Meandros Med Dent J

Case Report / Olgu Sunumu

Congenital Calvarial Lymphangioma: A Case Report

Konjenital Kalvaryal Lenfanjiom: Olgu Sunumu

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Abstract

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
Lenfanjiom, kalvaryum, yaş, kraniyoplasti

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Öz

Kalvaryal kavernöz lenfanjiom çok az sayıda yayında bildirilmiş ve son derece nadir rastlanan bir klinik antitedir. Lenfanjiomlar vücudun birçok yerinde görülse de lenfanjiomlarla sıklıkla boyun ve uzun kemiklerde karşılaşılır ve literatürde lenfanjiomların birçok tipi tanımlanmıştır. Lenfanjiomlar için birçok tedavi seçeneği tanımlansa da lokal nüks hala büyük bir problemdir. Pediatrik popülasyonda kafatası defektleri sıklıkla kraniyoplasti gerektirir. Allogreft veya otolog kemik greftleri kraniyoplasti için bir seçenektir. Dört aylık erkek bir hasta, kraniyal bilgisayarlı tomografide kalvaryal veya ekstrakraniyal dış kabuğu kalsifiye bir lezyon görünümünde, ağrısız bir kitlesel lezyon nedeniyle hospitalize edildi. Hastaya cerrahi rezeksiyon uygulandı ve patoloji ile kavernöz lenfanjiom tanısı doğrulandı. Biz bu olguyu görüntüleme bulguları ve mevcut literatürün değerlendirilmesi ile sunuyoruz.

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.

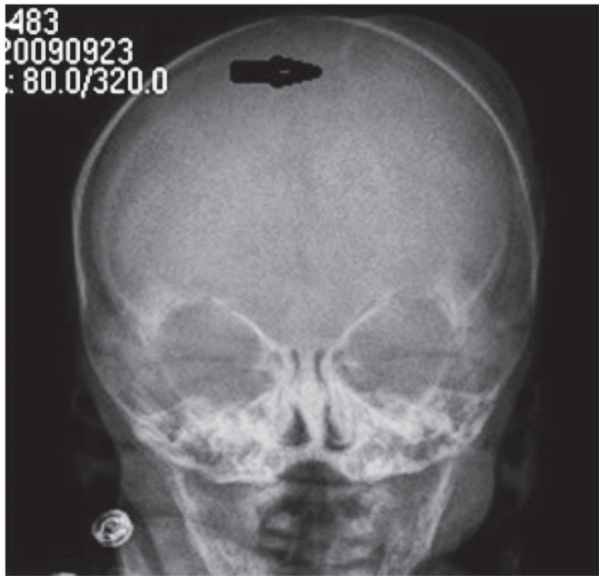

Figure 1. Skull anteroposterior view shows a bulging shadow with sclerotic rim in the left parietal area

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).

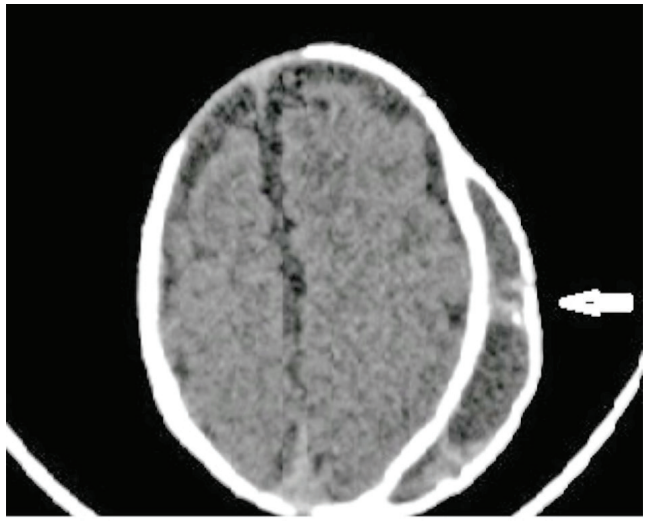

Figure 2. Crescent shape lesion looking like composed of solid and cystic portions is seen in the left parietal area. It seems to be the lesion developed in diploe, separating inner and outer table of the skull or the lesion developed in extracranial area with outer calcified shell

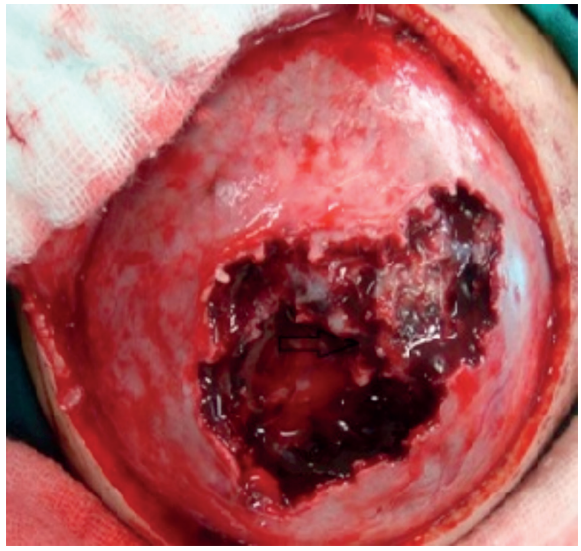

Figure 3. Intraoperative view of the lesion, after removing outer table of the skull

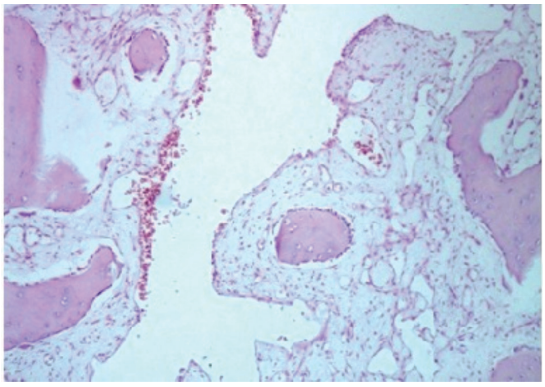

Figure 4. Enlarged lymphatic channels are seen with erythrocytes inside the lumen (hematoxylin and eosin x100)

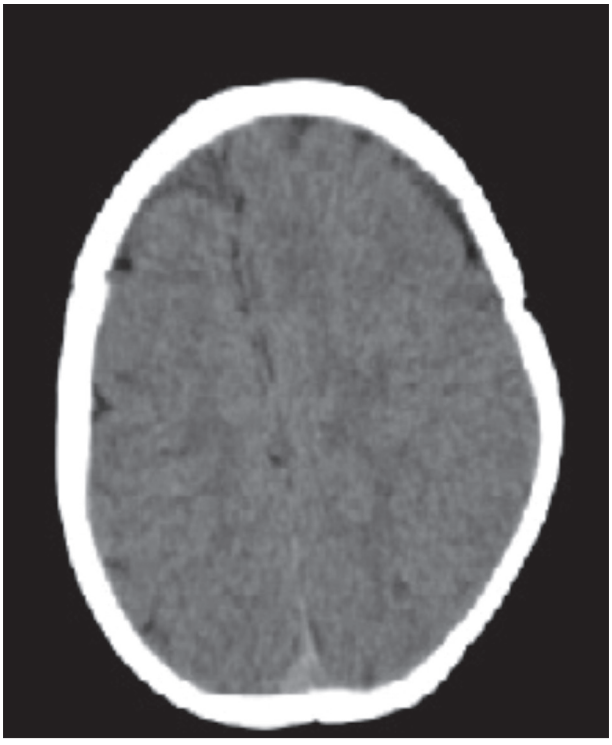

Figure 5. Follow-up computed tomography in 1 year shows no residual or recurred lesions. Bony defect is not seen in operation site

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

Surgical and Medical Practices: Z.K., B.Ç., Concept: A.Y., Design: Z.K., A.Y., B.Ç., Data Collection or Processing: Z.K., Analysis or Interpretation: B.Ç., Literature Search: Z.K., A.Y., Writing: Z.K.

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

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References

1. Di Emidio P, Chibbaro S, Mirone G, Balercia P. Double skull lymphangioma. Case report and review of the literature. Orbit 2009; 28: 293-6.
2. Ito E, Saito K, Nagatani T, Terenishi M, Kamei Y, Yagi S, et al. Lymphangioma of the skull base bones leading to cerebrospinal fluid rhinorrhea. J Neurosurg Pediatr 2008; 2: 273-6.
3. Kaya S, Colak A, Kutlay M, Gungor A. Lymphangioma of the clivus: Case report. Neurosurgery 2007; 60: 779.
4. Berkman MZ, Bek S, Ozcan D. Lymphangioma of the skull. A case report. Turkish Neurosurg 2001; 11: 56-9.
5. Willatt JM, Quaghebeur G. Calvarial masses of infants and children. A radiological approach. Clin Radiol 2004; 59:474-86.
6. Naguib MB, Al-Jazan N, Hashem T. Lymaphangiomatosis: A differential diagnosis of lytic lesions of the temporal bone. J Laryngol Otol 2008; 122: 23.
7. Okazaki T, Iwatani S, Yanai T, Kobayashi H, Kato Y, Marusasa T, et al. Treatmen of lymphangioma in children: Our experience of 128 cases. J Pediatr Surg 2007; 42: 386-9.
8. Maki DD, Nesbit ME, Griffths HJ. Diffuz lymphangiomatosis of bone. Australas Radiol 1999; 43: 535-8.
9. Kopperman M, Antoine JE. Primary lymphangioma of the calvarium. Am J Reoentgenol Radium The Nucl Med 1974; 121: 118-20.
10. Yoon SH, Park SH. A study of 77 cases of surgically excised scalp and skull masses in pediatric patients. Childs Nerv Syst 2008; 24: 459-65.
11. Alqahtani A, Nguyen LT, Flageole H, Shaw K, Laberge JM. 25 years experience with lymphangiomas in children. J Pediatr Surg 1999; 34: 1164-8.
12. Wallance MJ, Ross M. Bone lymphangiomatosis: Treatment with percutaneosu cementoplasty. Spine 2005; 30: 336-9.
13. Vural M, Açıkalın MF, Adapınar B, Atasoy MA. Congenital Cavernous Hemangioma of the Calvaria: A Case Report. J Neurosurg Pediatrics 2009; 3: 41-5.
14. Josan VA, Sogouros S, Walsh AR, Dover MS, Nishikawa H, Hockley AD. Cranioplasty in Children. Childs Nerv Syst 2005; 21: 200-4.
15. Moreira-Gonzalez A, Jackson IT, Miyawaki T, Barakat K, DiNick V. Clinic outcome in cranioplasty: Critical review in the long term follow-up. J Craniofac Surg 2003; 14: 144-53.

Letter to the Editor / Editöre Mektup

Place in Concomitant Use with Anti-EGFR Radiotherapy in Locally Advanced Head and Neck Cancer Treatment

Anti-EGFR Tedavilerin Lokal İleri Baş-Boyun Kanserlerinde Radyoterapi ile Eş Zamanlı Kullanımındaki Yeri

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Dear Editor;

Head and neck cancer (HNC) forms 8% of the adult cancers and each year approximately 600.000 new cases occur. In the course of application, about 40% of the cases are in early stage, 50% is locally advanced (stage III and IV) and 10% is in metastatic stage. More than 90% of the cases are squamous cell carcinoma and alcohol and smoking are common etiological factors. Human papillomavirus virus is a risk factor especially for oropharyngeal squamous cell carcinoma (1).

The anatomic localization of the disease, the stage of the disease, pathological features and the other diseases of the patient (comorbidity) are important factors to choose the treatment. Although there is not only one approach of treatment, surgical cure and radiotherapy have the similar survival results in early stage. For locally advanced cases, radiotherapy or concomitant chemoradiotherapy is widely used. In concomitant chemoradiotherapy implementations, cisplatin is the gold standard treatment. However, the toxicity related with chemotherapy (particularly cisplatin) restricts the application of chemoradiotherapy with especially the patients who have comorbidity. Epidermal growth factor receptor (EGFR) shows abnormal activity in several epithelial cancers including HNC. Therefore, there are so many done and ongoing studies, which are about different kinds of cancers with the agents who target this pathway (2).

Firstly, in the study of Bonner et al. (3) only radiotherapy (RT) and cetuximab treatments with RT are compared. In combination arm in comparison with only RT, a significantly better local-regional control is statistically provided (median 14.9 months against 24.4 months and p=0.005). Accordingly, while median survival is 49 months in cetuximab and RT arm for overall survival (OS), it has been found more superior in combination arm as 29.3 months (p=0.03) in RT arm.