Calvarial cavernous lymphangioma is an extremely rare disease entity with only a few reports. Lymphangiomas are frequently encountered neck and long bone but it may also seen in many part of the body and many tips of the lymphangioma have been identified. Several treatment options have been defined but local recurrence is still a big problem. In the pediatric population, postoperative skull defects may frequently require cranioplasty. Allografts or autologous bone grafts may be used for cranioplasty. A four-month-old male patient was hospitalized due to a painless head mass, which was revealed as a soft tissue lesion located in calvarial diploe or extracranial lesion with outer calcified shell on the cranial computed tomography. The patient underwent surgical resection, pathologically confirmed as cavernous lymphangioma. We report this case with imaging findings and review of literatures.

**Keywords**
Lymphangioma, calvarium, age, cranioplasty

**Anahtar Kelimeler**
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**Address for Correspondence/Yazışma Adresi**:  
Zahir Kızılay MD, Adnan Menderes University Faculty of Medicine, Department of Neurosurgery, Aydın, Turkey  
E-mail: zahirkizilay@hotmail.com  
ORCID ID: orcid.org/0000-0002-2021-0406

Introduction

The pathological identity of lymphangioma is still controversial, though it is now generally expressed as a disease of congenital venous and lymphatic malformations (1). The predilection sites are neck, axilla and mediastinum (2), usually developed in the soft tissue of those regions. Primary bone lymphangioma is very rare. Since the first description by Bickel and Broders in 1947, less than 30 bone
lymphangiomas have been reported (3). Especially, skull lymphangioma is extremely rare. It has been reported in only a few literatures (4), describing as an asymptomatic head lump incidentally detected or lump presenting with pain (5). We report a case of a four-month-old male patient who presented a skull mass confirmed as a cavernous lymphangioma.

Case Report

A four-month-old male patient referred to the hospital due to abnormal bulging of the left parietal area in the head. He had a normal neurological examination. However, skull X-ray series showed mass like lesion with sclerotic rim in the left parietal bone area (Figure 1). Cranial computed tomography (CT) was subsequently performed, revealing mixed density crescent shape lesion developed in diploe, separating inner and outer table of the skull. There were no abnormal findings in the brain (Figure 2). Approval forms were taken from parents of child. After preoperative preparations patient underwent surgical resection to remove the lesion. Thin purple colored calvarial bone covered the lesion under the scalp. After the removal of this thin bony layer, yellowish-white soft tissue mass was exposed. It contained brownish fluid loculation inside (Figure 3). All abnormal lesions including mass and surrounding bones were totally removed. Inner table of the calvarium was clear without involvement of the lesion.

Pathologic examination showed proliferation of the enlarged lymphatics with destructed bony trabeculae in some areas. Immunohistochemical studies revealed positive staining of CD31+ and CD34+. The pathological diagnosis was calvarial cavernous lymphangioma (Figure 4).

Follow-up cranial CT acquired in 6 months, which showed remodeling skull around operation site with complete filling up in postoperative defect. Residual or recurred lesions were not demonstrated. Follow-up CT in 1 year also showed same findings (Figure 5).
Discussion

Lymphangiomas are uncommon congenital tumors, usually discovered in neck and head. The cases in the literature are generally the ones with skeletal expanding (6). They are histologically benign, extruding surrounding normal structures without invasion or destruction when they grow in expanding pattern. However, lethal complications might be caused according to the location and growing behavior (7).

Lymphangiomas are classified into three groups as follows: cystic, cavernous and capillary. Cystic lymphangioma is formed of dilated thin walled spaces surrounded with regularly arranged endothelial cells that are full of eosinophilic proteinaceous fluid (3). Cavernous lymphangioma consists of large lymphatic channels, located inside the bones, soft tissues and organs. Capillary lymphangioma is characterized with including lymphatic channels in the size of capillaries. It is generally located in the skin but some of them have been reported to be located in the bone (4). The pathogenesis is not obvious yet. Infectious, toxic, neoplastic and congenital mechanisms are all thought to be possible (8). As the histopathologic sections of our case included large lymphatic channels, it has been diagnosed as cavernous lymphangioma and any predisposing factors could not be found.

Primary lymphangioma involving bone is rare. Only small number of cases have been reported in previous literatures. According to previous reports, bone lymphangiomas were discovered in the metaphysis and/or diaphysis of tibia, humerus, ilium, skull, mandible, and spine (3,9). Especially among them, involvement of skull is extremely rare, described just in a few literatures. The symptoms in bone lymphangioma is associated with the eroding of the bone by the enlargement of the lymphatics or the pressure of it. Patients may be presented by local pain, enduration or pathologic fractures or it can be diagnosed incidentally in the radiologic imagings that have been performed for another suspicion (2). Radiographs of the skull lymphangioma have been described in a few reports. Plain skull series typically demonstrate expanding change of calvarial bony tables, which might cause loss of bony coverage. CT reveals the low lesion associated with mixed lytic and sclerotic changes of the skull. Internal hemorrhage might be developed, causing changes of imaging nature on CT or magnetic resonance imaging (4,10).

Treatment is aimed for cosmetic or pain control as well as curative removal of the lesion. Total surgical excision, sclerotherapy and laser ablation have been popularly used for the treatment. As lymphangiomas have high tendency of local recurrence, total resection is known as the best treatment choice (1,3,11,12). Sclerotherapy and laser ablation are often used for the treatment of soft tissue lymphangiomas.
In the pediatric age, postoperative skull defects frequently require cranioplasty. Allografts or autologous grafts may be used for cranioplasty, although autologous bone graft should be the first option in pediatric age. The best donor source of autologous bone is referred as a split-thickness cranium. But, it is almost impossible to perform the procedure because of the thin and immature diploe of the skull in children younger than 3 years old. The second harvesting area for bone grafts is referred as an autologous split rib grafts. But, this procedure requires a second wound site and a prolonged operating time. In addition, autologous bone grafts can be absorbed partially. Allograft materials options for cranioplasty are metallic mash plates, methyl methacrylate, hydroxyapatite and porous polyethylene with high density (13-15). In the different serious, complication rate changes between 5% and 30%. Infection is the most important problem due to used allograft materials after total resection of calvarial mass (14).

In our case, we did not perform cranioplasty or the other options. Because his age was not suitable for autologous bone grafting and his inner table of the calvaria was clear. We thought that new bone might develop from his inner table of the calvaria and we saw remodeling on his calvarial bone in over time. We can ask a question ourselves for self-criticism in here, could a local recurrence develop in the surgery area? Of course, we could saw a local recurrence because recurrence is still a big problem.

We experienced an extremely rare case of skull lymphangioma, which was manifested as a soft tissue lesion located in calvarial diploe. It was clinically presented as abnormal bulging of the skull and total surgical resection was achieved without intermediate term evidence of local recurrence. Postoperative follow-up of lymphangiomas is important because of the local recurrence.

Ethics

Informed Consent: It was taken.
Peer-review: Externally peer-reviewed.

Authorship Contributions


Conflict of Interest: No conflict of interest was declared by the authors.

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