

Long-term follow up of a patient with primary cervical spinal cord meningeal melanocytoma

Dlouhodobé sledování pacienta s primárním meningeálním melanocytomem krční míchy

Dear Editor,

Among the uncommon tumors of the spinal cord, one of the rarest and unfamiliar is primary melanoma. Clinical course of primary spinal melanoma is unpredictable and little is known about the best management strategy and its recurrence. Larson et al reported that the average life expectancy of primary central nervous system (CNS) melanoma is approximately seven years after surgery with radiotherapy [1]. There is no study in the literature regarding long-term follow up of primary spinal melanoma to assess the pattern of recurrence and further management of such recurrent cases. We encountered a patient with melanoma originating from the meninges of the cervical spinal cord which recurred 11 years after total gross excision. The patient underwent a re-do surgery with a favorable outcome. The current case illustrates that patients with primary CNS melanoma should be kept on regular follow up as recurrence may happen even after years of gross surgical resection.

Sixty-eight-years-old woman was admitted to our hospital with complaints of gradually progressive numbness and weakness of both upper and lower limbs since the past six months. She was previously admitted to this hospital in 2005 with the same complaints and was diagnosed to have a C2–C4 intradural-extramedullary (IDEM) mass (Fig. 1A–C) which was managed by C2–C4 left hemilaminectomy and total gross tumor excision. Biopsy was consistent with melanocytoma. Post-operatively, the patient improved and remained symptom-free for the past 11 years when again she started to develop signs of spinal cord compression. Neurological examination revealed hypoesthesia below C5 dermatomal level with Gr III spastic quadriplegia (power 3/5 in both upper limbs and 2/5 in both

lower limbs) with exaggerated deep tendon reflexes and a positive Babinski sign on both sides. MRI of cervical spine revealed left hemilaminectomy defect at C2–C4 with an IDEM mass at the level of C2–C3 which was hyperintense on T1- and hypointense on T2-weighted images causing marked compression and displacement of the spinal cord with patchy edema (Fig. 1D–G). The solitary, solid, contrast-enhancing mass measured 25 mm × 15 mm in size.

In view of the possible diagnosis of recurrent melanocytoma, the patient underwent re-exploration through a standard posterior cervical midline approach. Intraoperatively, the dura was bulging and black in color due to the underlying intradural mass. The mass was indistinct, black and hypervascular, and was adhered to the dura and arachnoid, which was infiltrated (Fig. 2A). The tumor compressed, although it did not invade the spinal cord and was decompressed using a microsurgical technique. However, part of the infiltratively growing portion could not be removed.

Standard histopathological examination revealed dense fibrocollagenous tissue with infiltrating dyscohesive tumor cells. The tumor cells showed mild pleomorphism, high nuclear-cytoplasmic ratio, round to oval nuclei and moderate cytoplasm containing coarse melanin granules. MIB-1 index was 1–2% consistent with meningeal melanocytoma (Fig 2B–C).

Postoperatively, the patient was kept on ventilatory support for seven days after which gradual weaning started. Given the possibility of metastasis, CT images of chest/abdomen/pelvis and MRI of the brain and total spine were obtained which found no significant abnormalities. A thorough full-body dermoscopy, retinal examination and anoscopy did not identify any melanocytic lesions, consistent with

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a primary non-metastatic spinal melanoma. The immediate postoperative MRI scans of the spine confirmed tumor decompression with minimal residual findings; the signs of cervical myelopathy were still recognizable, although significantly decreased and at

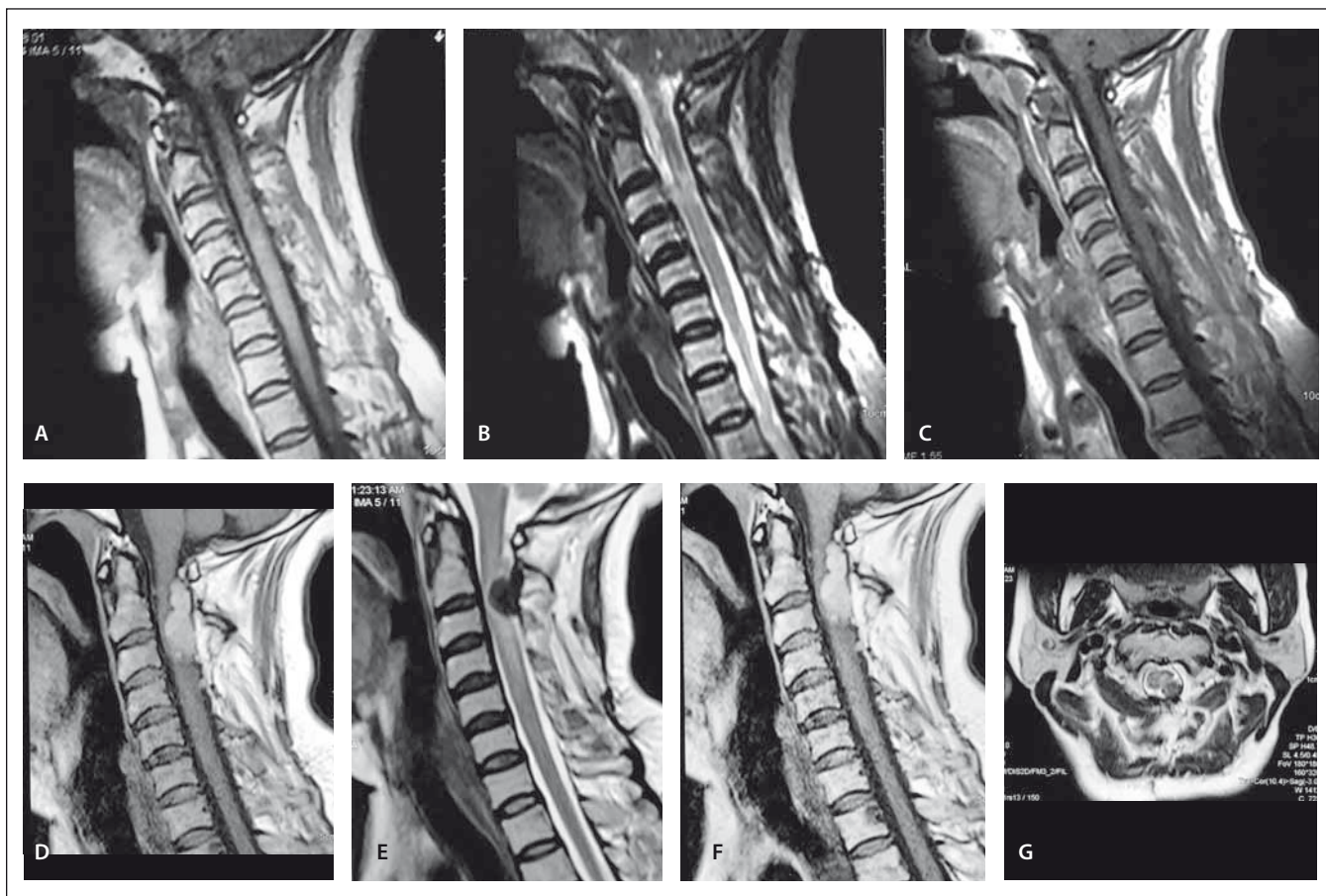


Fig. 1. A – T1W; B – T2W; C – contrast MRI of cervical spine done 4-yr after the first surgery showing no residual/recurrent lesion; D – T1 hyperintense; E, G – T2 hypointense; F – contrast-enhancing intradural-extramedullary lobular mass at C2–C3 11 years after the first surgery suggestive of recurrent tumor.

the time of this report, the patient was in remission from her melanoma. The patient underwent radiotherapy soon after her vital signs were stable. For monitoring of the patient, clinical exams and serial MRIs were performed every six months. Six months after surgical resection, the patient remained neurologically stable and gradually regained motor function in the muscles of the upper and lower limb (postoperative strength grade 4/5). The concomitant hypoesthesia improved in the postoperative follow up.

Hayward in 1976 established a guide for classification of primary CNS melanoma, which described how primary CNS melanoma is considered a diagnosis of exclusion [2]. Location in the spine is especially rare, with only a few cases described in the literature [3,4]. Within the spine, it presents as intradural extramedullary masses, mostly found in the upper cervical region, as the melanocytes are mostly concentrated in this region, with features of cord com-

pression such as numbness and spastic paresis [5].

On CT, these lesions present as well-defined, isodense to hyperdense, homogenous, and contrast enhancing mass lesions. Signal characteristics on MRI include isointense or hyperintense on T1-weighted images and isointense or hypointense on T2-weighted images which give a heterogeneous enhancement on post-contrast images and show blooming on gradient images [6].

For primary CNS melanocytic neoplasms, a complete tumor resection is preferred because it can cure well-differentiated and intermediate-grade melanocytomas (IMGs) and most melanomas. Radiotherapy is recommended for the incomplete resection of IMGs and melanomas. The risk of recurrence of low-grade melanocytomas is less clear and careful observation is warranted [7,8].

Survival rate for CNS melanomas is not exactly known [1,6] and the efficacy of

radiotherapy and chemotherapy in CNS melanoma is still controversial [9,10]. At the present time, there is no data concerning the effect of radiotherapy or chemotherapy on survival. Our case underwent complete surgical resection of the tumor and remained symptom-free for at least 11 years without radiotherapy and was monitored both radiologically and neurologically over the years when she again presented with recurrence. Thus, we can assume that in meningeal melanocytoma, the primary variable for survival is the extent of resection.

Despite extensive Medline search, we could not find any article regarding long-term follow up of any primary spinal melanoma to assess the recurrence and further management of such patients. Our case survived for more than 11 years which indicates that long term survival is possible, provided the tumor is meticulously removed. The current case illustrates that complete resection alone, without any adjuvant therapy, may result in a favorable outcome of primary spinal cord

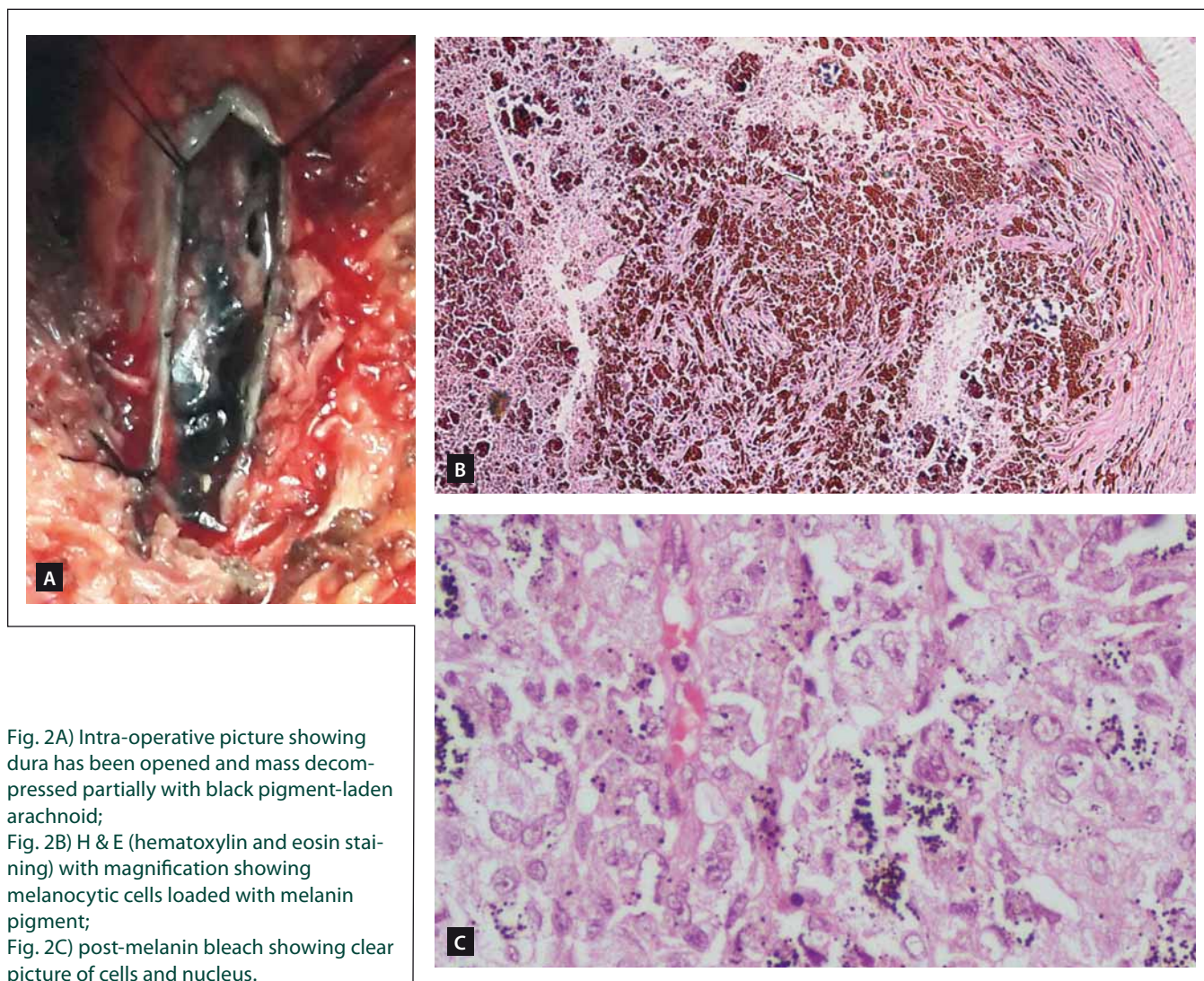


Fig. 2A) Intra-operative picture showing dura has been opened and mass decompressed partially with black pigment-laden arachnoid;
 Fig. 2B) H & E (hematoxylin and eosin staining) with magnification showing melanocytic cells loaded with melanin pigment;
 Fig. 2C) post-melanin bleach showing clear picture of cells and nucleus.

melanocytoma and in the event of the recurrence, re-exploration and gross total tumor excision along with concurrent radiotherapy may enhance patient outcome.

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