Rarity of a melanocytic low-grade malignant peripheral nerve sheath tumour with ganglioneuromatous component causing brain metastases as primary manifestation in a 60-year-old patient [Abstract]

K. Mueller, J. Tilgner, Ansgar Berlis, P. Fisch, B. Volk

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Rarity of a melanocytic low-grade malignant peripheral nerve sheath tumour with ganglioneuromatous component causing brain metastases as primary manifestation in a 60-year-old patient

K. Mueller (1), J. Tilgner (2), A. Berlis (3), P. Fisch (4), B. Volk (1)

(1) Pathologisches Institut, Abteilung Neuropathologie (Freiburg); (2) Pathologisches Institut, Abeilung Stereotaktische Neurochirurgie (Freiburg); (3) Pathologisches Institut, Abteilung Neuroradiologie (Freiburg); (4) Pathologisches Institut, Abteilung Molekularpathologie (Freiburg)

Objective: The presented case report puts emphasize on a clear-cut diagnostic procedure in cases of brain metastases of unknown origin.

The extremely rare case of a low grade melanocytic malignant peripheral nervous sheath tumor (MPNST) causing brain metastases is demonstrated.
Methods: We show the case of a healthy 60-year-old man who attracted clinical attention by a seizure. Magnetic resonance imaging (MRI) revealed two contrast enhancing and dura based masses frontal und parietal, suspicious for carcinoma metastases. A stereotactic biopsy was done to verify the diagnosis. Extensive pathomorphological and also molecular genetic analyses were done.
Results and clinical course: The histomorphological investigations showed an unusual pigmented neuroectodermal tumor which was classified as melanocytic MPNST with a ganglioneuromatous component. In addition to conventional histology and immunohistochemistry electron microscopy and molecular genetic analysis were necessary to find out the correct diagnosis. After neurosurgical resection of one of the lesions the patient got whole brain radiotherapy. In further MRI and positron emission tomography (PET) the presumable primary could be detected 6 months after resection at the right sciatic nerve. Familiar tumor syndromes such as neurofibromatosis could be ruled out. Clinical controls showed also further metastatic manifestations at intraspinal and cervical lymphatic localizations. Two years after the first symptoms the patient is at stable disease with Karnofsky score 90.
Conclusions: In case of/given unexpected results in investigating brain metastases a concise diagnostic procedure of tissue handling is necessary to avoid expensive and futile clinical investigations in search of the primary. The absolutely rare metastatic spread of a low grade MPNST to the brain as shown here before the primary is detected underlines the importance of a precise diagnostic and katamnestic workup.