# Epithelioid hemangioendothelioma

Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor that frequently occurs in soft tissues. Patients suffer from local recurrence and remote metastases because of its malignant potential.

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Covelli C, Parente P, Icolaro N, Dimitri LMC, Vigna B, Popolizio T, Graziano P. Primary Epithelioid Hemangioma of the Central Nervous System: A Case Report and Review of the Literature. J Neuropathol Exp Neurol. 2021 Jan 7:nlaa163. doi: 10.1093/jnen/nlaa163. Epub ahead of print. PMID: 33411905 [1].

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A 57-year-old male patient was admitted for several months of back pain. A computed tomography (CT) scan and magnetic resonance imaging (MRI) were suggestive of T12 hemangioma without the involvement of the spinal canal or posterior elements. Despite aggressive conservative treatments, such as medications or nerve blocks, the back pain worsened. The CT and MRI 2 months later revealed a lesion involving the vertebral body and posterior elements with extension into the epidural space and with spinal cord compression. The patient underwent surgery for bone cement-augmented percutaneous screw fixation followed by low-dose radiotherapy. Histological examination confirmed the diagnosis of atypical hemangioma, specifically an epithelioid hemangioendothelioma [2].

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A 58-year-old female presented to the hospital with respiratory distress several days after a right hallux amputation. A new lytic lesion within the fourth thoracic (T4) vertebral body and mediastinal lymphadenopathy was noted on chest computed tomography scan. A bone biopsy was performed, revealing bone and collagenous fragments only. Two months later, new imaging revealed approximately 60% lytic destruction of the T4 vertebral body with new right pedicle involvement. Surgical treatment was offered. Intraoperative frozen pathology indicated a hemangioma. An intralesional debulking and stabilization was performed. The right T4 nerve was sacrificed to gain access to the entire vertebral body. Curettage was then used to push the tumor away from the spinal canal into the vertebral body. The spine was reconstructed with 5-10mm beads of Simplex P bone cement (Stryker®, Kalamazoo, MI) which contained 40 grams of poly-methyl methacrylate and 1 gram of tobramycin. Five months after resection, the patient presented with computed tomography and magnetic resonance imaging findings of recurrent disease at T4 and spread to the adjacent T5 vertebral body with lytic changes. At 18 months following her second debulking surgery and radiation treatment, the patient was doing well with no pain or numbness. Long-term imaging compared to the patient's preoperative imaging displayed improvement in spinal debulking with minimal residual enhancement of tumor despite significant artifact [3].

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Ogita et al present a rare case of EHE that originated from nasal cavity and invaded intracranially through the anterior skull base.

A 27-year-old woman who presented a local physician with intermittent epistaxis and a facial pain

around her nose. Preoperative studies demonstrated that the tumor invaded into anterior skull base and the dura matter. Therefore, they performed combined skull base and transnasal surgery, which achieved complete resection of the tumor. Postoperative course of the patient was uneventful. No recurrence or distant metastases was observed in the patient for 2 years following the radical resection.

To date, four cases of EHE in the nasal cavity were reported. This is the first case in which EHE demonstrated invasive potentials with intracranial extension. Radical surgical resection plays an important role for better management of invasive paranasal EHE [4].

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Covelli C, Parente P, Icolaro N, Dimitri LMC, Vigna B, Popolizio T, Graziano P. Primary Epithelioid Hemangioma of the Central Nervous System: A Case Report and Review of the Literature. J Neuropathol Exp Neurol. 2021 Jan 7:nlaa163. doi: 10.1093/jnen/nlaa163. Epub ahead of print. PMID: 33411905.
2)
Kim CH, Kim SW. Rapidly Progressive Atypical Vertebral Hemangioma: A Case Report. Korean J Neurotrauma. 2020 Aug 20;16(2):320-325. doi: 10.13004/kjnt.2020.16.e24. PMID: 33163444; PMCID: PMC7607043.
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