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Case Report: Primary

Leptomeningeal Melanoma in a Patient with Neurocutaneous Melanosis

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Primary Leptomeningeal Melanoma in a Patient with Neurocutaneous Melanosis

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A 30-year-old woman was admitted to our hospital because of a generalised tonic-clonic seizure. Her past medical history was unremarkable except for a biopsy-confirmed Tierfell naevus located on the posterior side of the pelvic region. After the seizure she complained of headache and nausea.

On clinical neurological examination the patient showed no abnormalities except for non-fluent aphasia. In order to exclude underlying structural abnormalities a magnetic resonance imaging (MRI) scan was performed which showed a contrast-enhancing lesion located on the left frontal lobe.

■ What Is Your Diagnosis?

Diagnosis: primary leptomeningeal melanoma in a patient with neurocutaneous melanosis (Figures 1, 2).

PET-CT scan and dermatological examination showed no melanoma outside the central nervous system. Because of the mass effect from the tumour and the neurological abnormalities surgery was performed and the tumour was removed 8 days after admission. Surgical appearance of the leptomeninges displayed several large, dark brown lesions. A postoperative MRI scan showed complete removal of the tumour without contrast enhancement. Histopathological examination of the resected tissue revealed a circumscribed malignant melanocytic lesion with an increased number of non-atypical melanocytes in the surrounding leptomeninges.

Primary melanocytic neoplasms of the central nervous system are rare and arise from leptomeningeal melanocytes. They include benign circumscribed or diffuse tumours (melanocytoma, melanocytosis) and their malignant counterparts (melanoma, melanomatosis) [1]. The diagnosis is mainly based on histopathological findings. The incidence for melanocytoma is estimated to be 1 case per 10 million; for primary CNS melanoma 0.5 cases per 10 million [2].

A preoperative diagnosis of melanocytic neoplasms is generally difficult to establish. On MR imaging, most tumours have a low T2-weighted signal and a high T1 signal with contrast enhancement and FLAIR but tumours with higher percentages of melanin-containing cells might show hyperintensity on T1- and hypointensity on T2-weighted images [3]. Macroscopically, the tumour usually appears as a brown-to-black lesion that is firmly attached to the underlying meninges. Microscopically, a variable amount of melanin pigment is seen in the tumour cells at various stages of development [2].

Although meningeal melanocytoma is a benign condition, relapse and malignant transition into melanoma have been reported. Wang et al reported a case of a primary meningeal melanocytoma located at the temporal lobe in which malignant transformation was confirmed histopathologically 3 years after resection of the tumour [4]. For that reason, adjuvant radiation therapy is advised in both complete and incomplete resection. The 5-year survival rate for patients

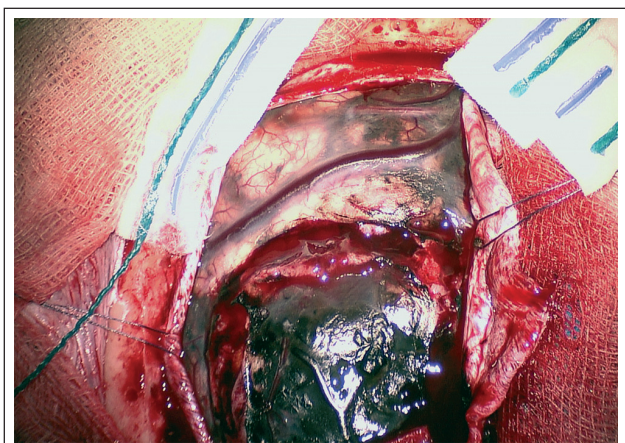


Figure 1. Intraoperative photograph after opening of the dura, showing a black leptomeningeal tumour in the left frontal region. Of note is the spotty black appearance of the arachnoid, which is not continuous with the tumour, consistent with neurocutaneous melanosis.

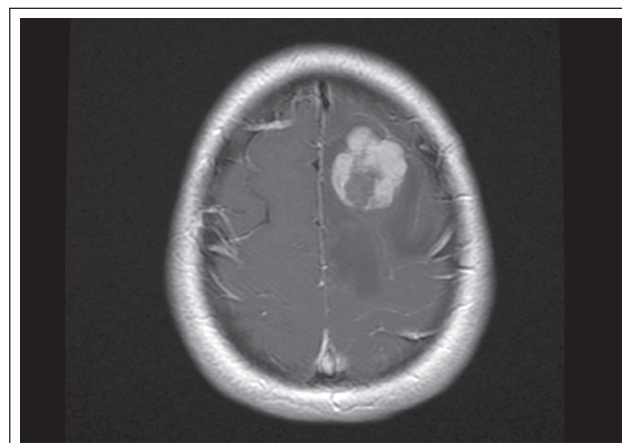


Figure 2. Transversal section of contrast-enhanced T1 MRI demonstrating a left frontal, inhomogeneously contrast-enhancing, and space-occupying lesion with oedema in the surrounding brain tissue.

with incomplete resection in combination with radiation therapy was 100 %, but only 46 % without radiation therapy [5].

Primary CNS melanoma is an aggressive tumour and may metastasise throughout the neuraxis and sometimes even to other organs [2]. Patients with complete resection have a better outcome, post-operative radiation therapy is advised in all cases [3]. Primary malignant melanoma of the CNS may be found in isolation or (as in our patient) in the context of neurocutaneous melanosis.

Our patient underwent postoperative radiation therapy. Three months after surgery the patient was free of symptoms and an MRI scan showed no recurrence.

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