

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Abstract

Introduction. Intravascular papillary endothelial hyperplasia is an unusual reactive proliferation of endothelial cells around an organized thrombus, which occurs either in a dilated blood vessel, hematoma, or preexisting vascular lesion. These tumors rarely affect the central nervous system. Symptoms depend on the localization of the process itself. Localization in the central nervous system is limited to the intracranial space. Localization in the spinal canal is extremely rare, and only a few clinical cases have been described so far in the literature. **Case report.** A 67-year-old female patient was examined neurologically initially due to bilateral weakness of the lower extremities, accompanied by a feeling of pain and muscle tension, dominantly in the upper legs, more to the right. The complaints were present a year ago, and before that period, the patient was in a stable state of health. Due to a severe neurological deficit and the need for detailed exploration, the patient was hospitalized. A diagnosis was performed, which showed the localization of the pathological process in the thoracic 5–6 region of the spinal column. Decompression surgery was performed, and with the *ex tempore* findings metastasis was ruled out. Definitive pathohistological findings proved Masson's tumor. After the operation, the neurological weakness recovered. **Conclusion.** Masson's tumor, although rarely localized in spinal canal, is curable if it is correctly diagnosed and if an adequate therapeutic approach is applied. The initially presented symptoms may resemble numerous neurological or systemic diseases, which requires the clinician to be continuously aware of such rare pathological processes.

Key words:
diagnosis; differential diagnosis; histological techniques; magnetic resonance imaging; neoplasms, vascular tissue; neurosurgical procedures; spinal canal.

Apstrakt

Uvod. Intravaskularna papilarna endotelna hiperplazija je neobična reaktivna proliferacija endotelnih ćelija oko organizovanog tromba, koja se javlja ili u proširenom krvnom sudu, hematomu ili u postojećoj vaskularnoj leziji. Ovi tumori retko zahvataju centralni nervni sistem. Simptomi zavise od lokalizacije samog procesa. Lokalizacija u centralnom nervnom sistemu ograničena je na intrakranijalni prostor. Lokalizacija u kičmenom kanalu izuzetno je retka, a do sada je u literaturi opisano samo nekoliko kliničkih slučajeva. **Prikaz bolesnika.** Bolesnica stara 67 godina je prvobitno pregledana od strane neurologa zbog obostrane slabosti donjih ekstremiteta, praćene osećajem bola i napetosti mišića, dominantno u natkolenicama, više desno. Tegobe su se pojavile godinu dana ranije, pre toga je bila stabilnog zdravstvenog stanja. Zbog teškog neurološkog deficita i potrebe za detaljnim ispitivanjem, bolesnica je hospitalizovana. Urađena je dijagnostika kojom je utvrđen patološki proces u torakalnom 5–6 regionu kičmenog stuba. Urađena je operacija dekompresije, a *ex tempore* nalazom isključena je metastaza. Definitivnim patohistološkim nalazom utvrđen je Masonov tumor. Neurološka slabost se povukla posle operacije. **Zaključak.** Masonov tumor, iako je retko lokalizovan u kičmenom kanalu, izlečiv je ukoliko se pravilno dijagnostikuje i primeni odgovarajući terapijski pristup. Prvobitno ispoljeni simptomi mogu ličiti na mnogobrojne neurološke ili sistemske bolesti, zbog čega kliničari moraju neprekidno imati na umu tako retke patološke procese.

Ključne reči:
dijagnoza; dijagnoza, diferencijalna; histološke tehnike; magnetska rezonanca, snimanje; neoplazme, vaskularno tkivo; neurohirurške procedure; kičmeni kanal.

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Introduction

Intravascular papillary endothelial hyperplasia (Masson's vegetative hemangiothelioma) is a sporadic pathological condition affecting the nervous system [1]. It was discovered by Pierre Masson in 1923. However, a later neoplastic description by Henschen was criticized, who said that this type of lesion is reactive rather than neoplastic in nature. Although there are still controversies regarding the histological structure of Masson's tumor (MT), most authors believe it is a type of organized thrombus surrounded by a reactive proliferation of endothelial cells [2]. In 1975, Clearkin and Enzingen [3] described a lesion with unusual and marked thrombotic reorganization; MT was then renamed and became intravascular papillary endothelial hyperplasia (IPEH). Today, in the literature, these two names are equated [4]. MT can be classified into three types: the primary or pure, the secondary or mixed form, and the extravascular form. The primary/pure form, which is the most common form, typically occurs in a dilated vessel, most usually a vein rather than an artery, and arises in subcutaneous soft tissue. The secondary/mixed form presents in preexisting vascular abnormalities. Finally, the extravascular form, which is the least common form, occurs in hematomas. The symptoms given by this tumor depend on the localization, so they can often mislead clinicians, which speaks in favor of the importance of a multidisciplinary approach. Localization of Masson's hemangioma in the region of the central nervous system is rare and mostly limited to the intracranial space. Certainly, the localization of the process in the spinal canal (SC) is extremely unusual, so in the literature, there are only a few clinical cases of MT with this localization [5–7]. We presented the first diagnosed case of a patient with thoracic (TH) localization of IPEH, unique in Serbia.

Case report

A female patient, 67 years old, was initially examined for weakness of the lower extremities in September 2023. At that moment, the complaints lasted for about a year, and a month ago, she had severe pain and a feeling of tightness in the muscles of her legs, most intensely in her upper legs, which is why she used an orthopedic aid for movement (a walker). Due to the need for a detailed exploration, the patient was hospitalized in the Department of Neurology of the Clinical Hospital Center "Dr. Dragiša Mišović-Dedinje" in Belgrade, Serbia.

The patient's complaints started about a year before coming to the examination. Initially, the patient felt pain in the lower extremities, predominantly in the upper legs, which progressed over time in intensity and impact on gait. There was also a feeling of tightness in the leg muscles, more pronounced in the upper legs, more to the right. The weakness of the lower extremities progressed, so the patient started using a walker to move around. Two weeks before admission to the Hospital, she sprained her ankle. In her personal history, she is being treated for arterial hypertension, she had a tendon rupture of her right shoulder caused by trauma, and she is allergic to iodine.

The neurological examination was dominated by severe spastic paraparesis, more pronounced on the right. Her gait was spastic, paraparetic, possible only with the help of a lumbosacral (LS) spine walker. The sensation was reduced below the TH spine 6 level. The rest of the neurological exam was normal.

A magnetic resonance imaging (MRI) of the LS spine was performed on an outpatient basis (self-initiated): the hemangioma was central in the body of lumbar (L)5, occupying the entire height of the body. Smaller hemangiomas were found posteriorly in the body of L5 and TH10. Disc protrusions at the L3–L4 and L4–L5 levels were in close contact with the corresponding radix without direct radicular compromise. An electromioneurography examination of the lower extremities was performed: a moderately strong neurogenic lesion was found in all examined muscles except in the tibialis anterior bilaterally.

During the hospital treatment, the following diagnostics were performed: the MRI of the TH part of the spinal column and computed tomography (CT) of the chest. On the MRI (Siemens Aera 1.5 Tesla) of the TH part of the spinal column (Figures 1–3), paravertebral to the left, next to the bodies TH6 and TH5, a tumor mass that lied on the mentioned bodies could be seen with a wide base, with dimensions 18 × 38 mm, and cranio-caudal up to 50 mm.

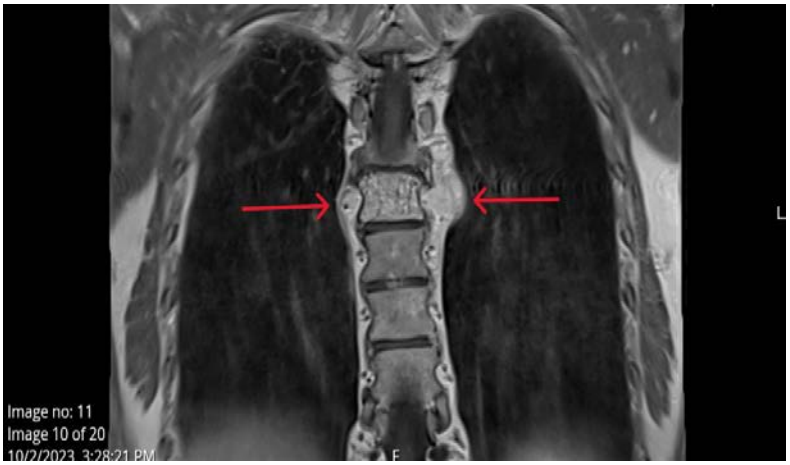

Fig. 1 – T2W coronal thoracic (TH): magnetic resonance imaging shows an affected TH 6 vertebra with a billateral tumor mass.

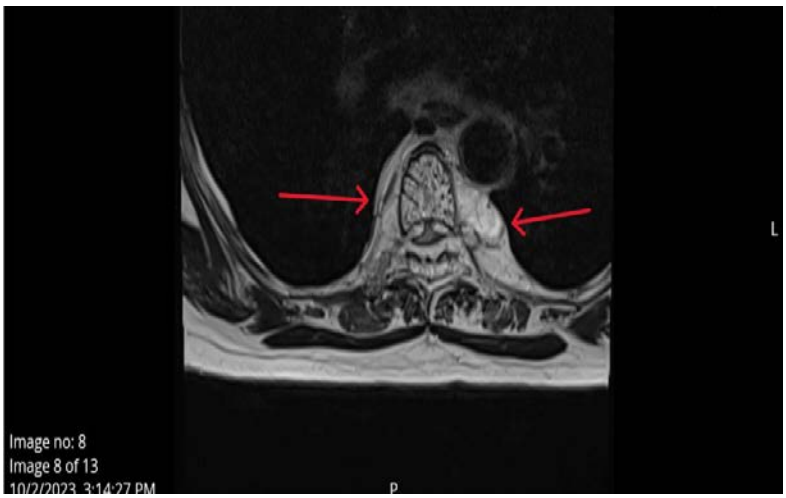

Fig. 2 – T2W axial: magnetic resonance imaging shows affected vertebra and paravertebral tumor mass more pronounced on the left.

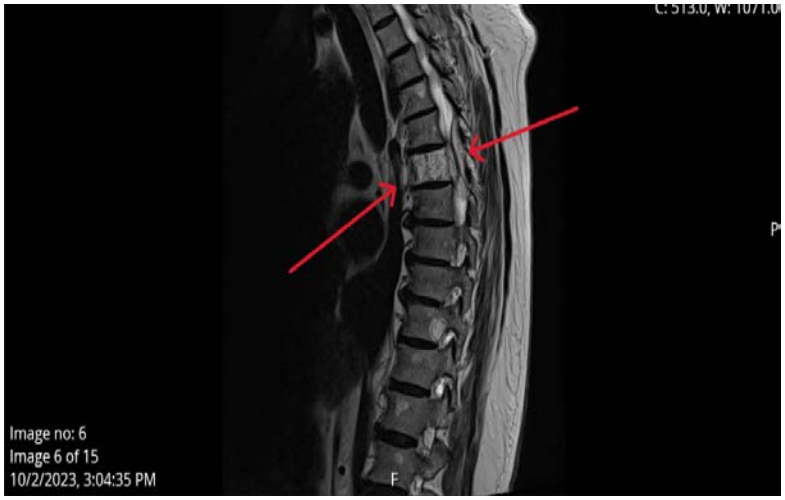

Fig. 3 – T2W sagittal: magnetic resonance imaging shows tumor mass in the spinal canal, complete obliteration.

There was an infiltration of the left underlying aspect of the body TH6, as well as the left pedicles, transversus, and lamina of this vertebra, without pathological fracture. There was an extension of the infiltrate into SC, which was critically stenosed at the level of the corpus TH6 with consequent compressive myelopathy in this segment. The differential diagnosis was thought to be in the direction of metastatic change of lung, breast, or pelvic region malignancy. The differential diagnosis, more precisely, was a suspected primary lung infiltrate with extension to underlying bodies or a package of pathological lymph nodes.

CT (Toshiba Aquilion One 320) finding of the chest was described as normal, without signs of consolidation and infiltration in the lung parenchyma. At the TH6 level of the vertebral body, the reduced bone structure could be seen with signs of destruction with edema and a soft tissue infiltrative component on both sides, more to the left. Differential diagnostic finding could have corresponded to a primary process on bone structures, but changes in the type of secondary deposits can not be ruled out either. Additional examination did not find any other pathological process.

After the orthopedist's examination, a decision was made on decompressive radical surgery. However, already during the extemporaneous biopsy, it was clear that it was not a metastatic tumor nor another malignant tumor, and tumor resection was performed while preserving the TH6 and TH5 vertebrae with stabilization on upper and lower levels (TH4 and TH7). MT was proven on the definitive pathohistological (PH) examination. The PH finding is given in the following lines. The microscopic analysis showed the following: several bone fragments with total dimensions of 3.5 × 3.5 × 1 cm; two soft tissue fragments with total dimensions of 2 × 1 × 0.5 cm. In the analyzed material, the following can be observed: fragments of trabecular bone, thin beds between which there is a vascular lesion represented by anastomosing blood vessels, thin wall, and dilated lumen with zones of papillary endothelial hyperplasia (Masson's tumor). The finding corresponds to an intraosseous hemangioma.

The postoperative course went smoothly. Within a month after the surgery, the neurological deficit gradually resolved.

Discussion

IPEH is a very rare benign pathological lesion that presents as a reactive vascular lesion with a tendency to expand and form a compressive mass. The mass was initially nominated as MT, but later, the name itself suffered numerous criticisms. Bearing in mind that the lesion itself is not malignant, considering the localization and the symptoms it causes, it still has the characteristics of a tumor. Therefore, the name was "softened" by introducing a term that actually corresponds to the PH structure of this pathological process. No preferential age of onset of IPEH has been shown, but data in the literature indicate there are differences in relation to gender. Namely, intracranial localization is significantly more common in women, with a ratio of 4:1 compared to men. Localization in SC, although extremely rare, affects men more often [4]. Usually, the articles published to date found IPEH in the skin and subcutaneous tissue of the head, neck, or extremities, and also in the oral mucosa, lips, sinus cavities, parotid gland, thyroid gland, lungs, superior *vena cava*, adrenal gland, renal vein, forearm, foot, intracranial. So far, only a few cases in SC have been published [8–12]. Our case report is the first to talk about the localization of the pathological process in the TH part of SC in Serbia and one of the few published worldwide. In our patient, it is most likely an MT due to a previous hemangioma of the vertebral body.

Considering the initially observed localization of the pathological process, as well as taking into account that the complaints occurred in a patient who was previously in a stable state of health, and bearing in mind the continuous progressive-deteriorative course of clinical symptoms, an expansive process with a possible primary cause was initially suspected with localization in the lung parenchyma or breast tissue, or in the pelvic organs. However, it turned out to be a mixed form of MT, formed on the site of a previous hemangioma in the body of the TH vertebra.

The pathogenesis of IPEH is still controversial. Different authors give different explanations for this lesion – from the fact that it is an overreaction to the normal process of thrombus reorganization to the fact that it is a benign proliferation of endothelial cells with secondary thrombosis and fibrin deposition [4]. Significantly, the diagnosis of IPEH itself is very demanding and challenging due to the presence of nonspecific MRI features. It is always necessary to consider cavernous/capillary hemangioma, Kaposi's sarcoma, endovascular papilloma and endothelioma, then schwannoma, neurofibromas, and arteriovenous malformations [12], and, in our case, secondary deposit as well. MRI and CT findings showed a pathologic substrate in our case, which was presented with tumor mass with infiltration of surrounded structures but without pathological fracture. In the essence of the pathological process, we found a hemangioma of the vertebral body, which transformed over time into a mixed form of MT.

The symptoms that appear depend on the localization of the pathological process. Namely, the localization of IEPH in SC can lead to pain in the chest and back, numbness of the lower extremities, paresis or paralysis, then bladder dysfunction as a result of compression of the spinal cord or cauda equina [1]. Treatment is considered in a situation where persistent pain and compressive symptoms occur. If diagnosed in time, complete surgical resection is possible, which is the most desirable treatment modality for the patient, with the most favorable outcome.

In our case, the patient underwent surgical treatment with complete resection, after which she recovered well, and further follow-up followed. Radiotherapy, as an adjuvant therapeutic modality, is considered when the lesion cannot be completely removed or when a recurrence occurs. To our knowledge, there is only one clinical case of IPEH localized in SC in which radiotherapy was applied after incomplete resection and with satisfactory success [13].

Conclusion

Although rarely localized in the spinal canal, MT is curable if diagnosed correctly and if an adequate therapeutic approach is applied. Better recognition of MT in the central nervous system can speed up and facilitate diagnosis. Certainly, magnetic resonance imaging is an irreplaceable method for preoperative orientation and operative plan. The significance lies in the domain of differential diagnostics because the initially presented symptoms may resemble numerous neurological or systemic diseases, which requires the clinician to be continuously aware of such rare pathological processes.

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