Primary melanocytic tumor

Primary melanocytic tumors of central nervous system represent rare tumors arising from melanocytes of the leptomeninges. These neoplasms include focal forms like melanocytoma and primary malignant melanoma and diffuse forms like leptomeningeal melanocytosis and primary leptomeningeal melanomatosis. The clinical diagnosis remains challenging, with clinical and radiologic features overlapping with other more common diseases [1].

Demographic, clinical and surgico-pathologic findings of five patients with melanocytic tumors seen between 1996 and 2003 were studied. In this study, five cases of primary melanocytic tumors have been reported: four cases of malignant melanoma and one case of melanocytoma. Three of the 5 cases were intracranial and 2 were spinal. The mean age in the present study was 26 years. Presenting features varied according to the location. Primary melanocytic tumor of CNS are rare. Whenever possible, complete surgical excision is the best treatment [2] —-

Primary melanocytic tumors of the central nervous system (CNS) are rare lesions, but primary sellar tumors are rarer. Only 10 cases have been reported, and they are often misdiagnosed as pituitary macroadenoma.

Zhan et al. report the case of a 54-year-old Chinese man who developed progressive bitemporal hemianopsia and visual loss. Magnetic resonance imaging (MRI) revealed an intrasellar and suprasellar clouded lesion adhering to the optic chiasm, hypothalamus, and pituitary stalk that was suspected of being a hemorrhagic pituitary macroadenoma. Because of the atypically giant, hemorrhagic, and upward-growing lesion, an initial transsphenoidal approach failed, and subsequent transfrontal craniotomy was adopted to achieve macroscopically complete resection. Histopathologic findings revealed a benign melanocytic tumor. Despite an extensive search, no other primary or secondary site was found. Considering the relatively benign lesion, effective surgery, and potential significant consequences of radiotherapy, the patient received no further treatment and is still alive at the 7-year follow-up. Primary sellar melanocytic tumors are exceptional lesions that are difficult to diagnose before operating and/or obtaining pathological findings. The pathological classification and extent of surgical resection may play a key role in the prognosis. Once this type of lesion is suspected, the transfrontal approach may achieve preferable exposure and resection. Complete surgical resection may be sufficient for relatively benign lesions; otherwise, stereotactic fractionated radiotherapy is indicated. More cases should be reported to improve the treatment strategy [3].

1)
Girolami I, Cima L, Ghimenton C, Zannoni M, Mombello A, Riva G, Cirielli V, Corradi G, Vogrig A, Di Stefano G, Novelli L, Gessi M, Eccher A. NRASQ61K mutated diffuse leptomeningeal melanomatosis in an adult patient with a brief review of the so-called “forme fruste” of neurocutaneous melanosis. Brain Tumor Pathol. 2018 Oct;35(4):217-223. doi: 10.1007/s10014-018-0328-x. Epub 2018 Aug 25. PMID: 30145692.
2)
Jaiswal S, Vij M, Tungria A, Jaiswal AK, Srivastava AK, Behari S. Primary melanocytic tumors of the central nervous system: a neuroradiological and clinicopathological study of five cases and brief review of literature. Neurol India. 2011 May-Jun;59(3):413-9. doi: 10.4103/0028-3886.82758. PMID: 21743173.
3)
Zhan RY, Zhou HJ, Ma YH, Cao F, Zheng XJ. Primary sellar melanocytic tumor mimicking hemorrhagic

pituitary macroadenoma: Case report and literature review. Br J Neurosurg. 2015 Jan 23:1-5. [Epub ahead of print] PubMed PMID: 25613300.