CASE REPORT

Eccrine spiradenoma: a rare adnexal tumour with atypical presentation: A case report

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Abstract

First described in 1934, eccrine spiradenoma (ES) is a rare, benign adnexal tumour arising from eccrine sweat glands. It commonly presents as a slow-growing nodule on the upper trunk, and head and neck region, mostly in the age bracket of 15-35 years, with no gender preference. While no established guidelines exist for optimal management of malignant ES, some therapies have been studied. The diagnosis of this entity is extremely important as it can harbour a malignant component with disastrous outcomes which may be missed due to its strong resemblance to benign lesions, such as a papilloma. Here, we present the case of a 35-year-old lady who presented with a papilloma-like growth on the upper medial aspect of the thigh which was diagnosed as eccrine spiradenoma upon excision.

Keywords: Eccrine Spiradenoma, adnexal tumour, dermal lesion, case report

DOI: https://doi.org/10.47391/JPMA.1199

Introduction

Eccrine spiradenoma (ES) is a rare, benign, adnexal tumour arising from the intradermal duct of the eccrine sweat glands, first described in 1934 by Sutton and later by Kersting et al in 1956.[1] It commonly presents as a painful, nodular, slow-growing mass on the upper trunk, and head and neck region, usually in the age bracket of 15-35 years, but can occur anytime during life without any gender or racial predilection.[2] Morphologically, it may resemble various other dermal lesions such as leiomyoma, dermatofibroma, angiolipoma, glomus tumour, papilloma and neurofibroma, hence a biopsy is essential for the correct identification of this rare tumour.[3] Generally, it is asymptomatic and solitary in most cases; however, approximately 22 cases of multiple ES have been reported in literature.[3,4] They may also occur as part of Brooke-Spiegler syndrome, an inherited autosomal-dominant disease caused by CYLD mutations on chromosome 16q, consisting of multiple ES, along with trichoepitheliomas, cylindromas, spiradenocylindroma and parotid tumours.[2,4]

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It is important to note that a malignant version i.e. eccrine spiradenocarcinoma also exists with metastasis rate of 50% and mortality rate of 37%, making the diagnosis of ES essential for its recognition.[2,5] Since it may mimic various benign dermal lesions, its excision and diagnosis can easily be delayed and missed. We herein describe a case of ES which presented as a papilloma in a 37-year-old female. Owing to the paucity of data, no guidelines for optimal management and follow-up of ES currently exists.

Case Report

A 37-year-old obese woman, with no comorbidity, presented to the general surgery clinic at the Aga Khan Diagnostic Centre, Garden East, in November 2018 with a swelling on her left upper inner thigh for the last one year which was unchanged since its appearance. The reason for her consultation was increased discomfort in the lesion, especially on walking, which was caused by rubbing of her medial surface of thighs. On examination, the lesion was 2x1cm, pedunculated, soft, non-tender and resembled a papilloma. The overlying skin was unremarkable and it did not appear to be infected (Figure 1). Due to her discomfort, she underwent surgical excision of the dermal lesion at the same centre under local anaesthesia. Differential diagnosis included a papilloma, dermatofibroma and neurofibroma. Intraoperatively, the mass was soft, fleshy, with a stalk and

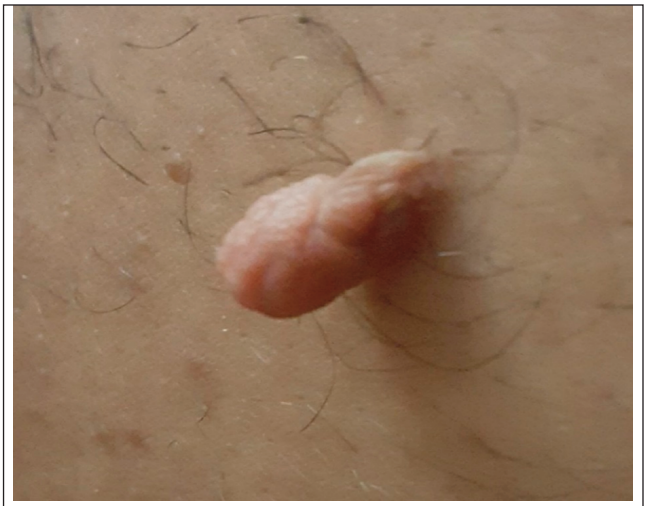

Figure-1: Gross appearance of pedunculated dermal lesion on the left upper inner thigh, clinically resembling a papilloma.

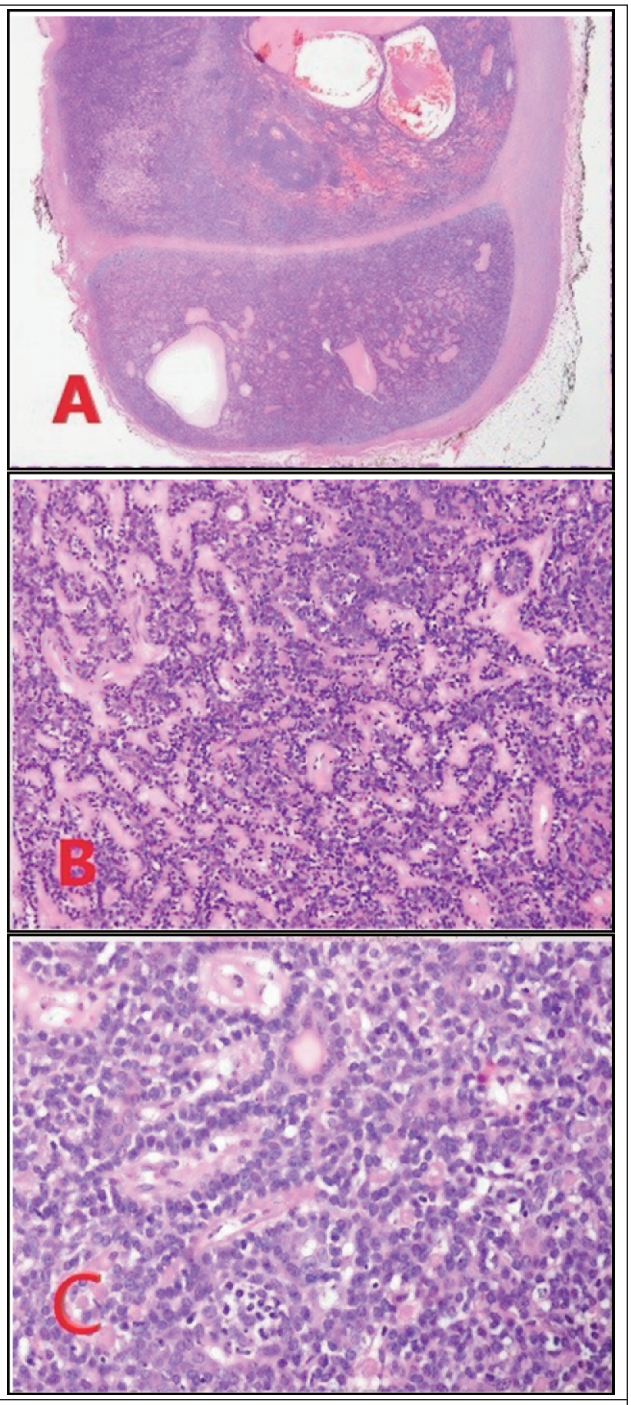

Figure-1: Gross appearance of pedunculated dermal lesion on the left upper inner thigh, clinically resembling a papilloma.

narrow base. The specimen was sent to the laboratory for histopathological examination. On gross examination, it was grey-white, firm, circumscribed and nodular. Cut surface showed a central dark brown area. Microscopically, the tumour was composed of closely opposed nodules surrounded by a thick fibrous capsule and contained cystic spaces of variable sizes. Also seen were interconnected trabeculae and compressed tubules which were separated by fibrous septae. Within these septae, deposition of basal lamina-like material was observed. Tumour cell population predominantly comprised of basaloid cells with scant cytoplasm and oval cells with finely dispersed chromatin. Scattered larger cells with clear cytoplasm were also present along with focal sprinkling of lymphocytes. A diagnosis of ES was established (Figure 2).

Due to limited cases of ES reported in literature, no standard guidelines for management and follow-up of benign ES exist. Our patient's post-operative course was unremarkable and no recurrence was noted at one year follow-up. She has been advised for a follow-up in case a similar lesion at the same or any other area appears.

Discussion

ES is a benign adnexal tumour commonly presenting as a small, bluish-pink to grey subcutaneous nodule, usually on the upper half of the body, typically accompanied with tenderness.[2] Our case was unusual in a number of ways. Firstly, it presented on the thigh, instead of the more common locations, such as the head, neck and upper trunk. In addition, the tumour was non-tender and had a pedunculated, papilloma-like configuration as opposed to usual nodular configuration. Clinically, ES may resemble a papilloma, neurofibroma, dermatofibroma, leiomyoma, angiolipoma and glomus tumour.[3] Histologically, papilloma exhibits acanthotic epidermis while the dermis is unremarkable. Neurofibroma, dermatofibroma, leiomyoma, angiolipoma and glomus tumour are mesenchymal tumours and show proliferation of neural cells, fibrohistiocytic cells, smooth muscle cells, adipocytes and pericytic cells respectively.[4,6] In contrast, benign proliferation of epithelial cells was seen in our case. Other benign adnexal tumours such as nodular hidradenoma and cylindroma are also included in the differential diagnosis. But typical arrangement of cells in trabeculae and cords with deposition of basal lamina-like material and sprinkling of lymphocytes were useful clues in reaching the diagnosis of ES.[4,6] Eccrine spiradenocarcinoma is another important differential which needs to be identified early, if present. Microscopically, the cells in eccrine spiradenocarcinoma show significant nuclear pleomorphism, frequent mitoses, lack circumscription and show destructive infiltration into adjacent tissue.[6] These features were not observed in our case.

While usually solitary, they can rarely present as multiple lesions and so far 22 cases of multiple ES have been reported in literature.[3] They may also occur as part of Brooke-Spiegler syndrome, consisting of multiple ES along

with multiple trichoepitheliomas, cylindromas, and parotid tumours.[4]

Histopathologically, ES is composed of intradermal lobules surrounded by a fibrous capsule without connections to the epidermis. The epithelial cells within the tumour lobule are arranged in intertwining cords. Two types of epithelial cells are present in the cords; the first type has small, dark nuclei located at the periphery of the cellular aggregates, and the second has large, pale nuclei arranged around a small lumen as were seen in our patient.[7]

The recognition of this entity is important because of the potential occurrence of its malignant version, i.e. eccrine spiradenocarcinoma. Rare, malignant transformation is possible, manifesting as sudden rapid growth, accompanied by erythema, increased pain, ulceration, necrosis, and appearance of satellite nodules.[8] The latency period before malignant transformation can range from six months to 70 years.[8] Apart from this transformation, de-novo appearance of malignant ES can also occur.[7] This can lead to devastating consequences, since these have metastases in 40-50% of the cases.[9] The most common metastatic sites include lymph nodes, lung, brain, liver, and bone.[9] This neoplasm is also notorious for a high rate of recurrence — up to 57% — which warrants wide surgical excision along with close follow-ups.[8,9]

Uptil now, a total of 102 cases of ES have been reported in the literature.[5] If encountered, it is recommended that both the benign and malignant forms of the lesion should undergo complete excision and followed closely for a recurrence in malignant cases.[5] A meta-analysis of the malignant cases also cautions regarding the potential for lymphatic spread, and the need for surgical clearance of the draining lymph nodes in suspected metastatic cases.[8] Radiation and hyperthermic chemotherapy have also been used as a measure for preventing local recurrence with very limited data.[5] Further, there are reports of ES being treated by CO_2 laser, intralesional botulinum toxin, and intralesional steroids.[10] Although an excision of a benign-appearing dermal and subcutaneous lesion may not be warranted straightaway, it is important to keep a close eye and warn the patient in case the tumour starts to exhibit signs of aggression, in which case a diagnosis of ES should be kept in mind so as to plan timely management. This is especially important since clinicians and patients may not always be keen on removing a harmless papilloma or inclusion cyst, which may be the topmost diagnoses in such scenarios.

Conclusion

Eccrine Spiradenoma is a rare entity which may be confused with other dermal lesions clinically, especially in cases with atypical presentation. It should always be considered in the differential diagnosis of all dermal lesions including papilloma. Careful histological evaluation and accurate diagnosis is essential for appropriate management and for exclusion of its malignant variant.

Consent Form: Informed consent was taken from the patient on phone and signed by her husband for publishing the case as the patient was out of city.

Disclaimer: None.

Conflict of interest: None.

Funding disclosure: None.

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