

Retiform Haemangioendothelioma on Genital Area in a 70-Year-Old Female: A Case Report

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Abstract

Haemangioendothelioma is a rare tumor that is both clinically and histologically intermediate between angiosarcoma and haemangioma. It is usually a solitary, slow-growing nodule on distal areas or extremities. The onset is frequently before 25 years of age with a male preponderance. Histologically, two components are important: dilated vascular channels and spindle cells elements with some pleomorphism and mitotic configuration. Treatment is wide excision and regional lymph node evaluation. As we reviewed the reported papers, we just found one reported case on genital involvement in a male; therefore, this could be a rare presentation of haemangioendothelioma on the genital area of an old woman. (*Iran J Dermatol 2008;11: 129-131*)

Keywords: haemangioendothelioma, retiform haemangioendothelioma, genital tumor, haemangioma, angiosarcoma

Introduction

Haemangioendothelioma is a rare tumor that is both clinically and histologically intermediate between angiosarcoma and haemangioma. It is usually a solitary, slow-growing nodule on distal areas or extremities. The onset is frequently before 25 years of age with a male preponderance. Histologically, two components are important: dilated vascular channels and spindle cells elements with some pleomorphism and mitotic configuration. Treatment is wide excision and regional lymph node evaluation. As we reviewed the reported papers, we just found one reported case on genital involvement in a male; therefore, this could be a rare presentation of haemangioendothelioma on the genital area of an old woman.

Case Report

A 70-year-old woman who was suffering from a tumoral lesion on her labia major referred to our dermatology clinic. This lesion had appeared about 3 months ago as a painless papule. On clinical examination, a 2.5 cm tender, erythematous, oozing and fragile tumor was observed (Figure 1). There were no lymphadenopathy or remarkable systemic problems. She was using Atenolol for hypertension and calcium for osteoporosis. Chest x-ray and

routine blood tests and urinalysis were normal. Differential diagnoses included: Dabskas' tumor, composite haemangioendothelioma, reactive angioendothelioma, angioblastoma Nakagawa, spindle cell haemangioendothelioma, Kaposi sarcoma, epithelioid haemangioendothelioma, Kaposi like haemangioendothelioma, granuloma pyogenicum, SCC, giant condyloma, metastatic tumor and amelanotic melanoma.

A biopsy of the tumor showed an ulcerated neoplasm covered by fibrinoleukocytotic exudates that was composed of vascular channels lined by plump epithelioid endothelial cells and small clusters and sheets of proliferated epithelioid cells forming small inconspicuous lumina (Figures 2,3).

Immunohistochemical study showed positive reaction of tumor cells for Von Willebrand factor and CD34, and negative for CD31.

Pathological diagnosis was compatible with retiform haemangioendothelioma.

Discussion

Some authors believe that haemangioendothelioma is a useful designation for vascular neoplasms that has an intermediate histopathologic appearance between haemangioma and angiosarcoma¹.



Figure 1: Oozing tumor on labia minor

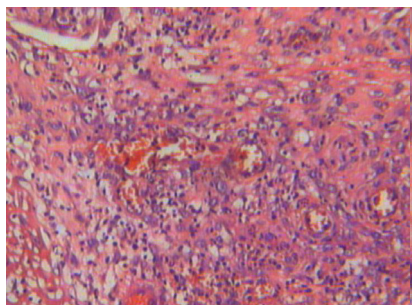


Figure 2: Vascular channels with sheaths of proliferated epithelioid cells (H & E staining, 40X)

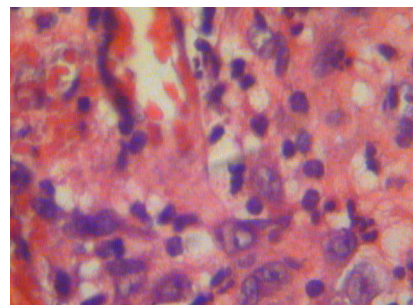


Figure 3: Plump epithelioid endothelial cells (H & E staining, 100X)

Haemangioendothelioma is a low grade angiosarcoma first described by Weiss and Enzinger² in 1982. It usually appears as a solitary, slightly painful soft-tissue tumor. It may occur at any age but is rare at childhood and affects both sexes equally. At least half of the cases are closely associated with or arise from a vessel, usually a vein³ and in some of these cases, the occlusion of the vessel accounts for most of the symptoms, such as edema or thrombophlebitis.

Haemangioendothelioma may occur at any site or organ; but in our search there was only one reported case on penile shaft involvement⁴ so this may be another rare case with genital involvement. Involvement of liver⁵, bones⁶, lungs⁷, spinal area⁸, intracranial cavity⁹, and brain¹⁰, mediastinum¹¹, and even nasal cavity¹² has been reported.

Epithelioid haemangioendothelioma is characterized by a proliferation of cord and nests of plump epithelioid cells embedded in a fibromyxoid or sclerotic stroma. Many of these cells contain vacuoles in their cytoplasm as a sign of primitive formation. Slight cellular pleomorphism and occasional mitotic figures may be seen. Rarely, large distinct vascular channels are seen.

A helpful feature for diagnosis is the presence of erythrocytes in the vacuoles of epithelioid haemangioendothelioma.

Several markers for blood vascular tumors and endothelial cell have been identified. Although CD34 marker was positive in our patient in immunoperoxidase staining, this is not specific for vascular tumors because some markers such as CD31-CD34 can also be detected on other dermal cells such as lymphatic endothelial cells (CD31) or fibrocytes (CD34) and haematopoietic precursor cells in bone marrow and circulation (CD34). The neoplastic cells of this tumor express immunoreactivity for factor VIII – related antigen

and Ulex europaeus lectin¹³ and usually do not express cytokeratin. Electron microscopic studies in haemangioendothelioma have demonstrated that neoplastic cells show characteristics of endothelial cell, with well developed basal lamina, pinocytotic vesicles and occasional Weibel-Palade bodies. They differ from normal endothelial cells by the abundance of intermediate filaments that fill the cytoplasm¹⁴.

Retiform haemangioendothelioma is treated by wide excision and clinical evaluation of the regional lymph nodes is necessary because this is the most common site of metastasis.

Successful palliative treatment with intraperitoneal OK-432 injection for this tumor with intractable ascites is reported¹⁵.

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