A malignant eccrine poroma in a pregnant woman: case report and review of the literature

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Summary

Malignant eccrine poroma is a rare cutaneous neoplasm that originates from the intraepidermal portion of the eccrine gland. It affects mainly elderly people while its occurrence in younger adults is extremely rare. We present the first reported case of a malignant eccrine poroma in a pregnant woman, with emphasis on its pathologic and immunohistochemical features. Early diagnosis and treatment of eccrine neoplasms are of crucial importance when pregnancy coexists, because of their tendency to aggravate under the influence of gestation-related changes.

Key words: Malignant eccrine poroma; Pregnancy; Estrogen; Progesterone.

Introduction

Malignant eccrine poroma (MEP) is a rare sweat gland tumor whose incidence is estimated to be less than 0.01% of all skin biopsy specimens [1]. It usually presents as a solitary, flat or nodular lesion often with ulceration predilecting the lower extremity. It may arise either de novo or on a pre-existing benign poroma [2].

We present the clinicopathological and immunohistochemical features of a malignant eccrine poroma which was diagnosed during pregnancy. To the best of our knowledge, this is the first reported case of this rare neoplasm in a pregnant woman.

Case Report

An otherwise healthy 32-year-old pregnant woman (gestational age: 33 wks) presented to our hospital with a skin lesion located in the posterior surface of the right lower limb (thigh). The patient reported that the lesion had been present for two years, and complained of pain, itching and increased size during the last month. Physical examination revealed an elevated lesion measuring approximately 1.5 cm. The lesion was excised with wide surgical margins and sent to our laboratory for pathological examination. Microscopic examination revealed a neoplasm infiltrating the dermis and developing in continuity with the overlying epidermal surface which was extensively ulcerated. The neoplastic cells were arranged in lobules with irregular and infiltrative margins (Figure 1), and consisted of relatively small to medium sized basaloid epithelial cells with an open chromatin pattern and eosinophilic or vacuolized cytoplasm, as well as larger cells connected by inconspicuous intercellular bridges. Keratin formations were not observed. There was also lack of peripheral palisading. Nuclear atypia in general was moderate to severe. Mitotic activity was prominent, with a count of 0 to 4 mitoses per high power optical field and occasionally accompanied by atypical forms. Well differentiated ducts lined with a single layer of ductular epithelial cells and containing PAS-positive/diastase resistant secretions in the lumen were noticed within the dermal component of the tumor (Figure 2). Central necrosis was also observed. Tumor cells stained positive for cytokeratin (CAM 5.2 antibody) and vimentin, but were negative for estrogen and progesterone receptors. The luminal ductal border stained positive with antibodies to EMA. Tumor depth was approximately 6 mm and surgical margins were free of neoplastic infiltration.

On the basis of these findings the diagnosis of malignant eccrine poroma was established and close follow-up for possible local recurrence or development of distant metastatic spread was recommended. The patient gave birth to a healthy child by cesarean section and is currently well and free of disease one year after the initial diagnosis.

Discussion

Malignant eccrine poroma is a cutaneous appendage tumor arising from the intraepidermal portion of the eccrine coil (acrosyringium) [2]. Since the first description by Pinkus and Mehregan in 1963 [3], most MEP reports have been either case studies or small series, with the recent study of 69 cases by Robson *et al.* [4] being the largest series of MEP to date. Most of the initially described cases were extremely aggressive neoplasms. However, more recent data suggest that the prognosis of these tumors is better than previously believed [4, 5].

The pathological and immunohistochemical features of MEP are those of epithelial cells with dermal ductal differentiation which is nevertheless less prominent in this appendage neoplasm than in many other eccrine neoplasms [2]. The histological diagnosis of MEP is based on a combined evaluation of cytology and architecture. Architecturally, an infiltrative tumor with a lobulated architecture is present both within and closely connected to the epidermis. The tumor cells show a variable degree of ductal or squamous differentiation without keratin formations. The lumina of well-formed ducts contains a

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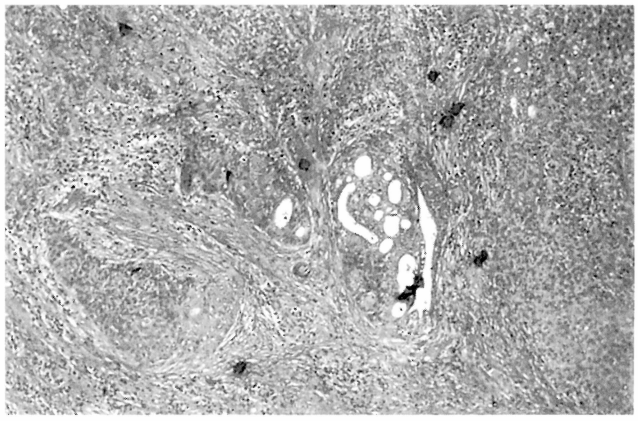
Fig. 1



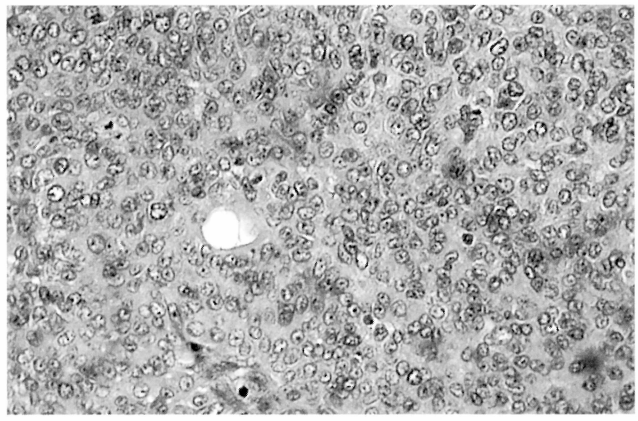


Fig.

Figure 1. — Histological section of the neoplasm showing the infiltrative margins (hematoxylin-eosin, x 100).
Figure 2. — Histological section of the neoplasm showing nuclear atypia, mitotic activity and small ducts (hematoxylin-eosin, x 400).

PAS-positive diastase resistant exudate, while the ductal border stains positive for CEA and/or EMA. Necrosis of the tumor, as well as ulceration of the overlying epidermal surface are often present and may be prominent, as in our case. Melanin is absent in tumor cells. There is also lack of peripheral palisading and of loose stroma with clefts, which aids in distinguishing MEP from basal cell carcinoma, especially when the latter exhibits features of eccrine differentiation [2]. Discrimination of MEP from its benign counterpart, eccrine poroma, is based on the presence of ill-defined infiltrative borders, individual cell necrosis, nuclear atypia and increased mitotic activity with atypical mitotic figures [2, 4]. These features, single or in combination, raise the suspicion of a biologically aggressive tumor [2], and were all present in our case.

MEP is a neoplasm affecting mainly elderly people and the mean age reported in previous series ranges between 50 and 80 years [4]. Its occurrence in younger adults is extremely rare while its sex prevalence remains controversial [4, 6, 7]. MEP has not been – to our knowledge – previously reported in a pregnant woman. Nevertheless, cases of rapidly growing eccrine poromas or other eccrine neoplasms during pregnancy have been previously described [8-10]. Pregnancy-related skin changes, including the enhancement of eccrine activity, are believed to be generally associated with the intense immunological, endocrinological, metabolic and vascular alterations that occur during that period. However, the exact mechanism relating pregnancy with alteration of eccrine activity remains partly unknown. Estrogen and progesterone receptors were negative in our case. Previous studies have suggested a possible involvement of sex steroids in the skin appendage function and the development of neoplasms, which is highly expectable due to the histogenetic similarities among the sweat gland and the breast [11]. Nevertheless, the precise distribution of hormone receptors in sweat gland neoplasms remains to be established.

In our case the growth of the histologically proven MEP was markedly increased during the last trimester of gestation. Early diagnosis and treatment of this neoplasm as well as of its benign precursor, eccrine poroma, are of crucial importance when pregnancy coexists because of the tendency of eccrine neoplasms to aggravate under the influence of gestational related changes.

References

[1] Maeda T., Mori H., Matsuo T., Nakagawa J., Yamauchi H., Tuziguchi H. *et al.*: "Malignant eccrine poroma with multiple visceral metastasis : report of a case with autopsy findings". *J. Cutan. Pathol.,* 1996, *53,* 566.
[2] Murphy G.F., Elder D.E.: "Cutaneous appendage tumors". In: Rosai J. (ed.). Non-melanocytic Tumors of the Skin, Atlas of Tumor Pathology. Washington, AFIP, 1991, 63.
[3] Pinkus H., Mehregan A.H.: "Epidermotropic eccrine carcinoma". *Arch. Dermatol.,* 1963, *88,* 557.
[4] Robson A., Greene J., Ansari N., Kim B., Seed P.T., McKee P.H. *et al.*: "Eccrine porocarcinoma (malignant eccrine poroma): a clinicopathologic study of 69 cases". *Am. J. Surg. Pathol.*, 2001, *25,* 710.
[5] Perna C., Cuevas J., Jimenez-Hefferman J.A.: "Eccrine porocaricnoma (malignant eccrine poroma)". *Am. J. Surg. Pathol.,* 2001, *26,* 272.
[6] Abenoza P., Ackerman B.A.: "Porocarcinomas". In: Abenoza P., Ackerman B.A. (eds.). Neoplasms With Eccrine Differentiation. Philadelphia, Lea & Febiger, 1990, 415.
[7] Goedde T.A., Bumpers H., Fiscella J., Rao U., Karakousis C.P.: "Eccrine porocarcinoma". *J. Surg. Oncol.*, 1994, *55,* 261.
[8] Guimera Martin-Neda F., Garcia Bustinduy M., Noda Cabrera A., Sanchez Gonzalez R., Garcia Montelongo R.: "A rapidly growing eccrine poroma in a pregnant woman". *J. Am. Acad. Dermatol.,* 2004, *50,* 124.
[9] Ban M., Kitajima T.: "A case of rapidly growing poroma during pregnancy". *J. Dermatol.*, 1997, *24,* 554.
[10] Gabrielsen T.O., Elgio K., Sommerschild H.: "Eccrine angiomatous hamartoma of the finger leading to amputation". *Clin. Exp. Dermatol.,* 1991, *16*, 44.
[11] Kariya T., Moriya T., Suzuki T., Chiba M., Ishida K., Takayama J. *et al.*: "Sex steroid hormone receptors in human skin appendage and its neoplasms". *Endocr. J.*, 2005, *52,* 317.

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