A Rare Case of Benign Tumor Deriving from the Eccrine Glands of the Forearm

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Abstract

Background: Cutaneous adnexal tumors are a rare cluster of neoplasms deriving from the adnexal epithelial parts of the skin, including the eccrine sweat gland epithelium. They are uncommon and heterogeneous histological entities. Generally, they present as solid or cystic lesions that seldom undergo malignant transformation. Due to their rarity in routine clinical practice, they often pose a diagnostic dilemma. Management usually involves appropriate wide local resection margins of the primary tumor. Recurrence after incomplete excision is well-known, as observed in our case.

Case presentation: A patient presented here with a recurrent cutaneous adnexal tumor on the dorsum of the forearm, histopathologically proven to be a syringoepithelioma (eccrine acrospiroma), which was planned to be managed by repeat surgical excision with adequate volume.

Conclusion: Eccrine acrospiroma is a solitary benign cutaneous tumor arising from the eccrine sweat gland epithelium. Only pathological analysis will lead to the diagnosis. The surgical management approach of adequate excision is the only effective treatment to prevent recurrence and malignant transformation.

Introduction

Syringoepithelioma is a nodular, non-encapsulated, single, solid or cystic (with serous or gelatinous, and occasionally hemorrhagic content), typically elevated mass of the skin situated in the dermis. Generally, this tumor is skin-colored and appears as a sleek lesion. It varies in size from 0.5 to 10 cm [1]. Hidradenocarcinoma commonly arises de novo, although it may derive from pre-existing eccrine acrospiroma [2]. Histopathological examination of biopsied specimens from the skin is essential for making a correct diagnosis [3]. The optimal therapeutic choice is total surgical excision [4-6]. The recurrence rate of syringoepithelioma is roughly 12% if not wholly excised [7], particularly in cases with irregular peripheral margins [2]. We present a case of eccrine acrospiroma confirmed by pathological analysis. The imaging appearance is nonspecific and does not allow for a proper diagnosis without histopathological examination.

Case Report

A 74-year-old man presented with a cutaneous mass on the dorsal surface of the right forearm that had been present for the last 6 years (Fig. 1). One year previously, a tumor with similar macroscopic features had been excised from the same location at another institution and histopathologically diagnosed as a benign eccrine acrospiroma. The recurrent soft tissue mass of the forearm was not associated with any clinical symptoms. There was no relevant history of trauma. Status localis revealed a 10×5 cm, solitary, irregular, lobular, painless soft tissue mass arising from the mid dorsal right forearm with overlying skin changes. The mass was slightly mobile and could be displaced laterally by palpation. The lesion appeared bluish to pinkish in color and was prominently raised without superficial areas of serous discharge or ulceration. Except for the described lesion, the status praesens objectivus showed no abnormalities. Vital signs and laboratory tests were unremarkable. Fine needle aspiration cytology from the soft tissue mass was reported as a benign cystic tumor.
A computed tomography (CT) study of the right forearm was performed. The CT analysis demonstrated no pathological changes in the forearm bones. A mixed cystic and solid appearance of the mass was discovered, with numerous variable-sized heterogeneous cysts of round or oval shape with local incomplete septa. The maximum size was 3×4 cm. The heterogeneous density of the cystic fluid content was noted, with iso- and predominantly hypodense substrate having density readings of about 43-48 HU and 16-26 HU, respectively (Fig. 2a). Solid mural components were present. Following the findings from the CT scan, a contrast-enhanced magnetic resonance imaging (MR) scan for further characterization of the soft tissue mass was planned.

A non-contrast-enhanced magnetic resonance study of the right forearm was performed (the patient refused administration of the gadolinium-based contrast agent for subjective reasons). Similar data were revealed as described earlier on CT. T1, T1-FatSat, T2, T2*-gradient-echo (GRE) weighted images, and DWI with ADC mapping in axial planes, as well as coronal proton density (PD) sequences, and sagittal T2-weighted non-enhanced sequences were obtained (Fig. 2b-f). The cystic component demonstrated heterogeneous T1 and T2 signals, with no signal loss on the fat-suppressed sequences. The heterogeneity of the signals is due to the different composition of the cystic fluid, and there is a layered sedimentation sign of the hemorrhagic component. The mural component showed low intensity on T2- and T1-weighted images. No diffusion restriction was found.

Following the data obtained, a preliminary diagnosis of “recurrent benign neoplasm of the skin of the right upper limb” (D23.6) was made, leading to the decision to perform an ultrasound-assisted trephine biopsy followed by histopathological analysis.

Pathological analysis was performed, with no immunohistochemical study carried out. A diagnosis of syringoepithelioma (eccrine acrospiroma) was made. Considering the patient’s age, comorbidity, and the absence of malignant transformation over the last 6 years, it was agreed upon to undergo periodic clinical follow-up.

**Discussion**

In the literature, syringoepithelioma has been reported under various names such as eccrine acrospiroma, clear cell acrospiroma, ductal epithelioma, solid syringoma, porosiryngoma, myoepithelioma of the sweat gland, clear cell epithelioma, clear cell hidradenoma,
superficial hidradenoma, solid-cystic hidradenoma, and nodular hidradenoma [8,9]. The abundance of terms indicates the diversity of the morphological structure of this tumor and reflects different views on its histogenesis [9]. This type of tumor is found in all cutaneous sites, but predominantly in the head, upper extremities, and trunk. Generally, lesions contain mixed solid and cystic components and range in color from reddish-blue to tan [1]. In the literature, the MRI appearance of eccrine acrospiroma is characterized according to the heterogeneity of this tumor group, which contains variable numbers of functioning sweat gland apparatus. The diversity of the T1 and T2 signals of the cystic content is perhaps due to the amounts of hemorrhage, cholesterol, and sweat gland secretions represented in the fluid. The mural component, noticed in our case, is a typical feature of this type of neoplasm. Histopathologically, it can be presented as papillary configuration of epithelial and clear cells with prominent central vessels or a sclerotic stroma, which can explain MRI contrast enhancement or the lack thereof. The ultrasound appearance correlates well with the MRI characteristics, generally showing a mainly cystic mass comprising complex fluid and a mural component with duplex ultrasound features correlating with the MRI enhancement pattern [6]. CT, in this case, is less informative than MR imaging and duplex ultrasonography. Histological analysis remains the gold standard for diagnosing eccrine acrospiroma [3]. Rarely, benign neoplasms can undergo malignant transformation. It is known that hidradenocarcinoma arises de novo or after malignant transformation from pre-existing syringoepithelioma in up to 7% of all cases [2,10]. Malignant neoplasms show more aggressive behavior [11]. In the absence of evident invasion into adjacent structures, no unique imaging characteristics appear to relate to the potential for malignant transformation, and the resected specimen must be histopathologically analyzed [6]. The gold standard for managing syringoepithelioma is wide local excision with at least 1 cm skin margin because less excision range will cause recurrence [4-7,12]. Recurrent masses are usually no more aggressive or atypical than primary lesions [1]. The prognosis of benign hidradenoma is good, and the recurrence rate after wide excision is nil [13].

Conclusion

Hidradenoma is a solitary benign soft tissue mass arising from eccrine sweat gland epithelium. The macroscopic features and radiologic appearance are not specific. Only careful histopathological examination will lead to the diagnosis. The surgical management approach of complete excision is the only effective treatment to prevent recurrence and malignant transformation. Subsequent regular follow-up is required due to the rare potential for malignant transformation.

Competing Interests

The authors declare that they have no personal or financial relationships that may have inappropriately influenced them in writing this article.

Author’s Contributions

Burko P. was the primary author. Juggath N. supervised and contributed equally to the final version of the manuscript.

Ethical Considerations

This article followed all ethical standards for research.

Funding Information

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Funding Information

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

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