Received: 31.01.2010 / **Accepted:** 26.04.2010
DOI: 10.5137/1019-5149.JTN.2950-10.1

Congenital Cavernous Hemangioma of the Skull

Kafatasının Konjenital Kavernöz Hemanjiomu

Engin YUCEL[1], Hampar AKKAYA[2], Doga GÜRKANLAR[1], Tarkan ERGUN[3]

[1]Baskent University, Faculty of Medicine, Department of Neurosurgery, Alanya/Antalya, Turkey
[2]Baskent University, Faculty of Medicine, Department of Pathology, Alanya/Antalya, Turkey
[3]Baskent University, Faculty of Medicine, Department of Radiology, Alanya/Antalya, Turkey

Correspondence address: Doğa GURKANLAR / E-mail: dgurkanlar2000@yahoo.co.uk

ABSTRACT

Calvarial cavernous hemangiomas (CHs) comprise 0.2% of benign neoplasms of the skull and frequently occur in the second and fourth decades. Their occurrence in infancy is extremely rare and they can initially be misdiagnosed as cephal hematoma or caput succadeneum that can occur due to the birth trauma, but trauma is not a predisposing factor. CT scan is more helpful than other neuro-imaging studies and untreated CHs of the skull may show progression. Only two cases of congenital primary CH of the skull without intracranial invasion have been reported in the literature. Herewith we reported a 4-month-old infant operated on due to a CH of the parietal bone and discussed the diagnostic and therapeutic modalities.

KEYWORDS: Cavernous hemangioma, Infant, Calvarium

ÖZ

Kafatasını tutan kavernöz hemanjiomlar (KH) , bu bölgenin iyi huylu tümörlerinin %0,2'sini oluştururlar ve sıklıkla hayatın ikinci ve dördüncü dekadlarında görülürler. Yenidoğanda görülmeleri ise oldukça nadirdir ve başlangıçta doğum travmasına bağlı gelişen cefal hematom veya caput succadeneum ile karıştırılabilirler. Ancak travma bu patolojinin oluşumunda rol alan faktörler arasında görülmemektedir. Bilgisayarlı tomografi diğer görüntüleme yöntemlerinden çok daha yararlıdır ve tedavi edilmeyen kavernöz hemanjiomlar yayılım da gösterebilirler. Literatürde bugüne kadar kafa içi invazyon olmaksızın sadece iki primer konjenital kavernöz hemanjioma rastlanmıştır. Biz burada parietal kemiğinde bulunan KH nedeniyle ameliyat edilen 4 aylık bir yenidoğanı sunduk ve tanı ve tedavi şekillerini literature eşliğinde tartıştık.

ANAHTAR SÖZCÜKLER: Kavernöz hemanjiom, Yenidoğan, Kafatası

INTRODUCTION

Cavernous hemangiomas (CHs) of the skull are rare tumors arising from the intrinsic vasculature of the bone and are mostly seen in middle age (3,7,8,9,11,13,14,21,25). The incidence of CH in infancy is quite low (6,16,17,25). They comprise 0.7% of all bone neoplasms and the most common site is the vertebral body, followed by the skull (12,15).

According to our knowledge, only two cases of congenital primary CH of the skull without intracranial invasion have been reported in the literature (23,25). Herewith we report a 4-month-old infant harboring a CH of the parietal bone.

CASE REPORT

A 4-month-old female infant, delivered vaginally, presented with a right 3cm solid mass lesion located in the parietal bone. This lesion was first noted at her birth and was diagnosed as caput succadeneum. However this solid mass had remained stable even after the resolution of caput succadeneum.

Her neurological examination revealed no abnormality except the right parietal solid mass lesion, three cm in diameter. CT showed a soft tissue density mass that expanded the diploic space on the right side. Both inner and outer tables were increased in the thickness and there were some defective areas on the cortex of the inner table (Figure 1).

The patient was operated on under general anesthesia. A right parietal craniectomy with total excision of the lesion and a margin of surrounding uninvolved bone was performed.

Histopathologic examination revealed dilated and proliferated vascular space between the bone trabeculae, consistent with an intraosseous cavernous hamangioma (Figure 2). Monoclonal antibody to CD34 highlights a single layer of flattened endothelial cells lining the dilated vascular spaces (Figure 3).

DISCUSSION

Calvarial CHs comprises 0.2% of benign neoplasms of the skull (24) and the parietal bone is the most common site (8). CH frequently occurs in female patients (21), especially in the second and fourth decades of their life (16,17,21). Only 9% of cases are observed during the first decade (6) and occurrence of CH in neonatal period is rare. There are only two reports of solitary CH in a neonate in the literature (23,25) and our case is the third harboring CH.

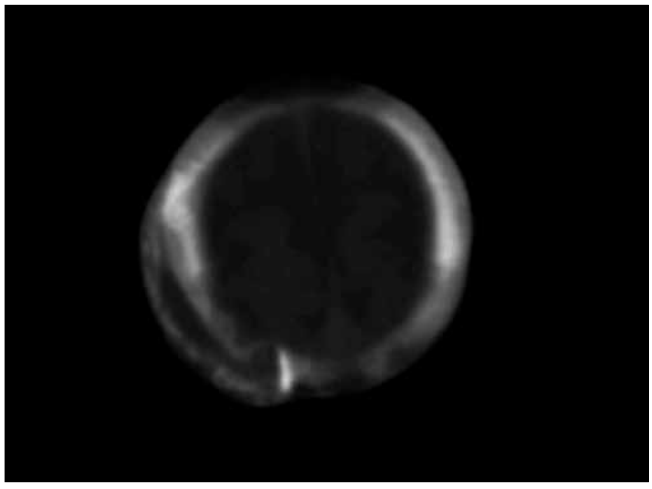

Figure 1: CT showed a soft tissue density mass that expanded the diploic space on the right side.

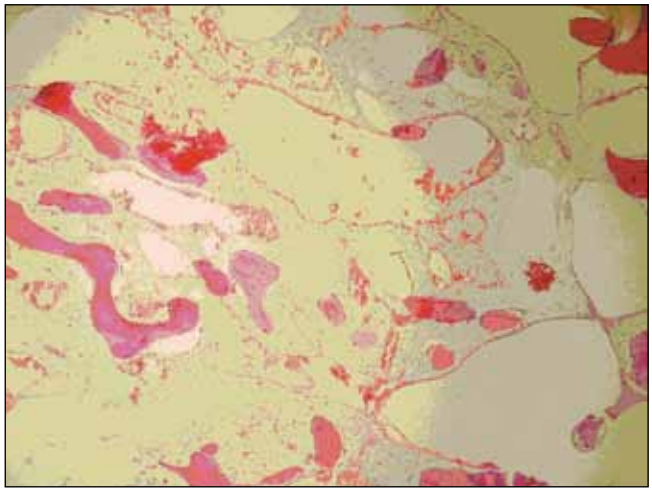

Figure 2: Dilated and partialy blood-filled vascular space between the bone trabeculae (hematoxylin-eosin, original magnification x50).

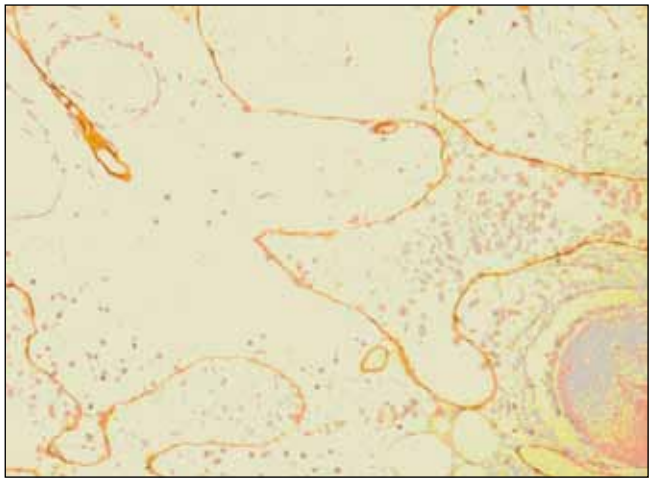

Figure 3: Monoclonal antibody to CD34 highlights a single layer of flattened endothelial cells lining the vascular spaces (original magnification ×200).

Calvarial CHs arise from the vessels in the diploic space and supplied by the branches of the external carotid artery, arising in the skull vault. The middle and superficial temporal arteries are the main sources of blood supply (13,14). Although trauma is not a predisposing factor (18,21,25), CHs can initially be misdiagnosed as cephal hematoma or caput succadeneum that can occur due to the birth trauma (23,25). Repeated hemorrhage may lead to an increase in tumor size (10). The bony trabeculae seen within this tumor is the result of osteoclastic remodelling and osteoblastic reinforcement by the growing vascular tumor (1).

Preoperative radiological studies include conventional radiography, computerized tomography and magnetic resonance imaging. Although prenatal ultasonography reported to be helpful in the diagnosis of calvarial CH, prenatal US of our patient revealed no pathology (12). Osteoblastic remodelling with trabecular bone following osteoclastic activity of the tumor results in typical sun burst appearance of CH (15,19). We performed CT of the skull before the operation, because CT scan is more helpful than other neuroimaging studies (22). We also performed CBC count in order to see the platelet levels due to the fact that in infants platelets may become trapped in the interstices of such lesions, leading to profound consumption coagulopathy (20).

Aneurysmal bone cyst, osteoma, giant cell tumor, fibrous dysplasia , sarcoma, meningioma, metastatic disease, Paget disease, dermoid and epidermoid cyst should be taken into consideration in the differential diagnosis of CH of the skull (4,5,22).

Untreated CHs may involve skull base and cause progressive exophtalmia, impairment of ocular functions and cosmetic alterations (2). So, we performed monobloc resection with removal of a rim of a normal bone in order to prevent recurrence (1,7,13,19). This technique also prevents excessive intraoperative bleeding (12,19).

CONCLUSION

CH of the skull is extremely rare in infants with the risk of intraoperative excessive bleeding. Although it may not show the characteristic features, CT is still the most helpful neuroimaging study. Prenatal US is not always helpful in determining CHs and they can initially be misdiagnosed as cephal hematoma or caput succadeneum.

REFERENCES

1. Ajja A, Oukacha N, Gazzaz M, Akhaddar A, Elmostarchid B, Kadiri B, Oukabli M, Boucetta M: Cavernous hemangioma of the parietal bone, case report. Neurosurg Sci 49:1-4, 2005
2. Awaya H, Ito K, Honjo K, Fujita T, Matsumoto T, Matsunaga N: Differential diagnosis of hepatic tumors with delayed enhancement at gadolinium-enhanced MRI: A pictorial Essay. Clin Imaging 22:180-187, 1998
3. Bergeson PS, Rekate HL, Tack ED: Cerebral cavernous angiomas in the newborn. Clin Pediatr 31:435-437, 1992

4. Bnerji D, Inao S, Sugita K, Kaur A, Chhabra DK: Primary intraosseous orbital hemangioma. A case report and review of the literature. Neurosurg 35:1131-1134, 1994
5. Buhl R, Barth H, Dorner L, Nabavi A, Rohr A, Mehdorn HM: De novo development of intraosseous cavernous hemangioma. J Clin Neurosci 14:289-292, 2007
6. Cervoni L, Artico M, Delfini R: Intraosseous cavernous hemangioma of the skull. Neurosurg Rev 18:61-64, 1995
7. Cuesta GM, Navarro-Villa C: Intraosseous cavernous hemangioma of the zygomatic bone. A case report (review). J Oral Maxillofac Surg 21:287-291, 1992
8. Hamilton HB, Voorhies RM: Tumor of the Skull. Neurosurgery 2nd ed. Volume 3. New York: McGraw Hill, 1996:1520-1521
9. Heckl S, Aschoff A, Kunze S: Cavernomas of the skull. Review of the Literature 1975-2000. Neurosurg Rev 25:56-62, 2002
10. Herter T, Brandt M, Szuwart V: Cavernous hemangiomas in Children. Childs Nerv Syst 4:123-127, 1988
11. Hoffman DF, Israel J: Intraosseous frontal Hemangioma. Head neck 12:160-163, 1990
12. Honda M, Toda K, Baba H, Yonekura M: Congenital cavernous hemangioma of the temporal bone, case report. Surg Neurol 59:120-123, 2003
13. Hook SR, Font RL, McCrary JA, Harper RL: Intraosseous capillary hemangioma of the frontal bone. Am J Ophtalmol 103:824-827, 1987
14. Khanam H, Lipper MH, Wolff CL, Lopes MB: Calvarial Hemangiomas: Report of two cases and review of the literature. Surg Neurol 55(1):63-67, 2001
15. Liu JK, Burger PC, Harnsberg H, Couldwell WT: Primary intraosseous skull base cavernous hemangioma, case report. Skull Base 13:219-228, 2003
16. Martinez –Lage JF, Capel A, Costa TR, Perez-Espejo MA, Poza M: The child with a mass on its head: Diagnostic and surgical strategies. Child's Nerv Syst 8:247-252, 1992
17. McAllister RM, Rutty GN, Hancock K, Sanders R: Cavernous hemangioma of the nasal bone. J Laryngol Otol 106:264-267, 1992
18. McIntyre NG, Brenner DM, Gluckman J: Primary Cavernous hemangioma of the frontal bone: A Case Report. S Afr Med J 52:537-538, 1977
19. Naama O, Gazzaz M, Akhaddar A, Belhachmi A, Asri A, Elmostarchid B et al: Cavernous Hemangioma of the skull: 3 case reports. Surg Neurol 70:654-659, 2008
20. Orenstein DM, Yonas H, Bilenker Hl, White RJ: Hemangioma thrombocytopenia syndrome. A case masquerading as an Encephalocele. Am J Dis Child 131:680-681, 1977
21. Peterson DL, Murk SE, Story JL: Multifocal cavernous hemangioma of the skull: Report of a case and review of the literature (review). Neurosurgery 30:778-781, 1992
22. Suzuki Y, Ikeda H, Matsumoto K: Neuroradiological features of intraosseous cavernous hemangioma, case Report. Neurol Med Chir 41:279-282, 2001
23. Vural M, Acıkalın MF, Adapinar B, Atasoy M: Congenital cavernous hemangioma of the calvaria. Case report. J Neurosurg Pediatrics 3:41-45, 2009
24. Wyke BD: Primary Hemangioma of the skull, a rare cranial tumor. Review of the literature and report of a case with special reference to the roentgenographic appearances. Am J Roentgen Radiotherapy 61:302-316, 1949
25. Yoshida D, Sugisaki Y, Shimura T, Teramoto A: Cavernous hemangioma of the skull in a neonate. Child Nerv Syst 15: 351-353, 1999