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Case Report

Multiple linear and zosteriform eccrine spiradenoma: A rare case

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Abstract Eccrine Spiradenoma (ES) is an uncommon tumor of eccrine sweat gland. It usually presents as a solitary, painful nodule located over trunk and proximal limbs. Multiple ES is a rare phenomenon, comprising less than 2% of all cases. We report a case of middle-aged female presented with multiple, painless tumors of varying morphology located in linear and zosteriform distribution diagnosed as ES. Prompt diagnosis of ES is pivotal due to risk of life threatening malignant transformation. This article highlights the rare presentation of ES and a brief review of this disease.

Key words
Eccrine spiradenoma; Zosteriform.

Introduction

Eccrine spiradenoma (ES) is an uncommon benign tumor of eccrine sweat gland origin first described in 1956. It usually affects adults with highest incidence in 2nd - 4th decade.[1] Both genders are equally affected. Classically it occurs as a small, blue, solitary, tender, nodule on trunk and proximal limbs.[2] Early diagnosis of eccrine spiradenoma is essentially important firstly because of its high recurrence rate, secondly, its ability of malignant transformation.[3] Multiple Eccrine spiradenoma can present in linear/ zosteriform/ blashkoid pattern however these presentations are extremely rare and only few cases have been reported in the literature.[4,5]

Case Report

40 years old female with no known co-morbids presented to us with complaint of multiple swellings all over the body since 15 years. They first appeared on her face and left forearm then progressed to involve her both arms, legs and trunk over the period of 5 years. Now since 10 years they are persistent with no change in size or symptoms noted by patient. There were no associated symptoms. Her past medical history and family history were unremarkable.

On examination multiple erythematous skin coloured papules, plaques, nodules and cystic lesions of varying sizes were present all over the face, some with surface telangiectasias **(Figure 1)**. Similar lesions were present on neck in linear pattern extending from center of chin to medial side of right clavicle **(Figure 2)**. Multiple, skin coloured nodules, arranged in linear pattern present over flexors and extensors of both forearms, arms and legs **(Figure 3,4)**. Similar nodules were present on left side of trunk in a zosteriform pattern **(Figure 5)**. On palpation they were soft in consistency, non-tender, compressible with minimal fluctuation. Diascopy showed blanchable telangiectasias. Needle aspiration was done to confirm the lymphatic or vascular nature of lesions but it was negative. Systemic examination was unremarkable.

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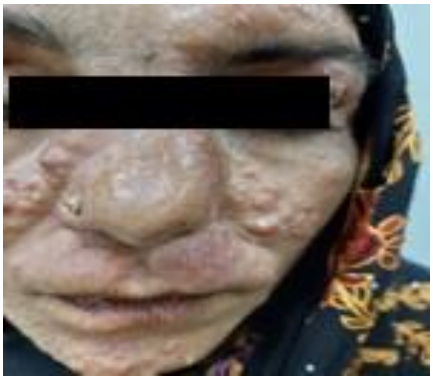

Figure 1 Lesions on face.

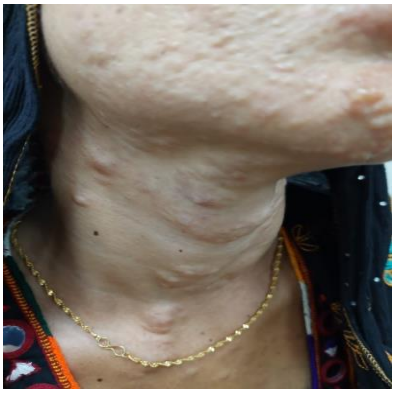

Figure 2 Lesions in linear distribution on neck.

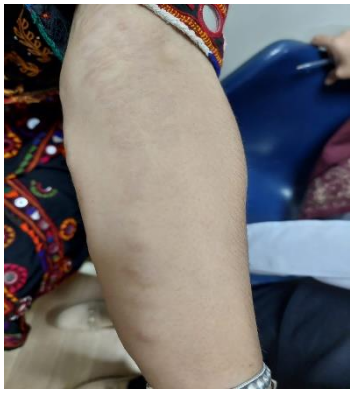

Figure 3 Lesion on forearm.

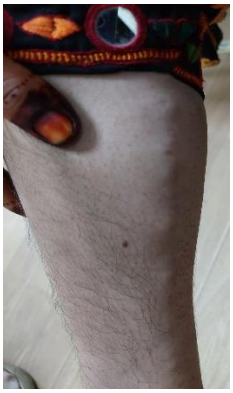

Figure 4 Leg.

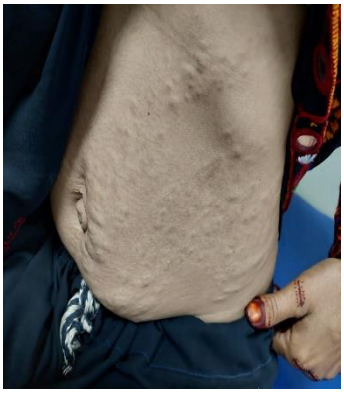

Figure 5 Trunk.

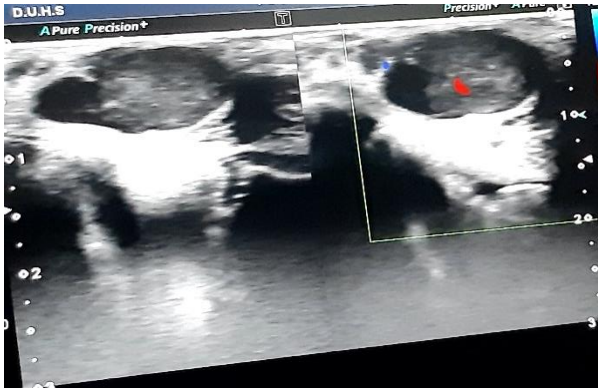

Figure 6

Ultrasound shows hypoechoic lobular masses located in dermis and subcutaneous fat layer. They were solitary on face and arranged in linear tract like pattern on limbs. Color Doppler of periorbital lesion

Figure 6 showed blood flow signal in central portion of mass, however no flow noted within the lobular masses of limbs. No connection with underlying vessels **(Figure 7)**.

Skin biopsy was taken from 2 different lesions located on face showed multiple well circumscribed, basophilic nodular lesions centered within the dermis and subcutis **(Figure 8,9)**. At higher magnification these neoplastic nodules were composed of two cell population with large pale staining cells in the centre and dark basaloid cells at the periphery admixed with lymphocytes suggestive of Eccrine spiradenoma. No evidence of malignant transformation **(Figure 10)**.

A final diagnosis of multiple linear/ zosteriform spiradenoma was made based on clinical and characteristic histopathologic findings. Patient was counselled about the course of disease and possible risk of malignant transformation. Regular follow up was advised to monitor disease activity.

Discussion

Eccrine glands are type of sweat glands that open directly to the surface, present all over the body, especially in the palms, soles and axillae.[4] ES is a benign neoplasm originating from the small transitional area between the secretory part and the coiled duct of the eccrine apparatus. It usually presents as a single, bluish, rounded,

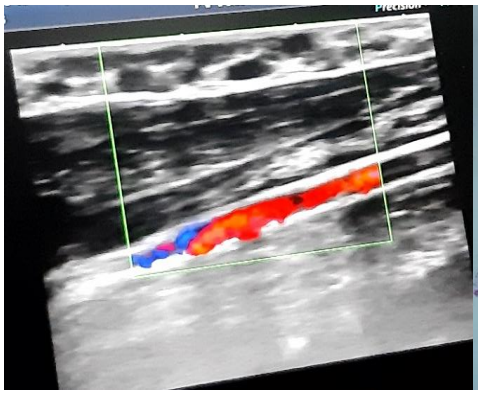

Figure 7

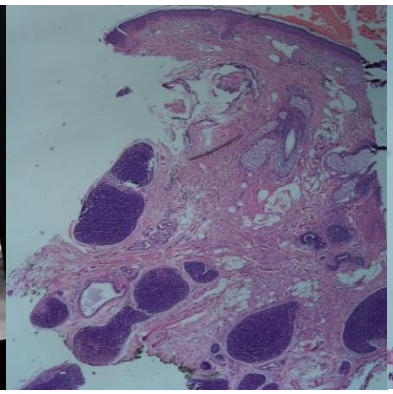

Figure 8

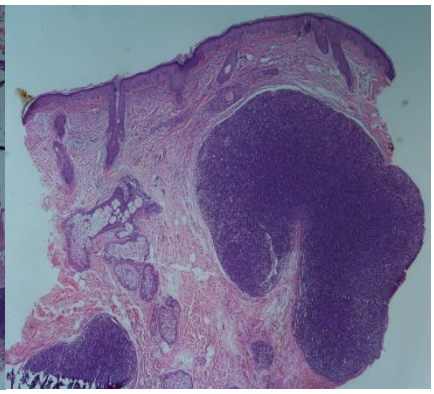

Figure 9

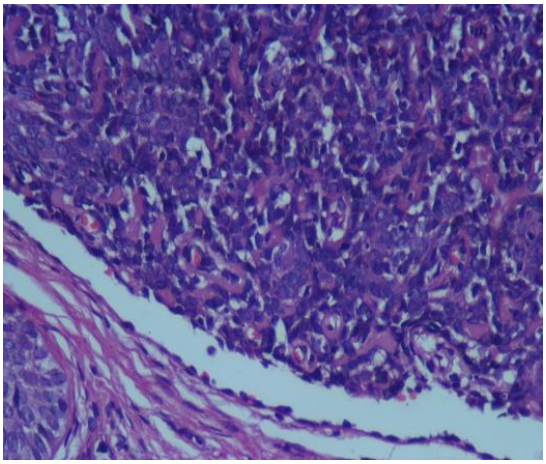

Figure 10

firm, and painful dermal nodule commonly involving trunk and proximal limbs.[5] It may mimics other skin tumors like papilloma, neurofibroma, dermatofibroma, leiomyoma and angiolipoma.[6]

More than 97% of ES presents as solitary, however multiple lesions arranged in linear/ zosteriform/ blashkoid may be seen in 2% of cases.[2] ES is rare with only 102 cases reported so far but multiple ES is even rarer with only 22 cases reported in the literature.[6]

Although ES more commonly occurs in young adults, studies shows that nodules can be present at birth specially when occurring as a part of Brooke Spiegler syndrome.[7]

Malignant transformation may occur in chronic, benign ES, however De-novo cases have also been seen.[1] Evolving lesions showing rapid increase in size, pain, ulceration, redness or increase in number of nodules should raise the possibility of malignant change.[3] The tumor may remain silent for six months to 70 years before malignant transformation.[6] Malignant tumors are highly aggressive with metastatic rate close to 50 percent occurring via both lymphatic and hematogenous route.[1] It commonly metastasize to lymph nodes, lung, brain, liver, and bone.[6]

Due to the clinical resemblance to other skin tumors biopsy is vital for making the correct diagnosis of ES.[8] Histopathology shows well-defined, multilobulated dermal and subcutaneous nodules known as “blue balls in the dermis”. These tumor contains two types of cells, larger cells with large pale nuclei present in the center and smaller cells with scanty cytoplasm and hyperchromatic nuclei arranged the periphery.[2] Features suggestive of malignant transformation are frequent mitotic figures with markedly atypical cells, necrosis, loss of the lobular pattern and absence of dual cell population.[2]

Surgical excision is the gold standard treatment for localized, cosmetically disturbing or painful tumors. Owing to the high recurrence rate up to 57%, wide surgical excision or Mohs

micrographic surgery offers the most conservative treatment choice.[1,6] For widespread lesions or malignant lesions radiotherapy, CO_2 laser or chemotherapy, are better options.[1] Use of intralesional botulinum toxin A and intralesional steroids is still under trials with no clear benefits.[4]

Conclusion

ES is a rare entity. Our case highlights the unusual presentation of this neoplasm occurring as painless lesions with varying morphology in a multiple linear and zosteriform pattern, which is extremely rare. Timely diagnosis of disease is crucial as it has potential for malignant transformation and high recurrence rate. Patient's education regarding the course of disease and close follow ups are very important to identify any aggressive change.

References

1. Englander, L., *et al*., A rare case of multiple segmental eccrine spiradenomas. *J Clin Aesthet Dermatol*. 2011;**4(4)**:38-44.
2. Nath, A.K., R. Kumari, and D.M. Thappa, Eccrine spiradenoma with chondroid syringoma in Blaschkoid distribution. *Indian J Dermatol Venereol Leprol*. 2009;**75(6)**:600-2.
3. Kanwaljeet, S. and T. Chatterjee, Eccrine spiradenoma: A rare adnexal tumor. *Indian J Cancer*. 2017;**54(4)**:695-6.
4. Ren, F., *et al*., Multiple Segmental Eccrine Spiradenoma with a Zosteriform Pattern: A Case Report and Literature Review. *Annals Dermatol*. 2015;**27(4)**:435-8.
5. Saha, M., *et al*., Multiple eccrine spiradenomas in zosteriform distribution involving three dermatomes. *Indian J Dermatopathol Diagn Dermatol*. 2016;**3(2)**: 75-6.
6. Salim, A., M.U. Tariq, and S. Zeeshan, Eccrine spiradenoma: a rare adnexal tumour with atypical presentation: A case report. *J Pak Med Assoc*. 2021;**71(2a)**:553-5.
7. Dhua, S. and D.R. Sekhar, A rare case of eccrine spiradenoma-treatment and management. *Eur J Plast Surg*. 2016;**39**: 143-6.
8. Kim J., H.J. Yang, and J.S. Pyo, Eccrine Spiradenoma of the Scalp. *Arch Craniofacial Surg*. 2017;**18(3)**:211-3.