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Neuroimaging Highlight

Intraventricular Primary Diffuse Meningeal Melanomatosis

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A 76-year-old man presented to our quaternary care neurosurgical center with a 3-month history of confusion, memory deficits and cognitive decline. He had no history of prior malignancy and his family history was unremarkable. Physical examination was notable for disorientation to time and place, without any focal neurological deficits. Detailed dermatologic and ophthalmologic examinations were unremarkable without any evidence of melanoma deposits.

Computed tomography (CT) imaging of the brain revealed an enlarged and avidly enhancing pineal lesion and obstructive hydrocephalus (Figure 1). Diffuse areas of leptomeningeal enhancement were seen, notably in the left cingulate sulcus, left temporal lobe and the roof of the fourth ventricle. We were unable to obtain magnetic resonance imaging due to concerns of incompatibility with a prior pacemaker insertion. Differential diagnosis based on imaging included pineal germ cell tumor, specifically germinoma given the central calcifications, pineal parenchymal tumor and cerebral metastasis.

Given his obstructive hydrocephalus, he was taken urgently to the operating theater for endoscopic third ventriculostomy and endoscopic biopsy. Intraoperatively, there was diffuse intraventricular dissemination of melanin deposits in the lateral ventricle and third ventricle (Figure 2). The pineal region was similarly stained with black melanin deposits and was biopsied. Pathology confirmed a melanocytic neoplasm. Molecular testing revealed MYC and FGR1 gain. The Ki-67 proliferative index was low. Testing for BRAF, PRAME, GNAQ, GNA11, KIT, NRAS and KRAS was negative. A diagnosis of primary diffuse meningeal melanomatosis was made. Postoperative CT demonstrated improved hydrocephalus.

The case was discussed at neuro-oncology conference, and systemic immunotherapy was recommended. However, the patient declined further treatment and opted for a palliative approach.

Primary diffuse meningeal melanomatosis is a rare neoplasm originating from leptomeningeal melanocytes with dissemination throughout the subarachnoid space.[1] It is considered a malignant neoplasm with a poor prognosis; however, published data are limited to case reports and small case series.[2] Substantial heterogeneity in immunohistochemical features, treatment approaches and clinical outcomes has been reported.[3] Furthermore, the prognostic significance and treatment implications of various molecular profiles remain incompletely characterized.[4]

This case illustrates the diagnostic and management challenges of intraventricular primary diffuse meningeal melanomatosis. These tumors can present with obstructive hydrocephalus often requiring urgent cerebrospinal fluid (CSF) diversion. Endoscopic third ventriculoscopy (ETV) allows for simultaneous CSF diversion and tissue diagnosis of pineal lesion tumors. However, if a patient's anatomy is not suitable for ETV, a staged approach with initial surgery for CSF diversion, either with external ventricular drain or a ventriculo-peritoneal shunt, followed by either stereotactic or open biopsy for tissue diagnosis may be required. To our knowledge, our intraoperative images are the first publication to demonstrate the in vivo appearance of intraventricular melanomatosis.

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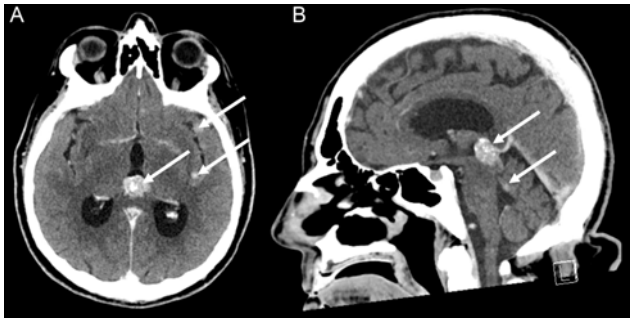

Figure 1. Preoperative computed tomography (CT) demonstrating enhancing lesions within the pineal region, roof of the fourth ventricle and left temporal lobe leptomeninges. Axial (A) and sagittal (B) views.

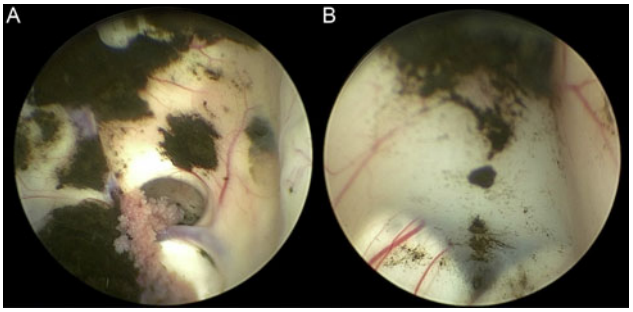

Figure 2. Intraoperative endoscopy demonstrating diffuse black melanocytic lesions within the lateral ventricle (A) and third ventricle (B).

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