Cilt 18/ Sayı 1

KANAMAYLA BAŞVURAN VE ATİPİK YERLEŞİMLİ İNFANTİL HEMANJİYOPERİSİTOM: OLGU SUNUMU

INFANTILE HEMANGIOPERICYTOMA IN AN ATYPICAL LOCATION AND WITH AN UNUSUAL PRESENTATION: A CASE REPORT

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ÖZET

Hemanjiyoperisitomlar çocukluk çağının nadir görülen damarsal tümörlerindendir. Bu çalışmada acil servise sol uyluk arka yüzünden kanama şikayeti ile başvuran 30 günlük erkek hastadaki ülsere hemanjiyoperisitom olgusu sunuldu. Cerrahi öncesi hemodinamik stabilitesi sağlanan hastada kitlenin ana besleyici damarları girişimsel radyoloji bölümünce embolize edildi. Embolizasyon sonrası kitle önemli bir kanama olmaksızın eksize edildi. Cerrahi sonrası dönem sorunsuz geçti ve bir buçuk yıl sonrası kontrollerinde nüks görülmedi.

ABSTRACT

Hemangiopericytomas are unusual vascular neoplasms that occur in children. We report a case of an ulcerated hemangiopericytoma on the left posterior thigh in a male infant who presented for evaluation of hemorrhage. After achieving hemodynamic stability and prior to surgery, the patient was referred to the interventional radiology department for embolization of the large nourishing artery of the hemorrhagic mass. The mass was excised with negligible blood loss. The postoperative clinical course was uneventful, and there was no recurrence during 18 months of follow-up.

INTRODUCTION

First described by Stout and Murray,[1] hemangiopericytomas (HPCs) are rare, vascular tumors thought to be derived from vascular pericytes. The majority of these tumors occur in the 5th and 6th decades of life and are therefore referred to as the adult type, but 10% of all cases occur in children and infants.[2,3] In adults, HPCs are usually located in the lower limbs, followed by the retroperitoneum, the head, and the neck. In childhood, the most common site is the head and neck, followed by the retroperitoneum and limbs.[4]

The congenital form is extremely rare, comprising only 3.3-7% of all HPCs.[5] The congenital form is generally referred to as the infantile type and comprises the HPCs that occur in children < 1 year of age. Infantile HPCs are detected at birth or during the first two months of life.[6] Infantile HPCs follow a more benign clinical course than the adult type or the type which occurs in children > 1 year of age. HPCs rarely cause profuse bleeding if located superficially.[7-9] Traumatic or spontaneous bleeding from the superficial lesions may be serious and require immediate intervention in children.[7-9]

In this report, we present the case of a 30-day-old male with an infantile HPC on the left posterior thigh causing abundant hemorrhage.

CASE REPORT

A 30-day-old male infant was referred to our hospital for evaluation of a bleeding, ulcerated mass on his left posterior thigh (Fig. 1a). He was born as a second child to a healthy mother after an uncomplicated pregnancy and normal vaginal delivery. His family history was negative for congenital anomalies. The lesion had existed from birth and was approximately 3 × 3 cm in size at birth. The initial diagnosis made by the pediatrician was a hemangioma. The lesion began to hemorrhage when he was 8 days of age. At 17 days of age, he required a blood transfusion that was administered at a pediatrics clinic. On physical examination, there was a 4 × 5 cm firm exophytic mass that was purple-black in color. The mass was not fixed to deep structures. There was a distal surface ulceration on the lesion.

The results of the infant's laboratory tests re-

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vealed the following values: hemoglobin, 7.99 g/dl; hematocrit, 21.97; activated partial thromboplastin time, 26.80 sec (normal range, 26.0-36.0 sec); prothrombin time, 13.50 sec (normal range, 11.00-15.00 sec); and platelet count, 132.20/mm^3 (normal range, 130.000-400.000/mm^3). Our presumptive diagnosis on admission was a vascular malformation. Forty cc of packed red blood cells were given immediately. The bleeding was controlled with a compressive dressing. An abdominopelvic ultrasonography was performed, and no evidence of hepatosplenomegaly was seen. Preoperatively, magnetic resonance angiography was obtained to delineate the lesion. On the magnetic resonance angiography images, we discerned a highly vascularized mass supplied mainly by the branch of the deep femoral artery in the proximal portion and by small branches from the superficial femoral artery in the distal portion (Fig. 1b). The large branch arising from the deep femoral artery, which supplied the proximal part of the mass, was occluded with an N-butyl-2-cyanoacrylate-lipiodol mixture by an interventional radiologist (Figs. 1b and c). The distal vessels could not be occluded because there were multiple small branches. The next day, the patient underwent surgery. The mass extended to the subcutaneous tissue, but there was no involvement with any of the underlying muscles or fascia. The mass was elevated above the deep fascia and excised; there was negligible blood loss. The defect resulting from the excision was closed primarily after subcutaneous undermining of the surrounding tissues. A HPC was diagnosed after the histopathological examination and immunohistological analyses of the excised specimen. The postoperative clinical course was uneventful, and there was no tumor recurrence during 18 months of follow-up (Fig. 1d).

HISTOPATHOLOGY

Microscopic evaluation of the excised specimen showed uniformly arranged, thin-walled, endothelial-lined, intercommunicating vascular channels and immature mesenchymal spindle cells located around the endothelial cells (Fig. 2a). There were central areas of myxoid and hyaline degeneration within the tumor, but there was no necrosis or lymphovascular invasion. The subcutaneous tissue was infiltrated with tumor cells. Tumor cells were separated from each other by rich reticulin fibers (Fig. 2b). Two mitotic figures were noted in 10 high-power fields, consistent with a low malignant potential. CD34 was diffusely positive in the vessels, but focally positive in the tumor cells (Fig. 2c). Factor-8–related antigen and smooth muscle antigen stained weakly in focal areas of the tumor. Desmin, S-100, and CD31 were negative. The histological findings and immunohistological analyses were consistent with the diagnosis of a HPC.

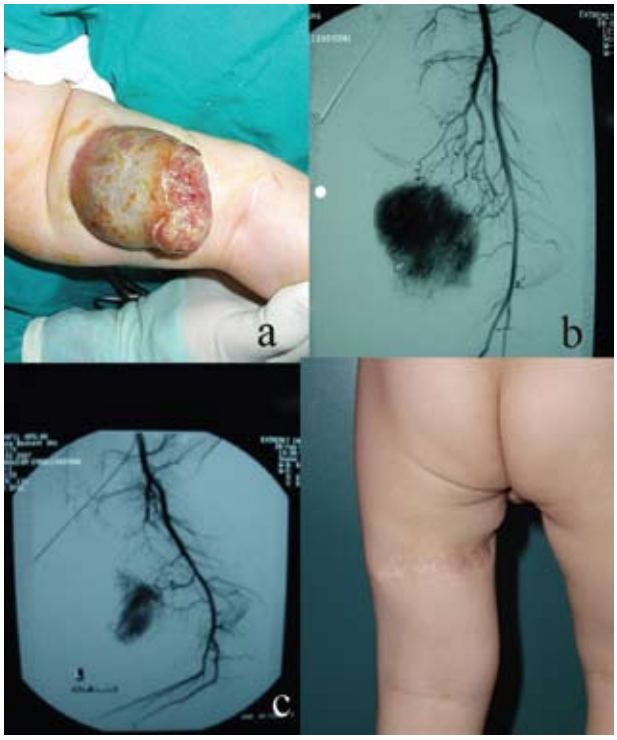

Fig. 1a: Preoperative view of the lesion.
1b: Nourishing pattern of the lesion before the selective embolization.
1c: View of the vascular leash after selective embolization.
1d: Postoperative view of the patient 14 months after surgery.

DISCUSSION

The diagnosis of HPCs is challenging due to the marked variability in the gross and histological appearance of the lesion, as well as the growth patterns exhibited by HPCs.[3] Congenital HPCs most often occur in the subcutaneous tissues, although a few cases in muscle and brain have been reported.[1,5,10] Childhood HPCs commonly occur in the head and neck, and rarely occur in the lower extremities.[4,6,11] Other rare locations for congenital HPCs include the nasopharynx, tongue, duodenum, retroperitoneum, and stomach.[5,12-15]

In the lower extremities, infantile HPCs are generally located in the subcutaneous tissues[4,11] without superficial ulceration.[7] Virden et al.[4] reported a subcutaneous congenital HPC arising from the medial belly of the long head of the biceps femoris muscle in the posterior thigh. Hamada and coworkers[11] reported a subcutaneous congenital HPC arising in the lower right leg. The only case of an ulcerated lower extremity congenital HPC was reported by Resnick and coworkers5 in the buttock of a 3-week-old boy. It appears that congenital HPC lesions are more prone to ulceration and bleeding when located closer to the epidermis. Also, if vascular channels are prominent, the tumor tends to be more hemorrhagic and dark-red in color.[7-9] As of this writing, only three cases of hemorrhagic congenital HPC have been

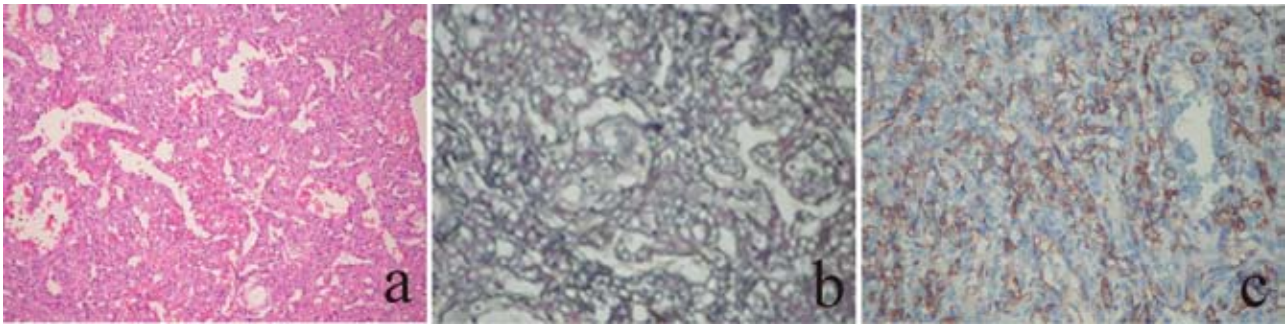

Fig. 2a: Microscopic view of the specimen (hematoxylin and eosin stain × 200).
2b: Immunologic stain showing rich reticulin fibers (Reticulin × 400)
2c: CD 34 immunostain of the specimen (CD 34 × 400)

reported; these were reported in the buttock, hand, and midline of the back.[7-9] None of those patients died of hemorrhage, but all required urgent surgery. The patient with the lesion on the right buttock had significant bleeding and hypovolemic shock.[7] Blood transfusions and emergency surgery were required to manage the hemorrhaging lesion.[7] Because of increased hemorrhaging of a finger HPC, Templeton and coworkers[8] had to amputate the affected digit. Hemostasis was accomplished with vasopressors, sustained direct pressure, surgical packing, and volume expansion. Emergency surgery was required for the patient with the HPC in the midline of the back.[9]

HPCs in adults have malignant characteristics and tend to metastasize.[3] In infants, HPCs have a more favorable behavior than in children > 1 year of age.[6,16] The clinical behavior in children > 1 year of age does not appear to differ from the adult counterpart and aggressive multimodality therapy is required. Tumors that arise early in childhood have lower recurrence rates and better prognoses than do those that arise later in childhood.5 However, aggressive behavior has been reported in childhood HPCs, albeit rarely.[5,17,18] Also, metastases of congenital HPCs have been reported.[19,20] Toren and coworkers reported the spontaneous regression of a huge gluteal HPC within 2 months. They advise the conservative approach to the CHP in the neonatal period.[21]

Congenital HPCs do not show the high mitotic count and necrosis as exist in adult HPCs.[3] A mitotic rate of 4 or more per 10 high-power fields indicates a rapidly growing HPC capable of recurrence and metastases.[3] However, no distinct histological criteria are capable of defining the grade of malignancy of HPCs in children to date.[16] In the biopsy specimen obtained from the patient reported herein, 2 mitotic figures were noted in 10 high-power fields, and there was no necrosis.

Complete surgical resection with a sufficient margin is the main therapy for congenital HPC lesions in patients whose lesions are completely resectable.[3,5,17] Incomplete excision may lead to local recurrence, especially for lesions around the larynx, tongue, trachea, and great vessels.[13,17] In patients with unresectable tumors, incomplete resection, recurrent lesions, and metastases, chemotherapy is considered the next line of therapy.[6,17] When the disease is considered not completely resectable at diagnosis, 10-12 weeks of chemotherapy can be administered to shrink the tumor.[6] Complete surgical resection with clear surgical margins was achieved in our patient. Therefore, chemotherapy was not required before or after the surgery. The results of chemotherapy and radiotherapy are not impressive. Radiotherapy is reserved for patients who have failed surgery and chemotherapy.[6, 22]

Our patient's pediatrician originally thought the lesion was a hemangioma. Hemangiomas are generally flat or undetectable at birth. They are usually soft and bright red. Congenital HPCs are dark red and firmer. Histologically, differentiation of HPCs from hemangioendotheliomas of childhood is facilitated by reticulin stain. Normal endothelial cells in hemangioendotheliomas show negative staining by reticulin. Although significant bleeding is not common in hemangiomas, congenital HPC may present the risk of a life-threatening hemorrhage.[7-9]

Selective catheter-directed embolization of the feeding arteries of an arteriovenous malformation before surgery has been used routinely.[23] Also combined percutaneous sclerotherapy and ligation of the main vessels for the treatment of venous malformation at difficult location such as oral and oropharyngeal area has been described in the literature.[24] However, selective arterial embolization of the HPC before surgery has not been previously reported. In our patient, selective arterial embolization reduced the bleeding during the surgery and the need for blood transfusion. We advise selective arterial embolization of the congenital HPC before surgery if a large nourishing artery is detected during the MR angiography.

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