Angiokeratoma of the vulva: a rare benign vascular tumor mimicking malignancy - case reports

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Summary

Angiokeratomas are rare benign dermal lesions of the external genital system and occur before the age of 50 years. Four cases of angiokeratoma of the vulva diagnosed at our institution in a ten-year-period are reported and issues of the differential diagnosis are discussed.

Key words: Angiokeratoma; Vulva; Benign; Lesion.

Introduction

Angiokeratoma is a variant of hemangioma and occurs almost exclusively on the external genital organs, usually the scrotum and the vulva [1]. Somewhat larger than senile hemangiomas, these lesions often are purple to brownish-black in color and occur primarily in women of childbearing age. Their peculiar appearance often prompts an excisional diagnostic biopsy, although they have no clinical significance.

Histologically these lesions present dilated endothelial-lined channels, separated by strands and cords of squamous epithelial cells, representing downgrowth from the overlying epithelium. Varying degrees of hyperkeratosis, acanthosis and papillomatosis of the covering squamous cell epithelium are present, along with a mild inflammatory reaction in the deep dermis [1]. Angiosarcomas and Kaposi sarcoma are included in the differential diagnosis of these vascular tumors.

We present four cases of angiokeratoma of the vulva and issues regarding the problems of the differential diagnosis and management of this rare lesion are discussed.

Case reports

Case 1

During the routine gynecological examination of a 28-year-old woman the appearance of two small painless vulvar lesions, bluish in color, were noted. Past gynecological history was unremarkable. The physical examination of the genital system was normal. Two purple lesions measuring approximately 0.5 cm each and having a verrucous surface were seen on the right labia majora and were excised. The clinical differential diagnosis included seborrheic keratosis, nevus and basal cell carcinoma.

Case 2

During the routine pap-test screening of a 32-year-old married woman, a purple cervical lesion and a purple vulvar lesion were noted. The clinical differential diagnosis included a nevus, a malignant melanoma, or a carcinoma. Histological examination of the cervical lesion revealed a hemangioma measuring approximately 0.5 cm while the vulval lesion was a angiokeratoma measuring 0.7 cm.

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Case 3

During a routine gynecological examination of a 40-year-old married woman a purple vulvar lesion measuring approximately 0.6 cm was observed. Because of a history of endometriosis of the ovary and peritoneum the differential diagnosis of the lesion included endometriosis and carcinoma.

Case 4

A 45-year-old woman presented to the Outpatient Department with a painful, bleeding lesion of the vulva. Past gynecological history was unremarkable and the physical examination of the genital system was normal. A purple lesion measuring approximately 0.6 cm and having a papular surface was seen on the left labia majora and excised with the propable diagnosis of a melanoma.

Pathology reports

A routine histological examination of all the lesions followed, sections were stained with hematoxylin and eosin and examined by light microscopy. The histologic examination showed similar morphology. Several enlarged erythrocytes containing vascular channels were found beneath an acanthotic epidermis (Figure 1). No nuclear atypia, mitotic activity or endothelial hyperplasia was noted. They were lined by a flattened endothelium with minimal fibrous stroma. Degenerative changes in the perivascular elastic tissue were not observed. The diagnosis of vulvar angiokeratoma was made in all cases.

Discussion

Angiokeratomas of the vulva are uncommon dermal neoplasms, easily mistaken for various benign and malignant conditions of the external genital organs. Most of the patients are between 20-40 years old [2]. The lesions are frequently unilateral and located on the left side of the vulva. The majority of patients present multiple dermal lesions. Angiokeratomas are usually smaller than 1 cm in diameter, with a verrucous surface and purple color [3, 4]. Rarely the color of the lesion may be red, gray, black or brown. Vulvar angioketomas are usually asymptomatic. However, intermittent bleeding and pruritis have been described. Patients with symptomatic angioketaromas often seek medical attention sooner than those patients with asymptomatic lesions [2].

Histologically the epidermis usually demonstrates

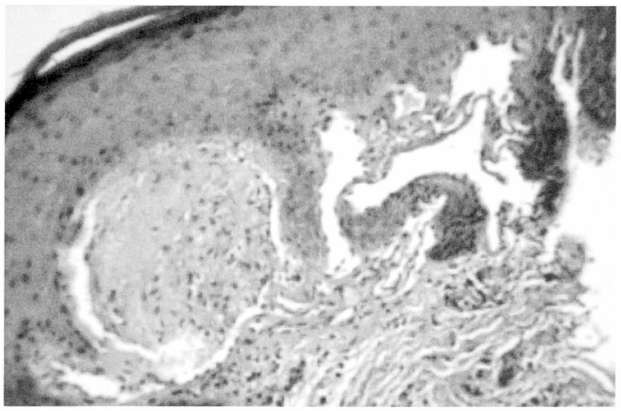

Figure 1. — Histological section of vulvar angiokeratoma: several enlarged vascular channels were found beneath an acanthotic epidermis (H&E x 250).

hyperkeratosis with occasional parakeratosis, papillomatosis and acanthosis with elongated rete ridges forming collarettes around the dilated vascular structures in the dermis [1, 2]. Within the papillary dermis dilated capillaries, venules and small veins are found. The elastic tissue around the dilated vascular structures is decreased. Thrombosis with organization and recanalization of the clot may be observed. Minimal inflammatory reaction and occasional lymphangiectasia is present in the papillary dermis. Finally there is an absence of dilated vessels in the reticular dermis.

Vulvar angiokeratomas occur principally on the labia majora [1-4] and a case has been reported with an ulcerated angiokeratoma of the clitoris [5]. The pathogenesis of the vessel enlargement observed in vulvar angiokeratomas may be the sequella of the degenerative changes occurring in the elastic tissue of these vessels; these changes could result from a primary congenital or idiopathic process or from a secondary process as a chronic inflammation or an increase of the venous pressure [2]. The majority of the patients present inflammation consistent with a secondary process. Some patients are able to correlate the presence of angiokeratoma with significant clinical symptoms as previous inguinal adenopathy, prior purulent bartholinitis or concurrent initiation of an oral contraceptive [2]. Other patients have a medical history of bleeding disorders.

The differential diagnosis of vulvar angiokeratomas includes infectious lesions (bacterial and viral), inflammatory lesions (prurigo nodularis), vascular lesions (angioma, lymphangioma, pyogenic granuloma), epithelial benign lesions (seborrheic keratosis, nevus, hidradenoma papilliferum), vulvar intraepithelial neoplasia and malignant lesions (basal cell carcinoma, squamous cell carcinoma, melanoma) [6-11]. The clinical features of these lesions may be similar and the histological examination of an excisional biopsy is necessary to confirm the diagnosis.

Seborrheic keratosis is a well-circumscribed lesion in comparison to angiokeratoma [1]. Vulvar nevi are commonly compound or intradermal; the nevus cells are located at both the dermoepidermal junction and in the dermis, and the morphology is typical microscopically [6]. Hidradenoma papilliferum is a small benign tumor of the apocrine glands occurring in the anogenital region of middle-aged women. They are firm and sharply circumscribed and present a typical morphology as well [6].

Basal cell carcinoma represent 2-5% of vulvar carcinomas [10]. Histologically the characteristic feature is the proliferation of palisading basal cells into the papillary dermis. Squamous cell carcinoma represents 90% of vulvar carcinomas that occur in older women on the anterior labia majora [10]. Malignant melanoma constitutes 5-10% of vulvar malignancies [11]. It usually occurs in older women between the sixth or seventh decade. The most common type is superficial spreading followed by nodular types. It presents a characteristic gross and microscopic morphology.

Angiosarcomas and Kaposi sarcoma are included in the differential diagnosis of vascular tumors. These malignant vascular tumors are typically cellular and present great cellular atypia, endothelial hyperplasia and mitotic activity [1]. In addition, they have fewer well-formed vascular spaces that are usually slit-like and which are infiltrative with poorly defined margins.

The therapy of choice is local excision or laser ablation although there is a possibility of blood loss due to the increased vascularity of these lesions [12].

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