

# Skull Base Enchondroma

**Maffucci syndrome** (MS) and **Ollier disease** (OD) are rare diseases characterized by multiple benign **enchondromas**. Incidence of skull base (SB) enchondromas and the risk of malignant transformation remain unknown. Most SB lesions are asymptomatic and surgical resection carries significant morbidity. Observation may be a reasonable strategy. We report our experience with observation of probable SB enchondromas in MS/OD patients.

Retrospective review of OD/MS cases with cranial imaging between 1995-2018 at one institution.

14 patients were included-3 MS (21.4%) and 11 OD (78.6%). Median age was 28 years (range 11-74) and 57.1% were female. Extracranial chondrosarcoma was reported in 3 (21.4%) patients. Seven (50%) patients with SB enchondroma or chondrosarcoma were identified on initial imaging. In patients with SB lesions, the indications for imaging were: headache (n=3), seizure (n=1), and diplopia (n=1); 2 cases were incidental findings. Most commonly involved structures: petroclival fissure (86%) and clivus (71%). Treatment included observation (6/7) and resection (1/7). Follow-up imaging was available for all SB lesions with a mean interval of 50.7 months (range 5-225) and was negative for progression in all cases.

Primary SB lesions in OD/MS patients frequently present in the petroclival junction. Cranial screening and close observation should be considered in MS/OD patients given increased risk of intra-axial intracranial tumors, de novo chondrosarcomas or malignant degeneration of previously known lesions. In asymptomatic patients, observation appears to be a safe strategy in this cohort. Further case accumulation and follow-up is required to better understand the long-term outcomes <sup>1)</sup>.

## Case reports

Enchondroma of the Skull Base in a Case of Ollier's Syndrome <sup>2)</sup>.

<sup>1)</sup>

Oushy S, Peris-Celda M, Van Gompel JJ. Skull Base Enchondroma and Chondrosarcoma in Ollier Disease and Maffucci Syndrome. *World Neurosurg.* 2019 Jun 21. pii: S1878-8750(19)31639-0. doi: 10.1016/j.wneu.2019.06.087. [Epub ahead of print] PubMed PMID: 31233929.

<sup>2)</sup>

Sabeti S, Yousefi F, Pourabdollah Toutkaboni M. Enchondroma of the Skull Base in a Case of Ollier's Syndrome. *Iran J Pathol.* 2015 Summer;10(3):237-42. PubMed PMID: 26351491; PubMed Central PMCID: PMC4539773.

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