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# Lymphangıomatosıs of the Skull Base

**Iplikçioğlu AC[1] and Karabağ H[2]***

[1]*Department of Neurosurgery, BHT Clinic Istanbul TEMA HASTANESİ Atakent Mahallesi, Turkey*

[2]*Department of Neurosurgery, Harran Üniversity faculty of medicine, Turkey*

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***Corresponding author:** Hamza Karabag Department of Neurosurgery Harran Üniversity, Şanlıurfa, Turkey Malakhov, Grodno State Medical University, Turkey

**Abstract**

Skull base lymphangioma is a rare disease and can cause pain, cranial nerve deficitis and CSF rhinorrhea. Neuroradiological examinations demonstrate lytic lesions in skull base. In most of the cases, complete resection of the lesion is difficult because of its multifocal nature of the disase. Radiotherapy and chemotherapy using agents as such as biphosphanates, interferon alpha and bevacizumab are other treatment options.

## Introduction

Lymphangiomas are uncommon benign congenital malformations characterized by infiltration and proliferation of lymphatic canals within the visceral organs and tissues [1]. They typically occur in the skin and subcutaneous tissues of the head and neck. These locations account for 70% of lymphangiomas. Lymphangioma of bone is rare disease and can be associated with massive bone loss called Gorham-Stout disease (vanishing bone disease) [2]. Skull involvement is extremely rare and presents as multiple or solid lesions [1,3]. In this report, we present a case of skull base lymphangioma.

## Case Report

A 39-year-old man was admitted to our clinic with a three month history of right ear ache and headache radiating from the right mastoid region to the right occipital region. At admission, the patient's neurological examination was unremarkable. A cranial computed tomography scan of the patient revealed a lytic clival lesion that extended from the right mastoid region to the right posterior skull base; the lesion was expanding to the right inferior occipital bone and the mastoid bone (Figure 1). The lesions were hypointense on T1-weighted cranial magnetic resonance imaging with heterogeneous contrast enhancement (Figure 2). Whole body bone scintigraphy revealed increased osteoblastic activity in the right occipital region. The patient underwent surgery via an endoscopic transnasal transsphenoidal approach. The lesion was excised partially due to the widespread nature of the tumour. The histopathological diagnosis was lymphangioma (Figure 3). The patient was closely monitored during follow-up, and no progression of the tumor was observed via control cranial magnetic resonance imaging and CT obtained one year after the diagnosis.

## Discussion

Lymphangioma of the bone usually occur in tibia, humerus, mandible, cranium and ileum [4]. Lymphangioma of the skull can be solitary or multiple. Kaya et al reported a case of skull base lymphangioma localized in the clivus [5]. Skull base lymphangiomas are usually multifocal and associated with dissemination of primary neck and vertebral lesions [6]. However, in our case no extraskeletal lesion was detected.

Lymphangioma of the bone usually become symptomatic with pain, pathological fracture and deformity due to the erosion of the bone [6]. Skull base lympangiomas also presents with pain, cranial nerve palsy and cerebro spinal fluid leakage [1,7]. In the case reported by Canady and Chou, neurological deficits developed due to acquired basilary impression and Chiari-I malformation [4]. Computed tomography showing the lytic lesions of skull base and magnetic resonance imaging showing the extension of the lesion and its relationship with crucial structures of the brain are the best diagnostic tools. Lytic lesions such as chordoma, chondroid chordoma, chondrosarcoma plasmocytoma, fibrous dysplasia, giant cell tumors, Paget's disease, eosinophilic granuloma and nasopharyngeal carcinoma should be considered in differential diagnosis [1].

Treatment regime depends on the localization, extension of the lesion and the symptoms. Complete removel of the solid lesions

is the best treatment. However, it is difficult to achieve a radical excision of multiple skull base lesions. Radiotherapy is usually used to control the progression of the lesion [8,9]. Medical therapy with agents such as bisphosphonates, vitamin D, interferon alpha 2b, bevacizumab is another option. Patients with CSF Leakage, should be treated for the repair of CSF fistula. Recently Wright et al. treated a patient with multifocal skull base lymphangioma by transoral clivoplasty resulting in complete resolution of pain [9].

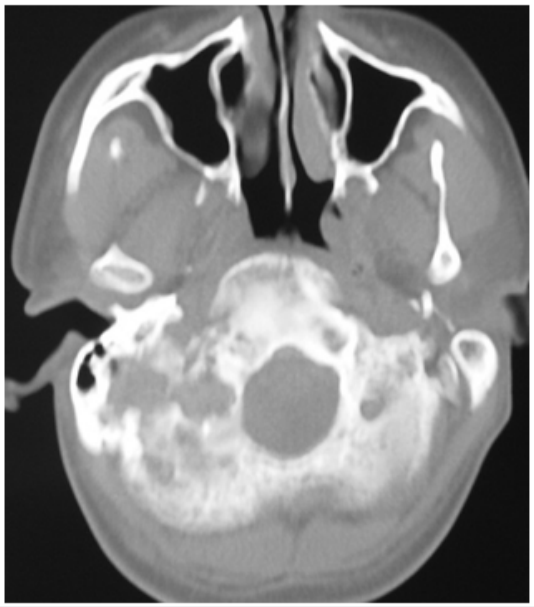

**Figure 1.** Axial CT scan showing osteolytic lesions of clivus and occipital bones.

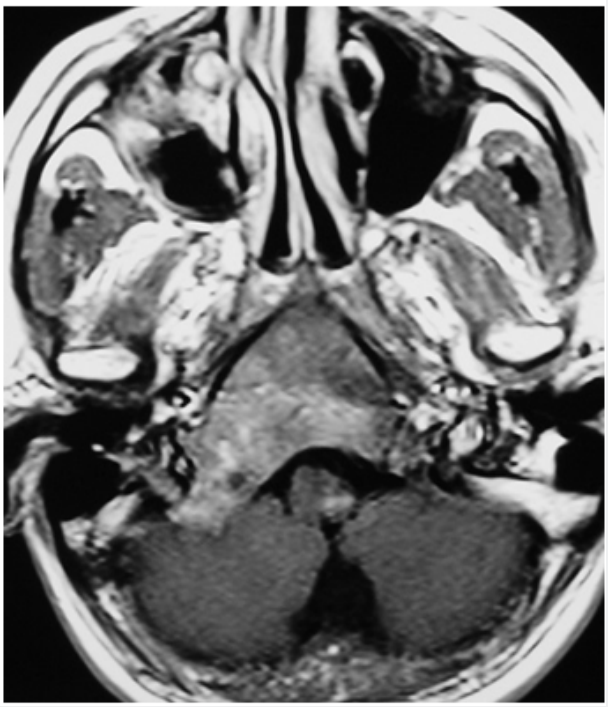

**Figure 2.** Axial T2 Weighted MRI after contrast administration demonstrating expanded clivus associated with intraosseous lesion with heterogeneous contrast enhancement.



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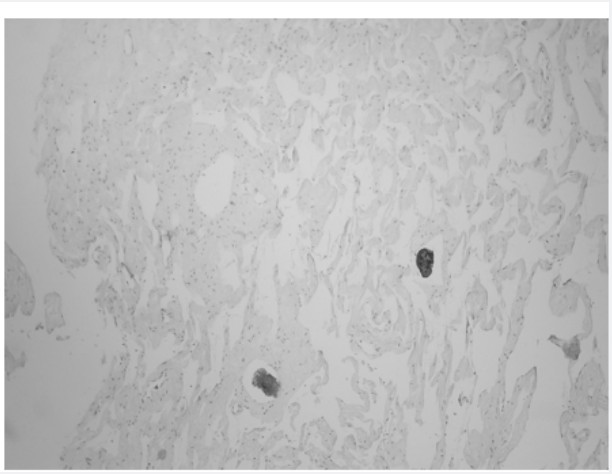

**Figure 3.** Histopathological examination shows multiply lymphatic channels in loose connective tissue. Flattened endothelial cells on the walls of the channels and infiltration of lymphocytes are also seen. X 100

## References


1. Ito E, Saito K, Nagatani T, Teranishi M, Kamei Y, et al. (2008) Lymphangioma of the skull base bones leading to cerebrospinal fluid rhinorrhea. J Neurosurg Pediatr 2(4): 273-6.
2. Takaya K, Sakamoto Y, Miwa T, Yoshida K, Kishi K (2021) Gorham-Stout disease with parietal bone osteolysis: a case series and review of literatüre. British Journal of Neurosurgery 35(1): 27-31.
3. Tibesar RJ, Rimell FL, Michel E (1999) Cystic hygroma of the skull base. Arch Otolaryngol Head Neck Surg 125(12): 1390-1393.
4. Canady AI, Chou SN (1980) Cervical lymphangiomatosis with progressive craniospinal deformity. Neurosurgery 6(4): 422-425.
5. Kaya S, Colak A, Kutlay M, Gungor A (2007) Lymphangioma of the clivus: case report. Neurosurgery 60(4): E779.
6. Sinard RJ, Welling DB (1995) Cervical lymphangioma with simultaneous skull base invasion and soft tissue regression. Ann Otol Rhinol Laryngol 104(8): 662-664.
7. Martinat P, Cotten A, Singer B, Petyt L, Chastanet P, et al. (1995) Solitary cystic lymphangioma. Skeletal Radiology 24(7): 556-558
8. Blei F (2011) Lymphangiomatosis: clinical overview. Lymphat Res Biol 9(4): 185-190.
9. Wright CH, Kusyk D, Rosenberg WS, Sweet JA (2017) Percutaneous transoral clivoplasty and upper cervical vertebroplasties for multifocal skeletal lymphangiomatosis resulting in complete resolution of pain: case report. J Neurosurg Spine 26(2): 171-176.



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