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Primary spinal cord melanoma – a case report

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Abstract

Authors present a case of a 57-year-old woman with primary spinal cord malignant melanoma. Intramedullary localization of primary melanoma is extremely rare. The patient presented neurological deficits such as lower limbs paresis and sensory loss. MRI examination showed intramedullar tumor located on the Th10 vertebra level. Surgical treatment with total removal of tumor was performed. Histopathological study confirmed melanoma. Subsequent chemotherapy was given. Tumor was successfully treated by neurosurgery; radio- and chemotherapy with disease free follow up of 9 months. Surgical treatment of melanoma in this location is extremely important as it leads to regression of neurological symptoms and improvement of the quality of life.

Key words: intramedullar melanoma, surgery, tumor.

Introduction

Primary melanomas located in the Central Nervous System (CNS) are rare and constitute of 1% of all tumors, while intraspinal location is even less frequent [5,6,12,14]. From the year 1906 only 39 cases of primary melanoma in spinal cord structures were reported in literature [15]. Melanomas located in CNS, especially those with intramedullar location, are usually metastatic [3,5,6,12,14]. The most frequent location for primary melanomas is thoracic part of spinal cord [9]. Main clinical symptoms caused by spinal cord melanoma are pain and neurological deficits, placed dependently on tumor location. In MRI scans a typical tumor gives hyperintensive signal in T1 scans and hypointensive in T2 [6].

We present a rare case of a female patient hospitalized due to progressing lower limbs paresis caused by tumor with intramedullar location on the level of Th10 vertebra.

Clinical presentation

A 57-year-old female was admitted to the Neurosurgical Department due to spinal pains and lower limbs pains with paresis progressing from 2 months. On admission, the patient presented lower limbs paresis, located mainly in the right extremity, with assisting superficial sensibility disorder represented by weakness below umbilical level and sphincters insufficiency. The patient's MRI spinal scans showed tumor in vertebral canal, with intramedullar location in the level of Th10 vertebra, showing post-contrast intensification. Radiological image suggested ependymoma or astrocytoma (Fig. 1).

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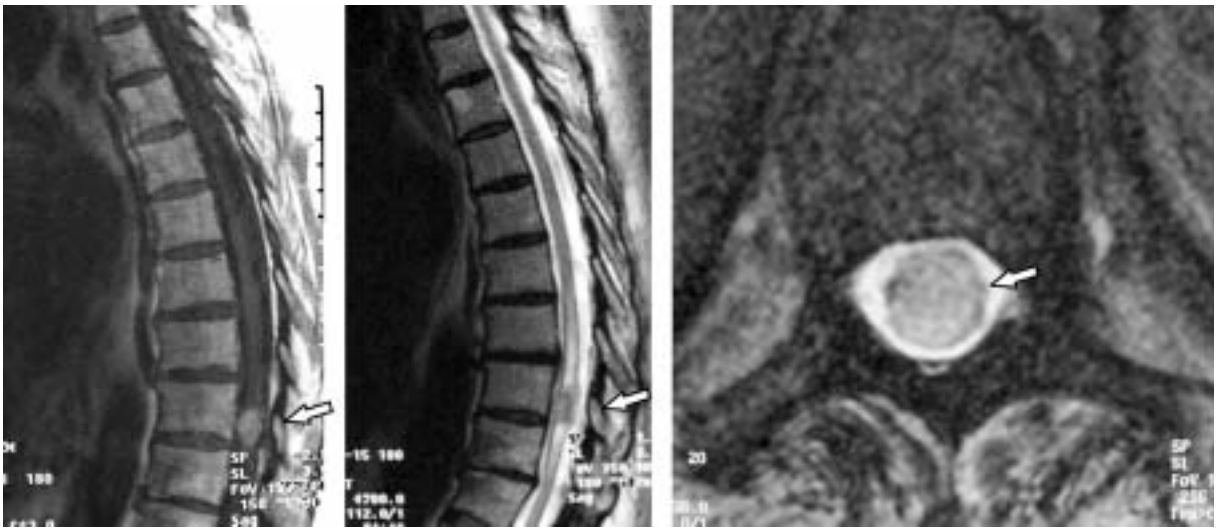

Fig. 1. Thoracic spine MRI examination (T1 and T2) and axial T2 image. Tumor located intramedullary in the level of Th10 vertebra (arrow).

Due to clinical symptoms of lower limbs paresis and tumor mass on radiological image, the patient was qualified for surgical treatment. Spinal cord was enlarged, edematous, with changed color and fully filled the spinal canal. The cord was cut in middle line. Semi-liquid, thick, gelatinous substance of black color was leaking under high pressure from the medulla incision. Subsequently, the rest – solid, color from grey to black – of the tumor was removed. The tumor with distinct borders was connected with medulla, however it was possible to separate it without bleeding. Macroscopically, tumor was totally removed.

Neurological state of the patient improved after surgery with gradual regression of paresis. The patient was subjected to rehabilitation. On discharge the patient was walking on her own, with the help of crutches. The patient was sent to the Department of Chemotherapy of Oncological Center in Łódź, Poland for further treatment. On the staging examination no other neoplastic foci were revealed, so the medulla location was confirmed as primary site. The patient was also treated with chemotherapy.

Follow-up examination was performed with magnetic resonance 6 and 9 months after the procedure.

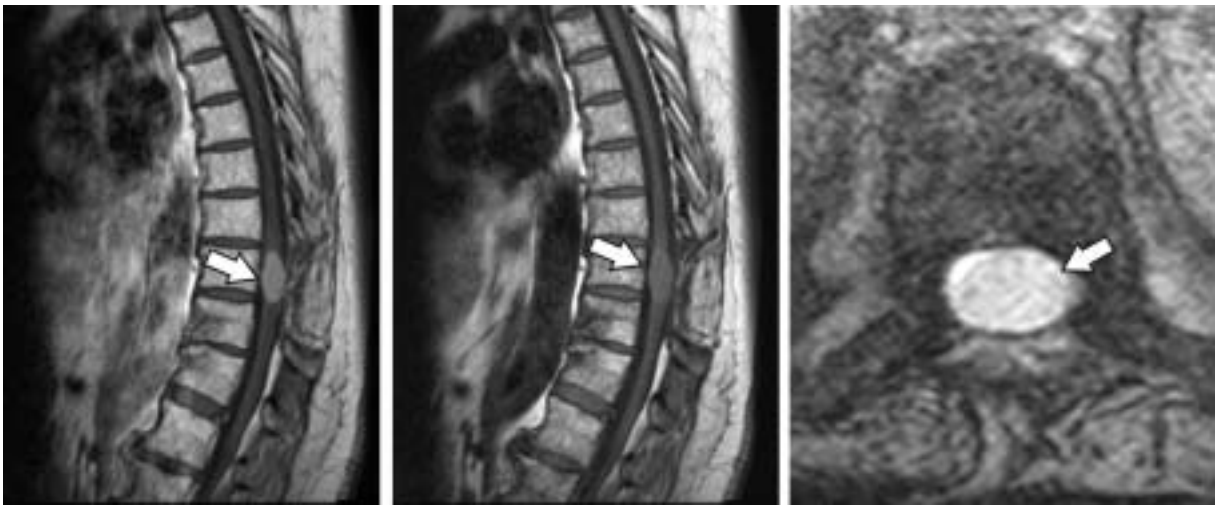

Fig. 2. Follow up MRI examination 12 months after surgery. Intramedullary tumor is present in previous location (arrow).

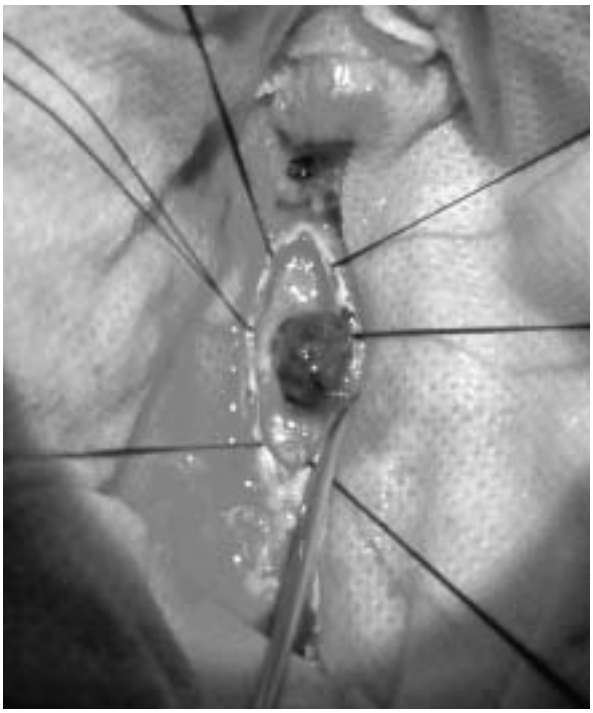

Fig. 3. Intraoperative image, visible dark red-black tumor.

On 9-month examination a tumor recurrence tumor was shown in the operated location. The second procedure was delayed due to the patient's good clinical state and lack of neurological deficits.

A year after the first procedure the patient returned to the Neurosurgery Department with massive paresis of lower limbs, intensified in the right extremity and urinary incontinence. MR examination of thoracic part of spine showed the tumor in the previous location. The second surgical procedure was performed. Following first surgery traces the medulla was reached. Vertebral canal was filled tightly, with indent in place of previous laminectomy. On a dorsal part of the medulla through the previous cut a part of red-black spindle-shaped tumor was visible with unequal surface. The tumor mass was larger than the one removed during the first procedure (Figs. 2 and 3). The tumor had mainly interspinal location, border was partly distinct, however the rest was blurred. Whole neoplasm was removed in part that was possible to be separated from the cord, while where border was not sharp, the tumor was removed with a slight margin of pathological tissue left (Fig. 4).

After histological examination malignant melanoma was confirmed. Lower limbs paresis gradually disappeared. Two weeks after the procedure patient had only a slight dysfunction of the right limb but was walking on her own with the help of crutches.

Histopathological study

On histopathological examination malignant melanoma was diagnosed. The tumor was densely cellular and composed of spindle, epithelioid, and pleomorphic cells with abundant cytoplasm with melanin deposits (Fig. 5).

The recurrent tumor displayed very similar morphology with majority of spindle cells (Fig. 6).

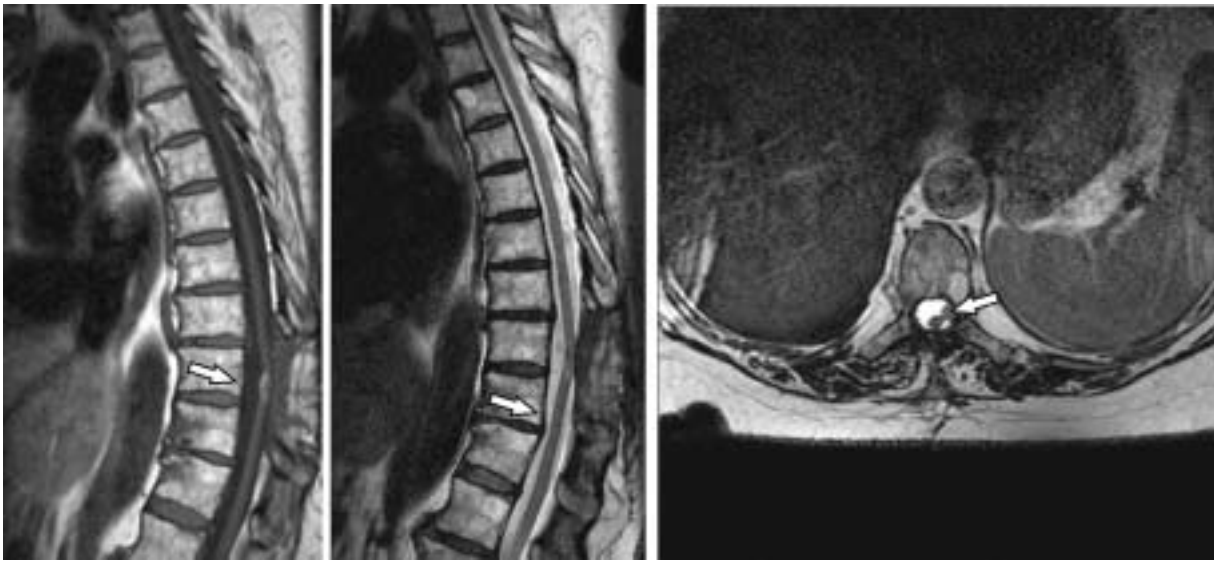

Fig. 4. Follow up MRI examination one month after second surgery. Presence of tumor remnants (arrow).

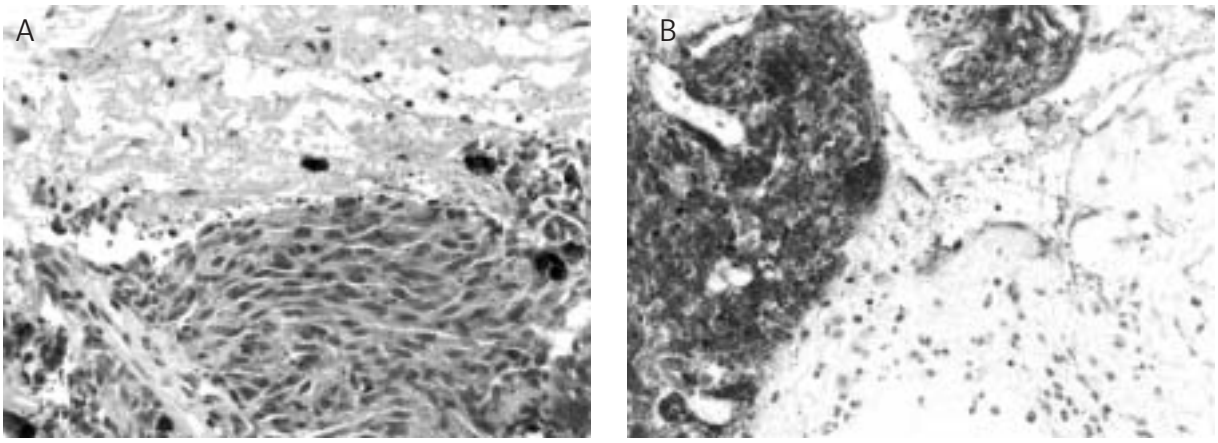

Fig. 5. Spinal cord invaded by the primary malignant melanoma that showed a wide spectrum of cells – some fields consisted of spindle cells **(A)** and other of epithelioid and pleomorphic highly pigmented cells **(B)**, HE, magnification × 200.

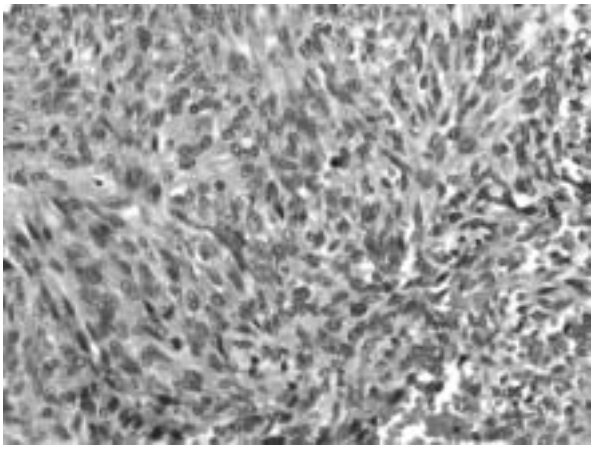

Fig. 6. The recurrent tumor showed the similar morphology with prominent spindle cells, HE, magnification × 200.

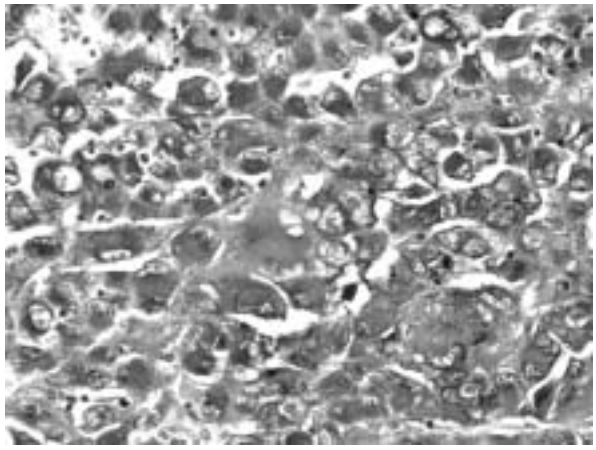

Fig. 7. The strong reactivity of HMB-45 in tumor cells, magnification × 400.

Immunohistochemical study in both primary and recurrent tumor revealed reactivity with HMB-45 (Dako-Cytomation, Denmark) and S-100 protein (Dako-Cytomation, Denmark) and high proliferative rate of Ki67 (Figs. 7 and 8).

Discussion

Primary melanomas are very rare and constitute only 1% of all melanomas located in the Central Nervous System (CNS) [5,6,8,12-14], while metastatic tumors are more frequent and make 3% to 16% of secondary neoplasm foci in CNS [1,3,11,17]. In the material of our Department from about 30 years, it was the first case of primary melanoma in interspinal location. Metastasis was excluded by detail examination of skin, endoscopy, and radiology. As only 39 cases of melanoma in such location have been described in literature since 1906, we believe it is worth sharing [15].

Primary spinal cord melanomas originate from melanoblasts, which derive from nervous chord during embryogenesis. Criteria for deciding whether it is a primary melanoma or not were presented by Hayward [8]: lack of melanoma outside CNS, lack of foci in other parts of CNS, histological examination confirming melanoma diagnosis. In literature, there are very few cases of melanomas in interspinal location [2,5-7,12-15,18]. In 1906 Hischberg was first describe

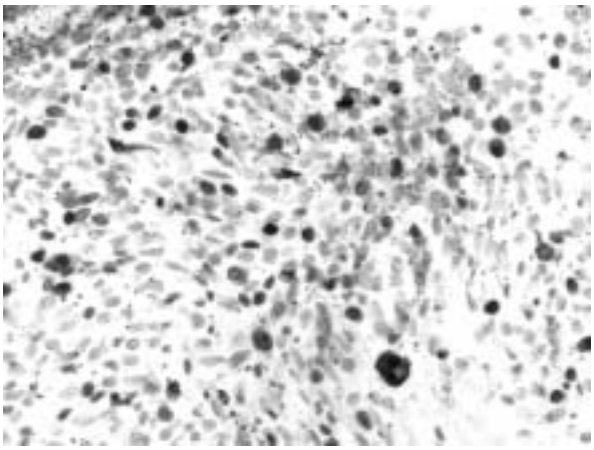

Fig. 8. The high proliferative rate of Ki67 in primary spinal cord melanoma – nuclear immunostaining and brown cytoplasmatic deposits of melanin in tumor cells, magnification × 200.

such location of melanoma [10,15]. Since that time 39 other cases of primary interspinal melanoma have been reported [15]. The most frequent location is thoracic part of the spinal cord [9]. The characteristic sign of melanoma on MRI is hyperintensive signal in T1 scan, which is connected with the presence of melanin, and hypointensive signal in T2 [6].

Primary CNS melanomas grow slow and seem to be less malignant than skin melanomas metastatic to the CNS [13]. Metastasis of skin melanoma grows very fast and causes death within average 6 months [4,16]. Whereas, primary CNS melanomas grow slow and metastasize within CNS, mainly in the same place where first found [13]. The average time of survival after surgical treatment and chemotherapy is 6 years and 7 months [12].

Treatment of primary malignant CNS melanoma is difficult. Due to frequent recurrence it is suggested to perform radical surgical resection, as far as it is possible in spinal cord tissue. In the case of our patient, it was impossible to perform total resection and complementary treatment with chemotherapy was implemented. It is said that when resection is suboptimal complementary treatment with chemotherapy and radiotherapy should be included, however their effectiveness is under debate [12].

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