Ectopic hidradenoma papilliferum with apocrine differentiation: A case report and review of the literature

Satyaveer Kumar Mathur, Ekta Boombak, Parul Tanwar, Rajeev Sen
Department of Pathology, Post Graduate Institute of Medical Sciences, Rohtak, Haryana, India

ABSTRACT

Hidradenoma papilliferum (HP) is a benign neoplasm involving anogenital mammary-like glands and occurs most commonly in the vulva or perianal region of adult white women. It is histopathologically similar to intraductal papilloma of the breast. The lesions are usually solitary, well-demarcated papules or nodules covered with normal skin and are generally less than 10 mm in diameter. The prognosis for HP is good. When these tumors are not located in the anogenital area, they are termed ectopic HP. We present a case of ectopic hidradenoma that presented as a mass on the right arm of a 65-year-old female. To the best of our knowledge, only a single case of ectopic HP has previously been reported on the arm.

Key words: Arm, benign neoplasm, ectopic, hidradenoma

INTRODUCTION

Hidradenoma papilliferum (HP) is a benign, cystic, papillary tumor that occurs almost exclusively in women on the skin of the anogenital region. Non-anogenital (ectopic) HP is rare.[1] It is because of its histopathological similarity to intraductal papilloma of the breast that HP is postulated to arise from the anogenital mammary-like glands.[2] Rare cases of HP have been reported in males and in extragenital locations, particularly in the head and neck.[1] We describe a case of a 65-year-old female with HP presenting as a right arm mass.

CASE REPORT

A 65-year-old female presented to the hospital with a mass on the right arm. It was a slow-growing mass, present since the last 5 years. Excision biopsy was performed with a clinical suspicion of a soft tissue tumor. Grossly, the mass was fibrofatty, measuring 10 cm × 8 cm × 4.5 cm and was partially covered by the skin. On serial sectioning, a grey-white tumor area was identified measuring 4 cm in diameter. Microscopy revealed a well-circumscribed nodule in the dermis with complex architecture. The nodule consisted of arborizing papillae with a fibrovascular core along with many tubular and cystic spaces [Figure 1]. The tumor comprised of a dual population of clear cells and small pale cells [Figure 2]. Histopathological findings were consistent with HP with apocrine change.

DISCUSSION

HP is a rare benign adnexal skin tumor, originating from the apocrine glands richly found in the anogenital region, vulval, perineum, axillae and periumbilical areas.[3] The tumor primarily affects Caucasian females, typically during the third to fifth decades of life.[3] In women, anogenital and ectopic tumors have only been reported after the onset of puberty. Hence, it is possible that estrogen stimulation may play a role in its pathogenesis in women.[3] The tumors are solitary, usually small and asymptomatic. Occasionally, the tumors can be large and elevated forming reddish-brown mass with ulcerated and bleeding surface.[3] The incidence of multiple lesions is reported as 5%, and all multiple lesions have been described as unilateral.[4]
Ectopic HP is slightly more frequent in women, and the age range has been wide (8-78 years).[7] Previously reported common sites for ectopic HP include the head and neck region, extremities, upper eyelid, external ear, nose and chest wall. Of all these sites, the head and neck region is the most favored site for ectopic HP, followed by the extremities.[3] Nineteen cases of ectopic HP have been reported in the literature [Table 1].[4]

There has been a single case of ectopic hidradenoma reported in the literature on the arm. To the best of our knowledge, this the second case report of ectopic hidradenoma presenting as a mass on the arm.

On histopathology, HP represents as adenoma with apocrine differentiation. The tumor is located in the dermis, well circumscribed, surrounded by a fibrous capsule and shows no connection with the overlying epidermis. The tumor shows tubular and cystic structures with papillary folds projecting into the cystic spaces. The lumina are lined by a double layer of cells consisting of an inner layer of cuboidal to columnar secretory cells and an outer layer of myoepithelial cells. Decapitation secretion is usually seen in the secretory cells.[8] The epithelial cells typically have histochemical characteristics of apocrine origin.

HP lacks cytonuclear atypia, mitotic activity or tumor necrosis. The prognosis for both anogenital and ectopic lesions is good. Regardless of the site, the treatment of choice is local excision.[9]

Despite occasional reports on malignization of anogenital HP, all the ectopic lesions have behaved in a benign fashion and have not recurred or metastasized. Recurrence of anogenital and ectopic lesions is unusual and is typically attributed to incomplete excision of the primary tumor.[3]

Depending on the location of these tumors, the likely histopathological differential diagnoses to be considered include tubular apocrine adenoma, clear cell (apocrine) adenoma, lipoma and intraductal papilloma and intraductal carcinoma of the breast with papillary pattern.[3]

To conclude, skin adnexal tumors, specifically HP, should be kept in the differential diagnosis when evaluating a tumor mass on the arm.

**REFERENCES**

3. Abudu EK, Umanah IN, Ekpo MD, Etuk EB, Onwuezobe IA,
Obi-Ihesie A. A giant ectopic hidradenomapapilliferum in a Niger delta region of Nigeria. Rare Tumors 2011;3:e50.


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