Eccrine Spiradenoma – A painful dermal nodule

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Abstract

Eccrine spiradenoma is an uncommon benign adnexal neoplasm occuring mainly in young adults. Here we report a case of eccrine spiradenoma with unusual presentation and concise review for the dermatopathologist.

Introduction

Eccrine spiradenoma also known as spiradenoma is an uncommon, well differentiated benign tumour historically designated as a tumour of eccrine differentiation, although current reconsideration indicates an apocrine process. It is one of the ten painful tumours of skin with characteristic histopathological findings.

Case Report

A 35 year-old woman presented with history of swelling in the lower part of posterior triangle of neck over right side since many years but from 6 months the swelling slightly increased in size with pain. Examination of the lesion revealed a tender, skin coloured, firm swelling in the subcutaneous plane ranging in size from 1.5 – 2.0 cms. So, initially a differential diagnosis of cervical lymphadenitis or angiolipoma was made. Routine haematological and biochemical investigations were normal.The patient underwent surgical excision under local anaesthesia and tissue was sent for histopathological examination. Histological findings demonstrated sharply delineated dermal nodules that display small rosettes and interwining cords (Figure 1). In higher magnification the tumor nodules were comprising of two cell types. First type being small, dark, basaloid cells with hyperchromatic nuclei at the periphery of the lesions and second type cells with large, pale vesicular nuclei located at the central area (Figure 2). Also, basement membrane like hyaline material was present in the stroma or within the cords along with lymphocytic infiltrate. PAS stain revealed eosinophilic PAS positive diastase resistant material in the lumen. So, the diagnosis of eccrine spiradenoma was prompted.

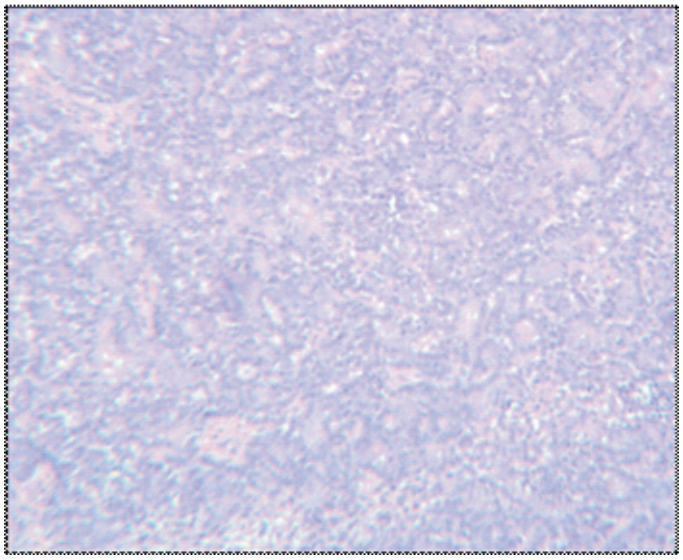

Figure 1. Tumor cells arranged in small rosettes with numerous lumen (H and E 200)X

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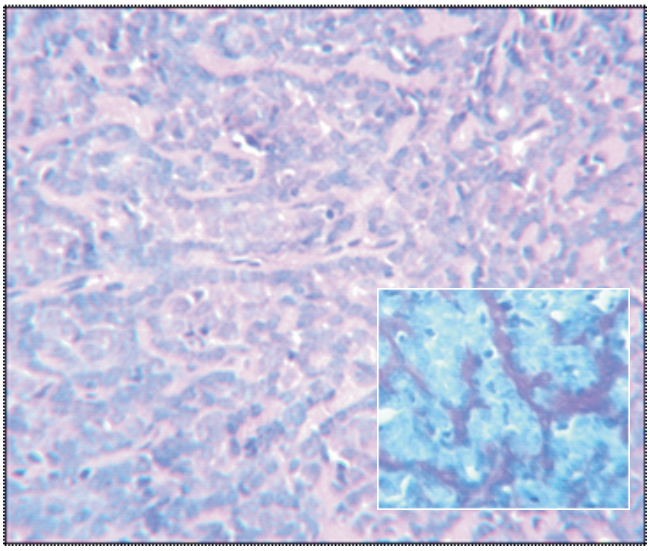

Figure 2. High powered view demonstrating two types of cells -small,dark, basaloid and larger cells with pale nuclei. Inset shows PAS positive material within the luminia (H and E 400 X)

Discussion

Eccrine spiradenoma is usually a rare solitary benign tumour; however, occasionally multiple lesions may be present that exhibit a linear or zosteriform pattern [1]. Chandeluz, in 1882, probably first described this tumour. Unna first coined the term spiradenoma. In 1956 Kersting and Helwig published the classic paper on spiradenoma in 136 patients [2]. Exact incidence is not known. Histologically,it consists of one or more large, sharply defined basophilic nodules in the dermis, also described as 'cannon balls' or 'big blue balls' in the dermis. It can occur in infancy but most commonly arises in persons aged 15-35 years. They usually occur in ventral aspect of trunk and proximal limbs [3]. Most lesions are found to be tender. The exact mechanism of pain is not clear [4]. The painful tumors of skin are limited and are remembered by acronym BLEND AN EGG which includes blue rubber bleb naevus, leiomyoma, eccrine spiradenoma, neuroma, dermatofibroma, angiolipoma, neurilemmoma, endometrioma, glomangioma, granular cell tumour. In Brooke-Spiegler syndrome, of which spiradenoma are a manifestation, the defective gene is the CYLD gene on chromosome. The CYLD seems to be a hot spot of mutations as novel mutations continue to be reported [5]. Meybehm and Fischer noted that spiradenomas and cylindromas express S-100 protien,ascribed to eccrine differentiation within their tubular and large, pale staining cells [6]. The tumour cells also express cytokeratins and the tubular structures are CEA positive [2]. The expression of p53 in malignant spiradenomas seems to be increased. Malignant transformation is rare and occurs more often in cases of multiple lesions than in solitary cases [7]. Correct diagnosis is critical due to the potential for malignancy. Treatment is surgical excision. Recurrences have been reported.

References

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