

Histiocytic sarcoma

Histiocytic [sarcoma](#) (HS) is a rare malignant [neoplasm](#) of macrophage-dendritic cell lineage that can occur at any site. The primary base of skull involvement is exceedingly rare.

Perez et al. presented the case of a previously healthy 56-year-old man who complained of headaches and showed localized neurologic symptoms. Magnetic resonance imaging demonstrated a hyperintense and enhancing mass involving the sphenoid bone and the clivus with an extradural component that compressed the distal pons. The differential diagnosis included chordoma or chondrosarcoma. Endoscopic trans-sphenoidal resection was performed. Microscopically, the tumor showed epithelioid and spindle morphology with atypia, mitoses, and necrosis. No osteoid, cartilaginous, or myxoid matrix was identified. By immunohistochemistry, the tumor was positive for CD68 (KP-1) and lysozyme, variably positive for CD4, CD11c, CD14, CD68 (PGM-1), CD45, and CD163, and negative for markers of epithelial, melanocytic, lymphoid, myeloid, muscle, and dendritic cell origin. Expression of PD-L1 by immunohistochemistry and BRAF V600E mutation analysis by PCR were negative. Tumor recurrence developed after radiation treatment with overwhelming progression into a largely infiltrating mass within 2 weeks with clinical deterioration, and the patient died 3 months later. To our knowledge, this represents the first case of primary HS of the clivus reported to date in the English literature, further expanding the spectrum of neoplasms seen at this site as well as the sites where HS can be seen. The overall prognosis of HS in the skull base is poor, with no standard treatment. Further research is warranted to develop effective treatment approaches, which in the future may rely on the expression of checkpoint inhibitors and/or specific molecular markers ¹⁾.

Chen CJ, Williams EA, McAneney TE, Williams BJ, Mandell JW, Shaffrey ME. Histiocytic sarcoma of the cavernous sinus: case report and literature review. *Brain Tumor Pathol.* 2015 Jan;32(1):66-71. doi: 10.1007/s10014-014-0191-3. Epub 2014 May 8. Review. PubMed PMID: 24807104.

¹⁾

Perez I, Gokden M, Day JD, Yaziji H, Pina-Oviedo S. Primary histiocytic sarcoma of the clivus with focal extension into central nervous system and neurologic manifestations: First description at an unusual site with an overwhelming and rapid progression. *Clin Neuropathol.* 2022 Mar-Apr;41(2):74-82. doi: 10.5414/NP301400. PMID: 34958302.

From:

<https://neurosurgerywiki.com/wiki/> - **Neurosurgery Wiki**

Permanent link:

https://neurosurgerywiki.com/wiki/doku.php?id=histiocytic_sarcoma

Last update: **2024/06/07 02:52**

