Vascular tumors of the female genital tract: a clinicopathological study of nine cases

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Summary

The aim of this study was to investigate the clinicopathological features of nine unusual vascular tumors of the female genital tract and to investigate the problems in the differential diagnosis from other genital lesions.

Material-Methods: In a review of 15,000 specimens of the female genital system examined in our Laboratory during the last decade, nine cases of vascular tumors were found.

Results: A capillary hemangioma of the endometrium, one capillary and one cavernous hemangioma of the cervix, six vulva lesions (one capillary, and one cavernous hemangioma and four angiokeratomas) and one hemangiosarcoma of the mons pubis in a 76-year-old patient were diagnosed.

Conclusions: The differential diagnosis of the vascular tumors must be made mainly from endometriotic lesions and melanomas. Immunohistochemistry (S-100, CD31, CD34, CKS, EMA) aid in the diagnosis. Local excision is adequate for the benign vascular lesions.

Key words: Hemangioma; Angiosarcoma; Female genital tract; Angiokeratoma.

Introduction

Vascular tumors are common soft tissue neoplasms and comprise 7% of all human benign neoplasms. They are the most common non-epithelial tumors of children [1, 2]. About 75% of these tumors are present from birth and 85% of the total present during the first year of life. They develop most commonly in women (women/men ratio is 3/1) and are usually located at the head and neck area. There is evidence that most vascular tumors represent congenital hamartomas or tumors arising in a congenital vascular defect [1, 2].

They are usually small tumors with a tendency to grow in size as the child develops and may reach several centimeters in diameter. In many instances they cease to enlarge at maturity and eventually regress [3]. Most vascular tumors are benign and represent some form of hemangioma, capillary or cavernous. There is evidence that in many women hemangiomas develop or grow in size during pregnancy, possibly because of local changes of uterine blood-flow [4]. Despite their wide distribution in the human body, genital vascular tumors are rare lesions [5]. Angiosarcomas (AS) are a rare type of malignant vascular tumors that comprise only 1% to 2% of all soft tissue sarcomas. AS may arise from endothelial cells of the vessels of almost any internal organ. Generally they are high grade aggressive tumors that tend to recur locally and metastasize early [6-8].

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Materials and methods

In our laboratory during the last decade (1990-2001), 15,000 specimens from the female genital tract were studied. Only nine cases of vascular tumors were found; one was located in the endometrial mucosa, two at the cervix and six at the vulva. One hemangiosarcoma of the mons pubis was diagnosed as well in a woman previously treated by radiotherapy for cervical cancer. Histological sections from paraffin blocks from our archives were studied by a streptavidin-biotin immunohistochemical method for the investigation of the expression of vimentin, factor-8, S-100, HMB45, MK1, CD31, CD34, Cyto-keratins, and EMA.

Case Reports

1) An endometrial vascular lesion was diagnosed in a specimen of total hysterectomy from a 63-year-old woman with metrorrhagia and uterine leiomyomas. A purple spot, measuring 0.7 cm was observed in the endometrial mucosa and the microscopic examination showed that it was a capillary hemangioma (Figure 1). *2)* A cervical lesion was observed during a routine examination as a purple spot and the diagnosis of "endometriosis" was made. The excisional biopsy showed a capillary hemangioma measuring 0.5 cm. Another capillary hemangioma of the cervix was found, presenting as a bluish-black lesion, 0.7 cm in diameter. *3)* The vulva lesions presented as purple or black spots measuring 0.3-0.6 cm and were excised with the probable diagnosis of "melanoma" or "endometriosis". Microscopic examination showed the morphology of angiokeratoma (4 cases) (Figure 2), cavernous hemangioma (1 case) and capillary hemangioma (1 case). *4)* One hemangiosarcoma of the mons pubis in a 76-year-old patient that developed after radiotherapy for cervical cancer was diagnosed. It measured 14 x 12 cm and infiltrated the labia majora as well (Figures 3 and 4). Extensive surgical excision was performed and the patient was

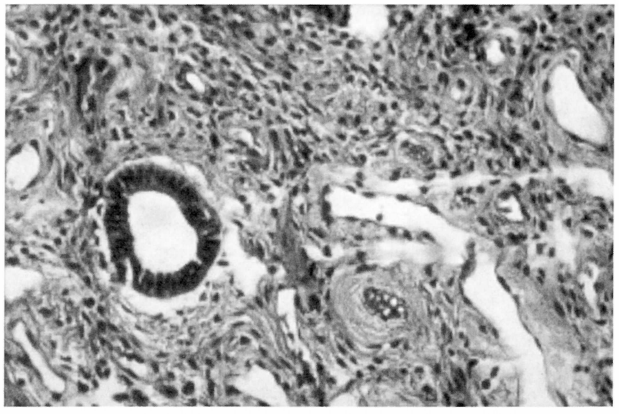

Figure 1. — Endometrial capillary hemangioma (H-E x 100).

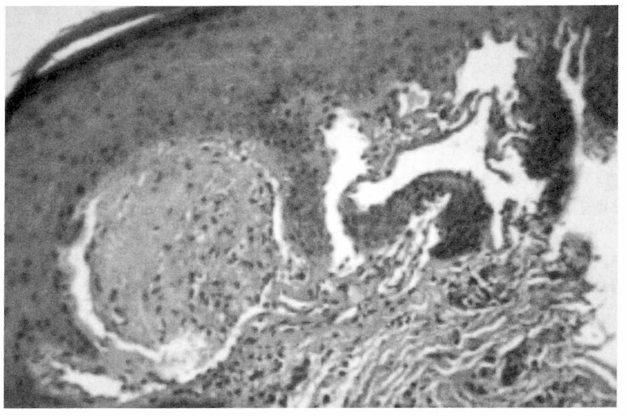

Figure 2. — Angiokeratoma of vulva (H-E x 250).

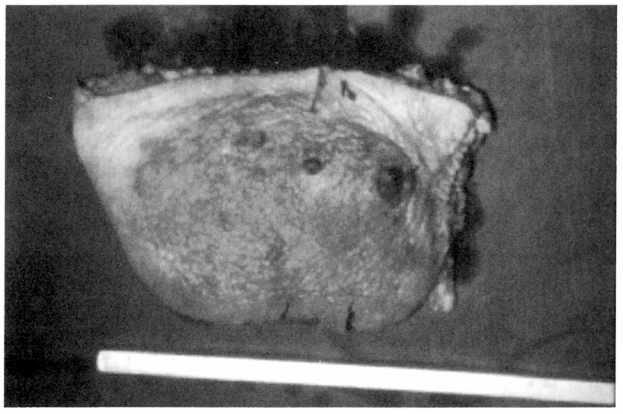

Figure 3. — Gross view of a hemangiosarcoma of the mons pubis.

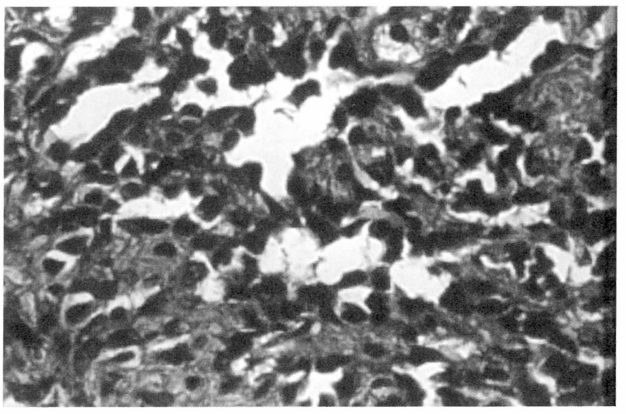

Figure 4. — Histological section of a hemangiosarcoma of the mons pubis (H-E x 400).

well a year after the diagnosis. The immunohistochemical study of all cases was positive for vimentin and factor 8 (markers of mesenchymal and vascular differentiation). The differential diagnosis of vascular tumors must be made from endometriosis and melanomas. Immunohistochemistry (S-100, HMB45, MK1, CD31, CD34, CKS, EMA) aid in the diagnosis.

Discussion

Vascular tumors present a wide distribution to the human body and they may even develop in muscular tissue; development in tissue of the female tract is rare [7] and in a series with a total of 570 reported hemangiomas only five cases were found located at the vulva [8]. Up until now there have been 20 case reports of hemangiomas of the cervix uteri [9], one case of hemangiopericytoma [10] and ten cases of ovarian hemangiomas reported [11]. There is only one case report of uterine hemangioma that developed in a pregnant woman [12] and had the morphology of an arteriovenous malformation located in the myometrium. In contrast our case developed in the mucosa and presents the typical morphology of a capillary hemangioma. More common are the vulva and perineal vascular lesions of the special type of angiokeratomas [13]. These lesions develop superficially in the skin of the area and present a typical morphology, such as in our cases. Grossly angiokeratomas may present a problem in the differential diagnosis from other lesions such as nevi, melanomas, dystrophic vulvar changes or condylomas. The histological examination solves the problem because of the typical miscroscopic features of the lesion.

Angiosarcomas occur predominantly in superficial tissue but can affect many tissues and organs. Based on a review of 366 AS from the file of AFIP's it is estimated that less than 25% of the tumors arise in soft tissues and only 7% arise in other locations [4]. Several authors have implicated radiation as a risk factor for the development of AS [14]. Over half of those cases qualifying as post-irradiation AS have occurred following radiotherapy for another malignant tumor such as carcinoma of the cervix [15], ovary [16], endometrium [17] and breast [18]. AS following radiation for genitourinary malignant tumors usually develop on the lower abdominal wall whereas those following radiation for breast carcinoma usually develop on the chest wall. The interval between the radiation and diagnosis has been approximately 12 years [14]. Fortunately, radiation-induced angiosarcoma is a very uncommon complication of radiation therapy. The overall prognosis of AS is poor, with reported 5-year survival rates of 10%-35% [19].

Conclusion

Hemangiomas of the female genital tract are rare small lesions which are usually incidental findings during routine gynecological examination. Complete excision is considered as an adequate therapeutic procedure. AS is a very rare and usually high-grade malignancy which demands extensive surgical excision.

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