Case Report

ERA'S JOURNAL OF MEDICAL RESEARCH

VOL.9 NO.1

DOI:10.24041/ejmr2022.19

ECCRINE SPIRADENOMA : CASE REPORT WITH AN UNUSUAL CLININAL PRESENTATION

Silky Rai, Nishi Tandon, Reeta Chaudhary, Nirupma Lal, Osman Musa Hingora

*Department of Pathology, Department of Surgery**

Era's Lucknow Medical College & Hospital, Era University, Sarfarazganj Lucknow, U.P., India-226003*

Received on : 26-02-2022
Accepted on : 22-04-2022

Address for correspondence
Dr. Nishi Tandon
Department of Pathology
Era's Lucknow Medical College & Hospital, Era University, Lucknow,-226003
Email: drnishitandon@gmail.com
Contact no: +91-9452296953

ABSTRACT

Eccrine spiradenoma is one of the rare, benign dermal neoplasm showing eccrine sweat gland differentiation. The literature search accounts for less than 100 reported cases of eccrine spiradenoma. Clinically, the lesion presents as solitary, intradermal, circumscribed, round or oval, firm, painful or tender nodule measuring from 0.3 - 5 cm in diameter. Rarely if ES, presents as multiple lesions, in combination with other types of adnexal tumors such as cylindromas & trichoepitheliomas, it can be considered as a part of the Brooke-Spiegler syndrome. Majority of these tumours appear on the head/face & upper trunk region of the body. Spiradenomas arise in early adulthood in most instances with few reports also of congenital cases and no sex predilection seen. The definitive diagnosis is made by excisional biopsy of the tumour. Complete tumor excision with clear surgical margins is considered the primary treatment for these cases. Here, we present a rare case report of a 58 years old male presenting with a soft, cystic lesion in the left lumbar region of the abdomen for past 2 years duration. Surgical excision of the tumour was done with clear margins and the specimen was sent for histopathological examination. Due to the overlapping histomorphological features, this tumour is often confused with cylindromas and trichoepitheliomas along with other vascular tumours. Eccrine Spiradenoma, although a benign tumour, but malignant transformation has been described especially in long-standing cases or multiple lesions. So, early diagnosis is very important to prevent recurrence and to identify any onset of malignant transformation.

KEYWORDS: Eccrine Spiradenoma, benign adnexal tumour, malignant transformation, Brooke-Spiegler syndrome, genetic testing.

INTRODUCTION

Eccrine spiradenoma (ES) is a rare, benign dermal neoplasm showing eccrine differentiation with first paper publication by Kersting and Helwig in 1956 (1,2). Clinically, the lesion presents as solitary, intradermal, circumscribed, round or oval, firm, painful or tender nodule measuring from 0.3 - 5 cm in diameter (3). Rarely if ES, present as multiple lesions, in combination with other types of adnexal tumors such as cylindromas & trichoepitheliomas, can be considered as a part of the Brooke-Spiegler syndrome, a rare autosomal dominant disease, having mutation of CYLD (Cylindromatosis) gene. Majority of these tumours appear on the head/face & upper trunk region of the body (1). Spiradenoma arise in early adulthood in most instances although there are reports of congenital cases too (4). No gender predilection has been reported (5). The mainstay for ES is tumor excision with clear surgical margins, however, literature documents many cases of recurrence along with its ability in long standing cases to undergo malignant transformation. Therefore, it becomes majorly important to make an early definitive diagnosis of ES by excisional biopsy. Being a painful lesion showing florid vascularisation on histology, ES can be easily mistaken for glomus lesions or angioleiomyoma (6). Hence, we report a noteworthy case of ES in a 58-year-old-male with focus upon its unusual clinical presentation followed by diagnosis made solely on the basis of histopathological examination of the excised lesion.

CASE REPORT

CLINICAL HISTORY A 58 year old male presented to the surgery OPD (Out Patient Department) of Era's Lucknow Medical College and Hospital, Lucknow, U.P., with the chief complaints of painless swelling in left lumbar region of the abdomen for the past 2 years. The swelling gradually increased in size over this period with a

history of watery discharge observed around 1.5 months back which stopped spontaneously. No history of pruritis, increased pigmentation or any other similar lesion was noted. Family history and medical history were insignificant. Local examination of the lesion revealed single, soft, globular swelling of size 2x2 cm, with smooth surface and round margins (Fig. 1). Routine blood investigations were normal. Clinically, the differential diagnosis of Sebaceous cyst and Lipoma were considered. Lesion was surgically excised and the specimen was sent to the histopathology laboratory for examination.

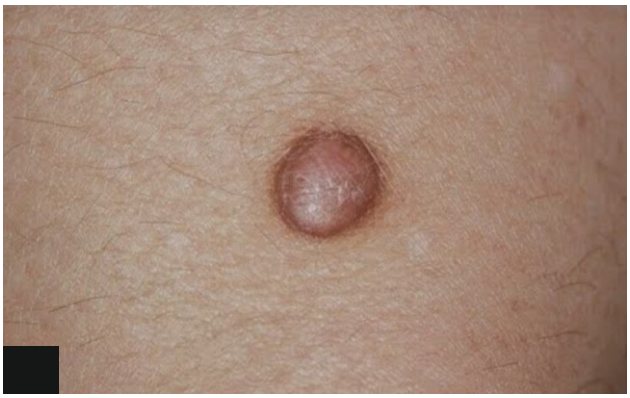

Fig. 1: Clinical image showing a 2x2 cm soft, globular, painless lesion present in the left lumbar region of abdomen.

GROSSLY

The specimen was solitary, gray-white to gray-brown, nodular, skin covered soft tissue piece measuring 2.0 x 1.5 x 1.0 cm. Cut section showed gray-white, solid areas. Routine tissue processing and H&E staining as per the protocol standardized in our histopathology laboratory was performed.

MICROSCOPICALLY

H&E (Haematoxylin and Eosin) stained sections showed an intradermal well-circumscribed, uninodular, basophilic tumor with fibrous capsule (blue ball) (Fig. 2). The overlying epidermis was intact with no connections to the tumor island. The neoplastic nodule was composed of dual cell population arranged in intertwining cords fashion. These tumor cells seen in the centre had large, pale nuclei and scant cytoplasm while tumor cells seen at the periphery had small, dark nuclei with scant cytoplasm (Fig. 3). Also seen were tumor cells surrounding the eosinophilic basement membrane material which was PAS (Periodic Acid-Schiff) positive (Fig. 4a) & Diastase resistant (Fig. 4b). The stroma between the cords of epithelial cells was seen to be focally edematous and hyalinized. Intratumoral lymphocytes along with widely dilated vascular channels were noted. However, no atypical mitosis was observed. There was no evidence of granuloma formation or any malignant focus seen in the tissue material examined. Based on the above histomorphological findings, final diagnosis of Eccrine Spiradenoma was made.

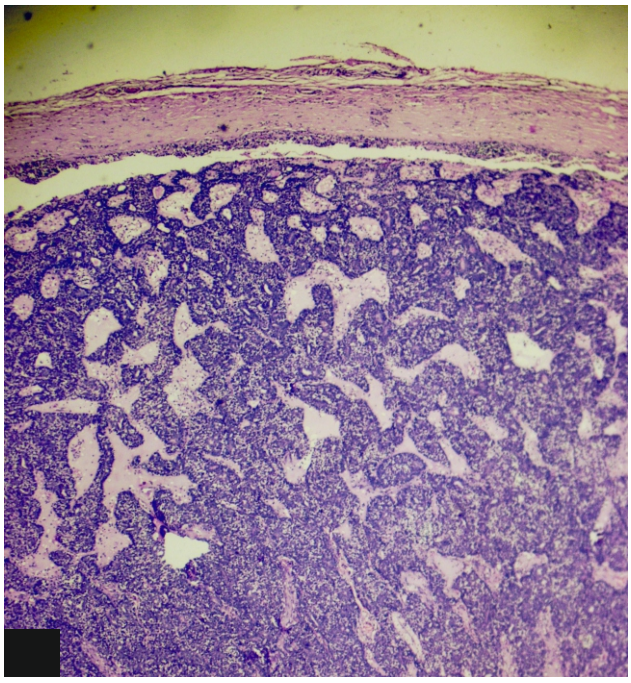

Fig. 2: (H&E, 4X) Histological examination revealed solitary strongly basophilic lobule of tumor cells in the dermis. The nodule was well-circumscribed & encapsulated.

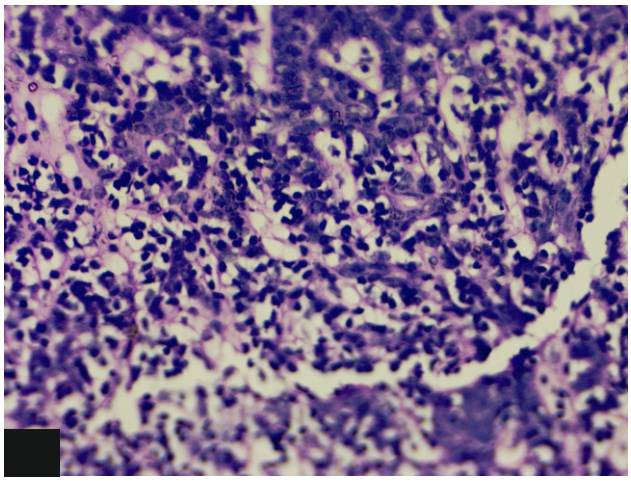

Fig. 3: (H&E, 40X) The biphasic neoplastic epithelial cell population (dark and pale) was evident in the tumor lobule. Dark cells were small, basaloid cells with hyperchromatic nuclei & scant cytoplasm located at the periphery whereas pale cells were larger with vesicular nuclei & ample pale cytoplasm situated mainly in the centre.

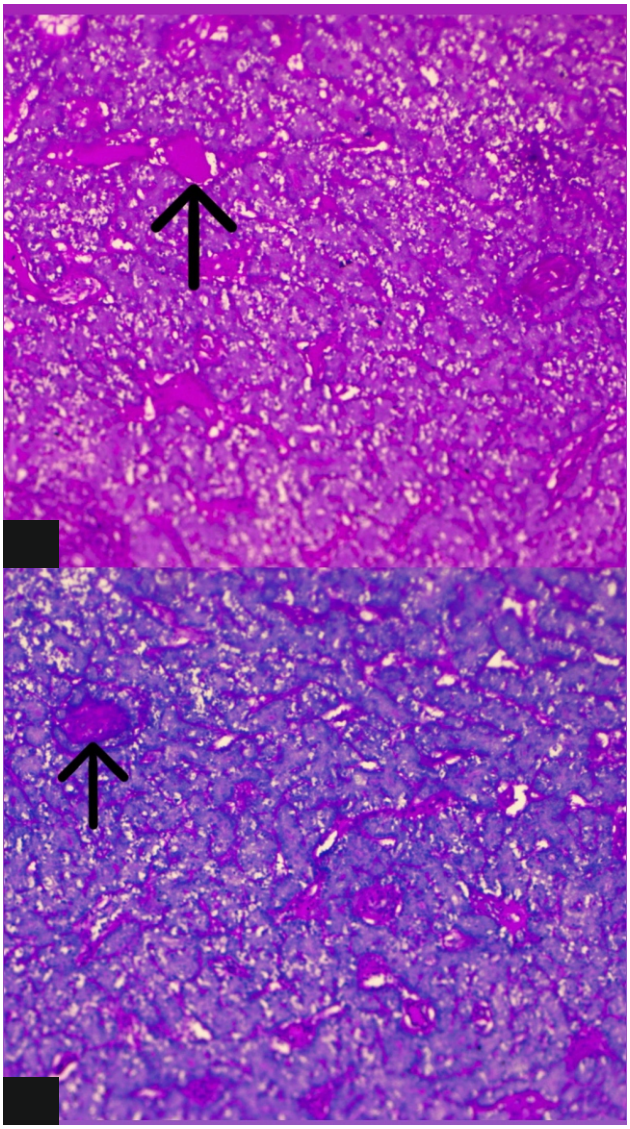

Fig. 4: (a) (PAS, 10X) PAS positive & (b) (PAS-D, 10X) diastase resistant basement membrane material was seen.

DISCUSSION

ES classically presents between the ages of 20-40 years with no sex predilection (6). However, in the current case, the patient was 58 years old male. The present case reports the location of ES in the left lumbar region along anterior axillary line approximately 10 cm left and lateral to umbilicus. However, it has predilection for the head/face & upper trunk region of the body (1).

Eccrine spiradenoma is a rare, benign dermal neoplasm showing eccrine differentiation (1,2). Clinically, the lesion presents as solitary, intradermal, circumscribed, round or oval, blue coloured, firm, painful or tender nodule ranging in size from 0.3 - 5 cm in diameter (3). However, in our case the lesion was soft, cystic and painless.

The clinical profile of the patient like male patient in the 6th decade of his life, unusual site of presentation along with soft, cystic and painless nature of the lesion makes it a case report with an unusual clinical presentation.

Grossly, ES is seen to be either uninodular or multinodular tumor centered within the dermis, commonly extending into the subcutaneous tissue (1). However, in the present case, the tumor was uninodular with no subcutaneous tissue present. The definitive diagnosis of ES is in line with the characteristic histopathological examination findings of ES as described for the current case.

Clinical differential diagnosis of sebaceous cyst was ruled out by the absence of cyst wall lining of stratified squamous epithelium with prominent granular layer and lamellated flakes of keratin (4). Also clinical differential diagnosis of lipoma was offered in current case which was ruled out histopathologically by absence of lobules of mature adipocytes separated by fibrous septae (4).

Histologically, it is very important to differentiate ES from cylindroma and trichoepithelioma because of the significant morphological overlap (1). Also, ES is highly vascular having widely dilated vascular channels that may result in a superficial resemblance to an angioma, hemangiopericytoma or glomus tumor (3). Even in the present case, all these histological differentials were ruled out.

Various histochemical and immunochemical stains can be used for the differential identification with ES showing positive staining for PAS, CK-7, CK-8, CK-18, CEA, EMA, S-100, SMA. Immunopositivity for IKH4 is supportive of eccrine differentiation over apocrine (4). The gold standard treatment for ES is complete excision of the tumour with clear margins. Low rates of recurrence has been documented by various literature studies (7). Other treatment options like radiotherapy, chemotherapy and carbon dioxide laser ablation have been proposed although no studies have substantiated an optimal practice (8). Even in the present case, wide local excision with clear margins was surgically performed.

Although, ES is a rare benign neoplasm of sweat gland, however chances of recurrence and malignant transformation increases in patients with long standing history or with multiple lesions. Signs of malignancy include an increase in size and number of lesions and change in colour and any new symptoms such as pain, ulceration and bleeding. Histological features indicative of malignant spiradenoma include loss of two cell populations, increased nuclear-to-

cytoplasmic ratio, hyperchromasia, atypical mitosis and necrosis (4). In our patient, histopathological examination of specimen revealed no such finding. Metastasis of malignant spiradenoma can manifest years after complete removal (4).

CONCLUSION

Eccrine spiradenoma may present congenitally or sporadically as tumour of the sweat glands with unclear etiology. Early accurate diagnosis is very important to prevent recurrence and more importantly to identify onset of malignant transformation. Although rare, malignant transformation has been described in literature especially in long-standing or multiple lesions. Therefore, if multiple lesions (Brooke-Spiegler syndrome) are present, genetic testing for CYLD / p53 tumor suppressor gene should be done. With recent and ongoing improvements in histopathological techniques, documenting the case reports of ES having interesting and unusual clinical presentations in the literature is the need of recent time by the pathologists, thereby, playing an integral role in the better understanding and setting up standard therapeutic protocols for this rare tumour.

REFERENCES

1. Elder DE, Massi D, Scolyer RA, et al. WHO Classification of Skin Tumours. 4th edn. Lyon, France: International Agency for Research on Cancer; 2018.
2. Kersting DW, Helwig EB. Eccrine spiradenoma. AMA Arch Derm. 1956 Mar;73(3):199-227.
3. Calonje JE, Brenn T, Lazar A, et al. Mckee's Pathology of the Skin. 5th ed. Elsevier; 2018. p.1570.
4. Lever WF, Elder DE: Lever's histopathology of the skin (11th ed.). Philadelphia: Wolters Kluwer Health/Lippincott Williams & Wilkins; 2015. p. 1083-1085.
5. Nath AK, Kumari R, Thappa DM. Eccrine spiradenoma with chondroid syringoma in Blaschkoid distribution. Indian J Dermatol Venereol Leprol 2009;75:600-2.
6. Zheng Y, Tian Q, Wang J, et al: Differential diagnosis of eccrine spiradenoma: A case report. Experimental and Therapeutic Medicine 8: 1097-1101, 2014.
7. Englander L, Emer JJ, McClain D, et al: A rare case of multiple segmental eccrine spiradenomas. J Clin Aesthet Dermatol 4: 38-44, 2011.
8. Andreoli MT, Itani KM: Malignant eccrine spiradenoma: a meta-analysis of reported cases. Am J Surg 201: 695-699, 2011.
9. Alhumidi AA. Simple approach to histological diagnosis of common skin adnexal tumors. Pathology and Laboratory Medicine International. 2017;9:37-47
10. Adulkar SA, Dongre AM, Khopkar US, et al. Zosteriform spiradenoma with spiradenocarcinoma: A rare entity. Indian Dermatol Online J. 2015 Dec;6 (Suppl 1):S30-2.
11. Rekhi B, Agarwal A. Cytopathologic features of an unusual case of multiple eccrine spiradenomas misdiagnosed as a malignant round cell tumor. J Cytol. 2017 Jan-Mar;34(1):39-42.
12. Ho KKF, Kwon L, Wong D, Hagley D. Giant vascular eccrine spiradenoma presenting as a thrombosed varix. J Vasc Surg Cases Innov Tech. 2019 Nov 26;5(4):583-585.
13. Cukic O, Jovanovic MB, Milutinovic Z. An Unusual Nodule on the Auricle: Eccrine Spiradenoma. Ear Nose Throat J. 2019 Oct-Nov;98(9):545-546.
14. Metovic J, Gallino C, Zanon E, et al. Eccrine spiradenoma of the nipple: Case report, differential diagnosis and literature review. Histol Histopathol. 2019 Aug;34(8):909-915.

Orcid ID:

Silky Rai - https://orcid.org/0000-0003-4861-8806

Nishi Tandon - https://orcid.org/0000-0002-3710-2743

Reeta Chaudhary - https://orcid.org/0000-0003-4861-8806

Nirupma Lal - https://orcid.org/0000-0001-9615-5426

Osman Musa Hingora - https://orcid.org/0000-0001-8901-7404

How to cite this article:

Rai S., Tandon N., Chaudhary R., Lal N., Hingora O. M. Eccrine Spiradenoma: Case Report with an Unusual Clininal Presentation. Era J. Med. Res. 2022; 9(1): 117-120.