Spinal Melanocytoma: A Case Report

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Abstract

Meningeal melanocytoma, a rare tumor of melanocytic origin, usually arises from intracranial structures but may also arise from the spinal cord. They are solitary low-grade tumors and do not invade the peripheral structures. They can occur in all the age groups and are more frequent in the fifth decade with a slightly female predominance. They localize frequently intradural extramedullary especially at the cervical or thoracic region. Most authors agree that complete surgical excision is the best treatment. The role of radiotherapy and chemotherapy still remain controversial. Here, we report a patient with a melanocytoma in the right cervicothoracic medullary region which was resected gross totally.

Keywords

Cervicothoracic; Spinal melanocytoma; Surgery

Introduction

Meningeal melanocytoma is a benign melanocytic tumor that originates generally from the melanocytes in the posterior fossa or along the spinal cord. They are benign but locally aggressive lesions and are very rarely associated with spinal localizations. This tumor generally occurs as an extraxial mass that compresses adjacent neural structures and produce myeloradiculopathy. We report a new case of a 62-year-old man presented with a walking difficulty.

Case Report

A 62-year-old man presented with a 3-year history of walking difficulty. His medical history was relevant for a prior operation for a cervicothoracic mass whose pathology had been reported as a meningioma 20-years ago. His neurological examination revealed a first motor neuron type hemiparesis with the muscle strength 4/5 and 2/5 at right arm and leg, respectively. Spinal cervical magnetic resonance imaging (MRI) showed an extramedullary tumor mass at C5-T1 level (Figure 1).

The patient was operated on with the aid of an operation microscope. Prior laminectomy defect was enlarged to the adjacent inferior lamina. Intradural extramedullary tumor tissue was black in color and hemorrhagic. Some of its fragments were sent for pathological examination before removal of the remaining mass by means of ultrasound aspiration. Gross total removal of the tumor was achieved. Microscopically the tumor was composed of polygonally shaped cells with cytoplasm rich in melanin. Melanocytic cells had round nuclei with prominent nuclei. The lesional cells were immunopositive for S100 protein, melen-A and MART-1 without expression of significant nuclear staining (Figure 2). The histological diagnosis was consistent with a meningeal melanocytoma. The patient was died 10 months after the operation because of myocardial infarction.

Discussion

Meningeal melanocytoma, a rare tumor initially named by Limas and Tio, describes a benign tumor of melanocytic origin arising from the leptomeninges [1]. It usually originates from intracranial structures and the spinal cord where it shows intradural and extramedullary localisation. The age of onset of spinal meningeal melanocytoma in the literature ranged between 16 and 75 years of age without sex predominance. The age of onset in our patient was within this range.

Spinal cord melanocytomas generally originate from the pigmented cells of the spinal leptomeninges at any level of the spinal cord but especially at the craniovertebral junction [2]. It may be diffuse.

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Received: August 26, 2014 Accepted: October 13, 2014 Published: January 14, 2015
or localized. The melanocytoma in our case was localized to the cervicothoracic region of the spinal cord.

Meningeal melanocytoma is an encapsulated, black mass adhered tightly to the neighboring dura [3]. Some meningiomas may appear as black due to the presence of hemosiderin resulting from intratumoral hemorrhage [2]. Intraoperative differentiation of pigmented schwannoma, pigmented meningioma and melanoma may be difficult [2,4-6]. The melanocytoma of our case was also black in color and attached to meninges. There was no capsule delineating the tumor although there was a clear distinction between the medullary parenchyma and the black tumoral tissue, contrary to the case reported by Turhan et al. [2]. The absence of a tumor capsule makes it impossible to claim that gross total removals exclude the possibility of minimal residual disease.

The distinction between a meningeal melanocytoma and a malignant meningeal melanoma can be very difficult to discern. Features favoring a meningeal melanocytoma diagnosis are a duration of symptoms of more than 1 year, imaging features similar to a meningioma, and uniform cytological features with a predominance of spindle cells and a low mitotic rate [7]. The Ki67 proliferation index of melanocytomas is less than 12% and there was no significant nuclear staining with Ki67 in our case. The differential diagnosis of spinal melanocytoma included intradural extramedullary tumors like primary or secondary melanocytic tumors (especially well differentiated melanocytoma and malignant melanoma) schwannoma, meningioma and melanoblastoma [2,5,8-13].

Long duration of symptoms and similarities in the clinical presentation may lead to wrong diagnosis of the melanocytoma as meningioma, schwannoma. We don’t have the pathology specimen of the prior operation so there is the possibility of its being mistaken as meningioma at that time. If we assume that the pathology result was correct, then it can be speculated that the meningeal melanocytoma transformed into melanocytoma which is a very rare condition [14]. Malignant transformation, described in the literature for some extramedullary cases, was not reported in any of the recurrent intramedullary melanocytomas, even when there was metastatic spread via CSF [8,10,14-17].

Both the prognosis and decision for the exact way of treatment of meningeal melanocytomas are difficult. First of all, the biological behavior of meningeal melanocytomas is variable [9,14]. Secondly, although gross-total removal of the tumor often results in favorable postoperative survival times, recurrence, invasion, or metastasis have been reported in such cases [6]. We resected the tumor gross totally without a plan for postoperative radiotherapy or any other treatment modality. Finally, prognosis of a meningeal melanocytoma is far more benign than a malignant melanoma of leptomeninges which must be considered in its differential diagnosis.

As a conclusion, melanocytoma should be considered in the differential diagnosis of spinal tumors and when they are diagnosed maximum effort should be spent at gross total resection. It must also be beared in mind that the lesion may recur and progress into a malignant melanoma so frequent follow-up of these patients is strongly advised.

References