KAPİLLER MALFORMASYONDAN KAYNAKLANAN RETİFORM HEMANJİYO-ENDOTELYOMA: NADİR BİR OLGU

RETIFORM HEMANGIOENDOTHELIOMA ORIGINATED FROM CAPILLARY MALFORMATION: A RARE CASE

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ÖZET

Retiform hemanjiyoendotelyoma son zamanlarda tanımlanan, karakteristik özellikli, düşük dereceli anjiyosarkom alt grubunda bir hastalıktır. Yüksek lokal nüks olasılığına karşın, uzak metastaz ihtimali düşüktür. Hemen hiç hastalıkla ilişkili ölüm olmaması ve sadece iki bölgesel lenf nodu metastazlı vaka bildirilmesi nedeni ile, geniş cerrahi eksizyon önerilen tedavi yöntemidir.

Çalışmada, 8 yaşındaki kızın sol kolunda bulunan kapiller malformasyon üzerinde oluşan retiform hemanjiyoendotelyoma sunulmuştur. Olgu literatürde yer alan retiform hemanjiyoendotelyoma olguları ışığında tartışılmıştır. Buna göre kapiller malformasyondan kaynaklanan bilinen ilk retiform hemanjiyoendotelyoma olgusudur ve literatürdeki en genç olgulardan biridir.

Tedavide yüksek nüks oranı nedeniyle anatomik sınırlar elverdiğince geniş eksizyon yapılmalıdır. Geniş lezyonlarda, nükslerde ve lenf nodu metastazlarında radyoterapi düşünülebilir.

Anahtar kelimeler: Retiform hemanjiyoendotelyoma, çocuk, kapiller malformasyon

ABSTRACT

Retiform hemangioendothelioma is recently defined, a distinctive variant of low-grade angiosarcoma of the skin. It has a potential of high local recurrence but low distant metastasis. Because there have been no tumor-related deaths and only two case of regional lymph node metastasis; wide surgical excision is the most suggested treatment method.

We document a case of retiform hemangioendothelioma that is originated from the capillary malformation of the left arm skin of an 8-year-old girl. Our case is discussed in the background of previously reported cases concerning retiform hemangioendothelioma in English-written literature. As far we know this is the first case of the retiform hemangioendothelioma originating from the capillary malformation and she is the youngest case.

Key words: retiform hemangioendothelioma, child, capillary malformation

INTRODUCTION

Retiform hemangioendothelioma (RH) is a very rare variant of low-grade, well-differentiated cutaneous angiosarcoma. The entity first described by Calonje et al. in 1994 with report of 15 cases.1 Thereafter, few case reports have been published in the literature. Differential diagnosis with conventional angiosarcoma is necessary to avoid aggressive therapy.[2-5]

In this paper we present a case of RH that is unusually occurred on capillary malformation consisting skin. To the best of our knowledge, this is the first case describing a RH originating from the capillary malformation. This is also one of the youngest case of RH in English-written literature.

CASE REPORT

An 8-year-old girl admitted to dermatology clinic for the evaluation of a painful, slowly enlarging wound which was appeared on her left elbow about four months ago. During the initial examination, dark livid macular lesions were inspected all over the left thoracic region, shoulder and arm. They all diagnosed as capillary malformations (nevus flammeus). There was also a 25x25 mm solitary crusted ulceration on the left elbow, with a same size mass beneath. There were no other complaints or physical signs. No trauma was remembered by the patient to the area. No significant past medical history determined. (Figure 1-A)

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A 6-mm punch biopsy was taken from the margin of the ulcer. Histopathologic examination was exposed that aborizing blood vessels in a retiform pattern were beneath the stratified squamous epithelium and in a collagen connective tissue. Moderate lymphocytic infiltrate was conspicuous both within the endovascular papillae and the dermis. Between the retiform pattern arranging CD34 (+) vascular endothelial cells with wide cytoplasm, in some areas, hobnail like endothelial cells were present. No significant atypia or mitotic figures were seen. Histological diagnosis was reported as RH with these findings. (Figure 2)

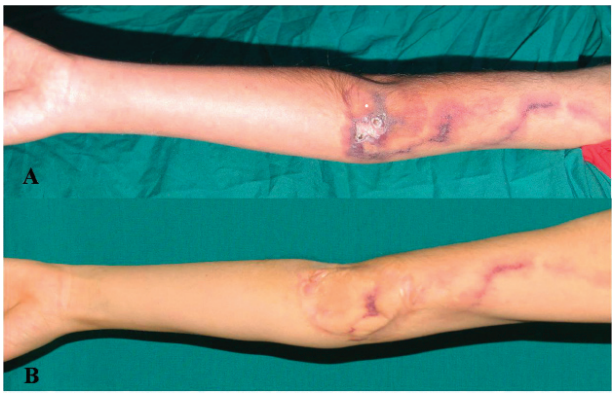

Figure 1.

A. Preoperative view of the the tumor which was occurred on capillary malformation of left elbow.

B. Postoperative 3rd year control view of the area which was reconstructed with proximally based V-Y advancement flap.

Case was consulted to our clinic for further investigations and treatment. X-rays of the elbow were normal. Magnetic resonance imaging of the area showed a 25x20 mm non-encapsulated subcutaneous mass which was taking up contrast agent, nearby the olecranon and the ulnar nerve. (Figure 3) Laboratory examinations including liver function test results and complete blood count were within normal range. Axillary ultrasound inspection detected no evidence of regional lymph node involvement. Computed tomography scan of her chest and abdomen revealed no metastatic disease.

Surgical excision with 20 mm margin was applied. There was no involvement of the ulnar nerve and the bone, so these anatomical structures were preserved. The defect was reconstructed with proximally based V-Y advancement flap. The postoperative histopathological examination was verified preoperative diagnoses and showed the histopathological tumor-free margins. The patient received no adjuvant chemotherapy or radiation. The case followed up closely for 3 years with no recurrence or metastases. (Figure 1-B)

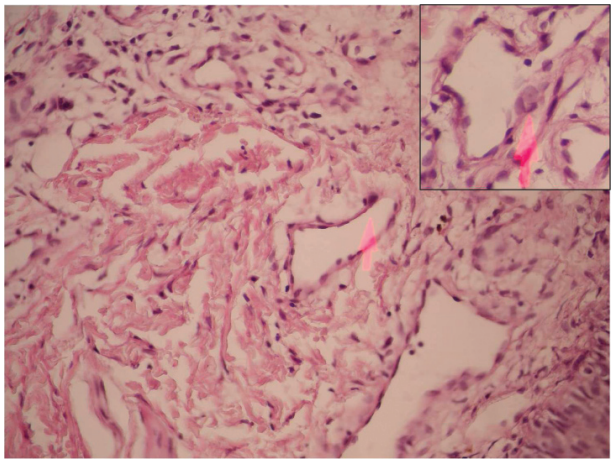

Figure 2. Retiform pattern with hobnail endothelial cells. Arrow indicates the hobnail endothelial cells (hematoxylin-eosin, magnification ×20). Upper right window also shows hobnail endothelial cells clearly (hematoxylin-eosin, magnification ×40).

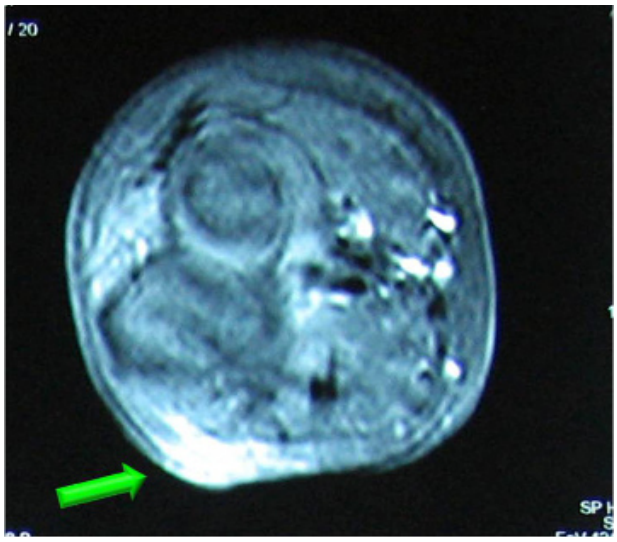

Figure 3. Magnetic resonance imaging (T2) view of the tumor which was taking up contrast agent, in close proximity to the olecranon and the ulnar nerve.

DISCUSSION

RH is a variant of low-grade angiosarcoma of the skin which recurs frequently, but has a very low metastatic rate.[2] It is first described by Calonje et al. in 1994 with report of 15 cases.[1] Few additional case reports (20 cases) have been added to the literature until now. RH has no distinctive clinical appearance. The lesions were exophytic, dermal, or subcutaneous slowly growing nodules or plaques measuring 1-30 centimeters in diameter. They especially don't suggest the vascular nature of the tumor. Tumors were appeared on the lower limbs (45%), upper limbs (20%), trunk (20%), genital region

(6%), and head (9%). RH usually occur as a solitary tumor, but in a single case, multiple lesions developed in different anatomic sites.6 RH affects usually young and middle aged adults. The age of the reported patients in English Literature ranged from 9 to 78 years. It rarely occurs in infancy. So that, there is only 3 cases under 10 years old and our case is the youngest one. There is also a female predilection (16/9).[2-5]

The etiology of RH is unknown. Although the majority of cases arise spontaneously with no known precipitating event, one case has been described in a setting of chronic lymphedema, another after radiation treatment[1] and HHV8 DNA sequences were claimed in a different case.[7] Medical histories of the some patients consisted of previously treated cervix cancer (2 cases),[1,7] thyroid papillary cancer,[8] adrenocortical neoplasm (incidentaloma).[3] In our case the tumor is unusually occurred on capillary malformation consisting skin. Although there is a possibility of this case to represent a mere coincidence, we cannot rule out a possible relation between both entities. Also accumulation of these rare cases in the literature will help for forthcoming etiological studies.

Characteristic histopathologic findings are essential, because the clinical features don't provide help about the exact diagnosis. The distinctive microscopic features of the disease are characterized by infiltrative vascular spaces arranged in a pattern similar to the rete testis with a prominent lymphocytic infiltrate and spaces lined by hobnail or papillary projections of endothelial cells. The vascular spaces infiltrate collagen bundles of the reticular dermis, and a myxoid background may be present.[2-5] Immunohistochemically, the reaction of the tumor cells with endothelial markers (CD31, CD34, factor VIII related antigen) are positive but D2-40 and VEGFR-3 is controversial.[9]

The term "low grade angiosarcoma" refers to a group of vascular neoplasms histopathologically stay intermediate between hemangioma and angiosarcoma. Differential diagnosis of RH includes tumors with hobnail endothelial cells primarily, such as targetoid hemosiderotic hemangioma (hobnail hemangioma) and Dabska's tumor. The former reveals the most similar clinical and histologic features with RH. Dabska's tumor occurs in infants and young children, most frequently on the head and neck as a small solitary lesion, consisting of a brown to violaceous papule surrounded by a thin pale area and a peripheral ecchymotic ring. Histologically, both tumors are composed of a proliferation of small round endothelial cells with a prominent lymphocytic infiltrate. But Dabska's tumor shows no retiform architecture and is composed of interconnecting cavernous vascular spaces, lymphangioma-like dilatation of the vessel.[2-5]

The most important differential diagnosis of RH is angiosarcoma. The differential diagnosis between these two diseases are important to preclude aggressive treatment. Angiosarcoma has an 85% mortality rate over 5 years whereas RH has local aggressive features but rare regional metastasis and no mortality. Both RH and angiosarcoma are infiltrative. However, in cases with overlapping features with angiosarcoma the diagnosis primarily is based on the degree of cytologic atypia and high mitotic rates. Other tumors that have to be considered for differential diagnosis are spindle cell hemangioendothelioma, tufted angioma, microvenular hemangioma.[2-5]

Because only two patients had regional lymph node metastasis[1,10], but there were neither distant metastases nor tumor-related deaths (except one controversial case)[11], all patients in the literature were treated with surgical excision with histopathologically tumor-free margins. There is a single case in that multiple lesions developed in different anatomic sites.[6] As mentioned by previous authors, current treatment choices are wide surgical excision considering the anatomical limits and radiotherapy for the cases with extended tumor sizes, local recurrence and regional lymph node metastasis.[1-5]

CONCLUSION

RH is considered to be a low grade vascular tumor so that it is important to distinguish it from conventional angiosarcoma to avoid overaggressive treatment.

The case reported here is similar to those in previous reports of RH. However, this case is unique, because the tumor developed on capillary malformation consisting skin and she was one of the youngest case in English literature.

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