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Case Report

Primary scattered multifocal melanocytomas in spinal canal mimicking neurofibromatosis

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Abstract

BACKGROUND CONTEXT: Meningeal melanocytoma is an extremely rare pigmented tumor derived from leptomeningeal melanocytes. By and large, it is considered to be a well-differentiated and slow-growing benign lesion. Generally, meningeal melanocytomas are solitary lesions, and the occurrence of the primary multifocal form in the central nervous system is exceedingly rare; it has been previously reported in only six cases.

PURPOSE: The present report illustrates a 41-year-old woman with primary multifocal meningeal melanocytoma in the spinal canal. Contrary to earlier reports, the tumors presented with a scattered appearance mimicking neurofibromatosis.

STUDY DESIGN: This study is a case report and review of literature.

METHODS: On admission, the cerebral magnetic resonance images of the patient were normal, whereas the spinal magnetic resonance images showed scattered multifocal nodules mimicking neurofibromatosis. Surgical resection of the responsible lesions was scheduled. In addition to this case presentation, relevant previous reports were reviewed, and the challenging diagnosis, management, and prognosis of meningeal melanocytoma are discussed.

RESULTS: Gross total resection of the two largest lesions was achieved, and histopathological examinations confirmed the diagnosis. Despite the benign histopathological findings, the patient had an aggressive clinical course. On follow-up at 18 months after surgery, she succumbed to the disease.

CONCLUSION: Clinicians should be alert to a potential aggressive clinical course of meningeal melanocytoma, despite its benign histopathological nature. Of particular note is multifocality and diffuse leptomeningeal hyperpigmentation, which may suggest a poor prognosis. A combined treatment including surgical resection and adjuvant radiotherapy should be considered, and long-term close follow-up is necessary.

Keywords: Magnetic resonance imaging; Melanocytoma; Meningeal melanocytoma; Multifocality; Neurofibromatosis; Prognosis; Spinal tumor; Surgery

Introduction

Meningeal melanocytoma is an extremely rare pigmented tumor of the central nervous system (CNS) derived from leptomeningeal melanocytes, with the majority located in the posterior fossa and spinal canal [1–3]. Meningeal melanocytoma is generally considered as a well-differentiated and slow-growing benign tumor, corresponding histologically to World Health Organization (WHO) grade I [3].

FDA device/drug status: Not applicable.

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However, malignant progression with infiltrative growth or leptomeningeal spread secondary to malignant transformation has also been reported in previous studies [4–7].

Generally, meningeal melanocytomas are solitary lesions, and the occurrence of the primary multifocal form in the CNS is exceedingly rare; only six previous cases have been reported [8–13]. Ali et al proposed that primary multifocal meningeal melanocytoma should be considered as a distinct pathological entity, and multifocality may portend an aggressive clinical course and a poor prognosis [8]. Herein, we report another case with primary multifocal meningeal melanocytoma in the spinal canal. Contrary to earlier reports, the tumors presented with a scattered appearance mimicking neurofibromatosis. The relevant literature has been reviewed, and the challenging diagnosis, management, and prognosis are discussed.

Case report

A 41-year-old woman presented with a 7-month history of pain in the left lumbosacral region and radiating pain to the left lower extremity. Over the month prior to admission, the patient indicated that the pain had increased impairing her gait, and it was exacerbated by coughing or sneezing. She also complained of a new onset of numbness in the anterior surface of the left thigh in the final 10 days prior to admission. Physical examination revealed a loss of sensation in the distribution of the L2 and L3 dermatomes. No motor deficit, sphincter dysfunction, or cutaneous abnormality was noted. Vision and hearing were both normal. There was no remarkable finding in the patient's past medical history, and she denied a family history of CNS tumors. She had no history of hypertension, diabetes mellitus, cigarette smoking, polyarthritis, stroke, or cardiac disease. The baseline laboratory data were normal. Cerebral magnetic resonance imaging (MRI) was requested and revealed no abnormality. Spinal MRI demonstrated scattered multifocal nodules in the thoracolumbar segments (Fig. 1). The T1-weighted images showed homogeneous hyperintensity, and the T2-weighted images showed homogeneous hypointensity. After the administration of gadolinium-diethylene triamine pentaacetic acid (Gd-DTPA), the tumors were homogeneously enhanced; nevertheless, the contrast enhancement of the nodules was evidently not revealed by visual assessment because of strong T1 hyperintensity on unenhanced images. Additionally, diffuse leptomeningeal enhancement was noted. The largest mass at the T12–L1 vertebral level was extradural, and the other masses appeared to be intradural extramedullary. A suspected diagnosis of neurofibromatosis was made.

A surgical strategy including laminotomy at the T11–L1 vertebral level and tumorectomy of the two largest masses via a posterior midline approach was adopted. An epidural tumor was encountered at the T12–L1 level, and the other tumor was found subdurally at the T11–T12 level after dural incision. The tumors as well as the local dura mater were black pigmented. The tumor had a tight dural attachment, and its vascularity was derived from dural vessels. The two largest responsible masses were grossly removed as total excisions. The pathological and immunohistochemical examinations confirmed a diagnosis of multifocal melanocytoma (Fig. 2). Microscopically, the diffusely distributed black melanin-pigmented deposits obscured the observation of cell morphology and microstructure, necessitating a bleaching procedure. The tumor was highly cellular and composed of monomorphic spindle or epithelioid cells arranged in a fascicled or nested growth pattern, with variable amounts of melanin pigment in the cytoplasm. Tumor cells had prominent oval-shaped nucleoli without pleomorphism. Mitotic activity and necrosis were absent. Immunohistochemical examinations supported the diagnosis, with positive staining for antimelanoma monoclonal antibody (HMB-45), vimentin, and S-100 protein, but negative staining for glial fibrillary acidic protein, epithelial membrane antigen, and synaptophysin. The proportion of Ki-67 positive cells was approximately 4%.

Given the WHO classification as grade I and the reported indolent biological activity of meningeal melanocytomas, no adjuvant radiotherapy or chemotherapy was recommended. Immediately after surgery, the patient's radicular pain significantly improved. The numbness and anesthesia in the anterior surface of the left thigh were relieved 1 month after the operation. Unfortunately, at follow-up 18 months after surgery, the patient presented with cough and dysphagia and lapsed into a coma within 1 week. Brain MRI at the local institution revealed multiple nodules with hyperintensity on T1-weighted images and hypointensity on T2-weighted images in the posterior fossa, which were presumed to represent intracranial metastasis. Given the disease progression and the poor Karnofsky score (20%), no further intervention was considered and the patient succumbed to the disease 2 weeks later.

Discussion

Meningeal melanocytoma is an extremely rare tumor arising from leptomeningeal melanocytes, and it consist of approximately 0.06–0.1% of all CNS tumors [14]. This entity can be found in any spinal region including intramedullary, intradural extramedullary, and extradural compartments. Meningeal melanocytoma is the histologically benign variant of a continuous melanocytic tumor spectrum in which primary malignant melanoma represents its malignant counterpart, their aggressiveness being entirely different. As distinct from malignant melanomas, meningeal melanocytomas are generally considered as a benign tumor with a non-aggressive clinical progression and non-proliferative histological characteristics. Meningeal melanocytoma corresponds to grade I in WHO 2007 classification of CNS tumors [15]. Given that the malignant progression has been described in a few isolated case reports, it is crucial for the clinicians to be aware of the potential aggressive behavior of meningeal melanocytomas.