

CASE REPORT

Multiple Eccrine Spiradenoma- An Unusual Clinical Presentation

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Abstract:

Eccrine spiradenoma is an uncommon benign adnexal tumour of apocrine differentiation. It is commonly seen in young adults with no sex predilection on upper ventral aspect of body as a solitary papule or nodule. Here we are reporting a rare case of Multiple Eccrine Spiradenoma (MES) in a 29 year male patient, who presented with multiple nodules on the anterior chest wall.

Keywords: Benign, Adnexal, Nodule

Introduction:

Eccrine spiradenoma is a benign adnexal tumour with eccrine differentiation. According to the recent concept this dermal tumour is considered to originate from cells which show apocrine and thiecoepitheliomatous differentiation that is folliculosebaceous apocrine complex hair follicle differentiation [1, 2]. Most of spiradenomas arise in 15-35 years of age group with equal sex distribution [1-3]. These lesions usually occur on anterior aspect of upper half of the body as a solitary grey, pink to bluish nodules. Presentation of spiradenoma as a multiple nodule is rare [4]. To the best of our knowledge only few cases of multiple spiradenomas have been reported in literature.

Although the exact etiology of these benign lesions is not known, however some authors opine that these tumours develop from defect in the tumour suppressor gene. Spiradenoma of Brooke-Spiegler Syndrome (BSS) is associated with mutation in gene CYLD1 located on chromosome 16 at position 12.1 [1-3]. Malignant transformation and

metastasis to distant organs of eccrine spiradenoma is very rare [4]. Due to rarity of multiple eccrine spiradenoma in a linear distribution, we present this case for awareness of unusual clinical presentation and review of literature.

Case Report:

A 29 year old male patient presented with multiple swellings over anterior chest wall since four years which were gradually progressing and associated with mild pain. There was no significant co morbid illness, past history and family history. On examination these swellings were multiple papulo nodular, soft in consistency, non tender and varying in sizes from 0.5 to 1 cm.

Surgically excised mass was sent for histopathological examination. Specimen showed single skin covered fibro fatty tissue mass measuring 5 x 1.5 x 1 cm with skin flap measuring 5 x 1.5 cm. Fibro fatty tissue showed five nodules of varying sizes, larger measuring 1 x 1 cm, and smaller measuring 0.5 x 0.5 cm. Cut section of nodule were pale yellow, homogenous (Fig. 1).

Multiple sections studied showed structure of skin comprised of epidermis and dermis. Epidermis appeared unremarkable. Dermis showed multiple well circumscribed encapsulated tumour nodules which were not connected to the epidermis (Fig. 2). Within the nodule, tumour cells were arranged in nests, cords and sheets. Two types of cells were noted, peripheral cells were round to polygonal with round to oval condensed nucleus and scant

eosinophilic cytoplasm. Centrally arranged cells were larger round to oval with round to oval vesicular nucleus with prominent nucleoli and scant to moderate amount of eosinophilic cytoplasm. At foci, cystically dilated spaces filled with eosinophilic proteinaceous material were seen. Also seen many scattered lymphocytes throughout the lesion (Fig. 3). Based on these histological features diagnosis of benign adnexal tumour—multiple eccrine spiradenoma was rendered.



Fig.1: Skin Covered Fibrofatty Tissue with Varying Sized Nodules

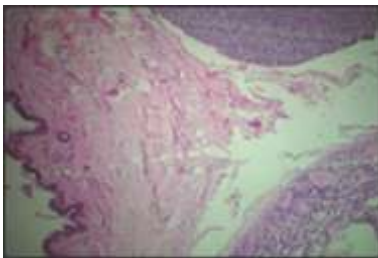


Fig.2: Photomicrograph Shows Epidermis and Dermis. Dermis Showed Two Well Circumscribed Encapsulated Tumour Nodules Which Were Not Connected to the Epidermis. (H&E 100X)

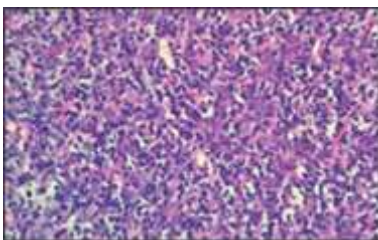


Fig.3: Microphotograph shows Two Distinct Types of Cells. (H&E 400X)

Discussion:

Eccrine spiradenoma classically presents in 2nd to 3rd decade of life usually occurs on trunk and extremities with soft to firm, round to ovoid shaped, blue-red dermal or subcutaneous nodules ranging from 0.5 to 5cms in diameter. These eccrine spiradenoma can occur as a single or multiple lesions. In some cases these multiple lesions present in a linear or zosteriform distribution [5-7]. To the best of our knowledge only few cases of multiple spiradenomas have been reported in literature. In present case nodules were of varying size from 0.5 to 1cm and presentation was in linear distribution.

Exact etiology of eccrine spiradenoma still remains controversial. The literature shows, MES is due to an abnormal clone of multipotent stem cells of the folliculosebaceous apocrine unit arises during embryogenesis, subsequently producing proliferation of abnormal cells resulting in nodule formation and possible malignant transformation. In our case, one year of follow up examination showed no evidence of recurrence.

BSS is an uncommon autosomal dominant disorder, which is characterized by multiple adnexal neoplasms specially trichoepitheliomas, cylindromas and spiradenomas [5]. It is caused by mutations in *CYLD1* gene on chromosome 16q12q13. However, mutations in *CYCD* gene are also found in familial cylindromas and familial trichoepithelioma. When patient presents with multiple adnexal neoplasm histopathology play an important role in distinguishing between BSS, familial cylindroma, MFT [3-5].

The eccrine spiradenoma shows many characteristics histopathological features which include sharply circumscribed basophilic nodules in dermis surrounded by fibrous capsule. Basaloid cells composed of two distinct morphologies,

small cell type with condensed dark nucleus located at the periphery of the nodules and centrally arranged large cells with vesicular nucleus with prominent nucleolus with scant cytoplasm and intervening tissue shows lymphocytic infiltration throughout the nodule [2, 3, 6-8]. In our case multiple nodules in linear distribution were present on the anterior chest wall and also displayed all these distinctive histological features with two types of cells in the tumour nodule and scant lymphocytic infiltrate in the intervening stroma.

Eccrine spiradenoma may be easily mistaken for cylindroma. The diagnosis can be easily done histologically. As a general rule, cylindroma tend to present on head and neck and not painful. However, eccrine spiradenomas present on trunk and extremities are generally painful. Histologically, cylindroma is differentiated by presence of basaloid cells arranged in islands, like a jigsaw puzzle and PAS positive densely eosinophilic material around tumour islands and absence of scattered lymphocytes which are often present in eccrine Spiradenoma [3, 5, 7].

The literature revealed that immuno histo-chemical staining of these eccrine spiradenoma shows positive immunoreactions for cytokeratin and S100 [3, 5-7].

Conclusion:

Eccrine spiradenoma is a dermal tumour of sweat gland; etiology is genetic which requires further investigations. Diagnosis is of paramount importance because of potential malignant transformation, particularly in the case of multiple, symptomatic and in long standing cases. Hence, follow up of these patients presenting with eccrine spiradenoma should be done. The correct diagnosis can be achieved with histological criteria when combined with pertinent clinical information.

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