Zosteriform spiradenoma with spiradenocarcinoma: A rare entity

Satish A. Adulkar, Atul M. Dongre, Uday S. Khopkar, Raghuram Maddala[1], Prachi V. Gole, Siddhi Chikhalkar

Department of DVL, Seth GS Hospital, Mumbai, [1]Department of DVL, J.L.N H and RC, Bhilai, Chhattisgarh, India

ABSTRACT

Eccrine spiradenoma (ES) is an uncommon well-differentiated benign tumor originating from the sweat glands. It usually occurs as a single lesion in adults. Multiple ES in a linear or zosteriform distribution are rare. Spiradenomacarcinoma is an extremely rare tumor, which develops in an existing benign spiradenoma of several years of duration. We report a case of a 23-year-old- female patient with multiple spiradenomas arranged in zosteriform pattern and malignant transformation occurred in one of the lesions over a period of 10 years.

Key words: Malignant eccrine spiradenomas, multiple spiradenomas, spiradenocarcinoma, zosteriform spiradenoma

INTRODUCTION

Eccrine spiradenoma (ES) is a slow growing well-differentiated tumor. Although it is uncommon the malignant counterpart is extremely rare, and our case is peculiar in its distribution pattern also with typical histopathology.

CASE REPORT

A 23-year-old- female patient presented with multiple small asymptomatic skin colored to reddish raised lesions over the chest and back of 10 years of duration. Lesions appeared first when she was 13 years old. These lesions gradually increased in number and size over a period of 10 years. Two months ago one of the nodules on the left scapular area started growing rapidly in size accompanied with pain.

Examination revealed multiple skin colored, erythematous, grouped, non-tender firm papules, and nodules varying in size from 0.5 to 5 cm over left chest and back in segmental pattern [Figure 1]. A few of the lesions showed bluish tinge over the surface. An erythematous tender nodule of approximate size 4 cm × 5 cm was noted on the back on left side of body [Figure 2]. There was no regional lymphadenopathy or hepatosplenomegaly.

Histopathological examination of the small nodules on the back revealed multiple well-circumscribed basophilic tumor islands located in dermis and subcutaneous tissue [Figure 3]. The nodules were composed of basophilic cells. On higher magnification two types of cells were seen. Dark, basaloid cells with hyperchromatic nuclei and cells with large pale vesicular and ovoid nuclei were seen. A few tubules were also seen [Figure 4]. These features were suggestive of benign spiradenoma.

However, biopsy from large erythematous painful nodule over back showed large aggregates of tumor cells with mild to moderate nuclear atypia, abnormal mitotic figures, and necrosis en masse [Figure 5]. At places features of benign component and gradual transformation into a malignant one were noted suggestive of spiradenocarcinoma. She was investigated

Access this article online
Website: www.idoj.in
DOI: 10.4103/2229-5178.171057
Quick Response Code:

Address for correspondence:
Dr. Satish A. Adulkar, Plot 12 B, Dhangavalinagar Pipla Road, Hudkeshwar, Nagpur - 440 034, Maharashtra, India.
E-mail: dr.satish.adulkar@gmail.com

Cite this article as: Adulkar SA, Dongre AM, Khopkar US, Maddala R, Gole PV, Chikhalkar S. Zosteriform spiradenoma with spiradenocarcinoma: A rare entity. Indian Dermatol Online J 2015;6:30-2.

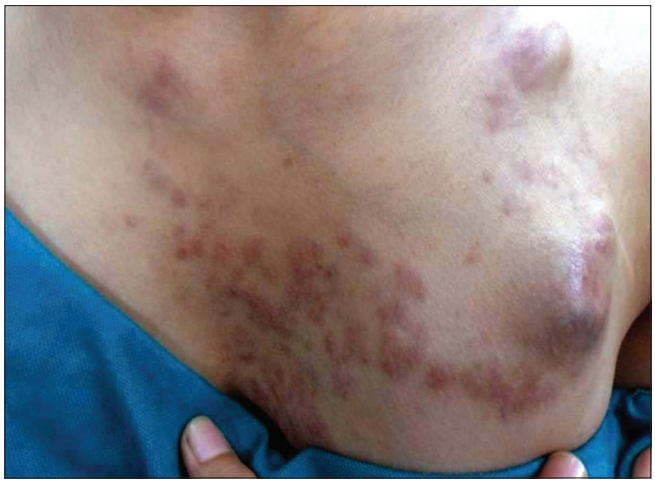

Figure 1: Erythematous to skin-colored small and large nodule on the left chest

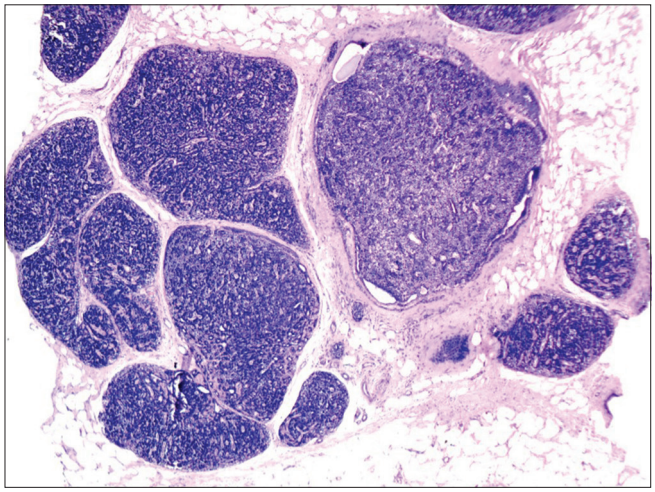

Figure 3: Multiple, basophilic, well-circumscribed tumor islands (H and E ×100)

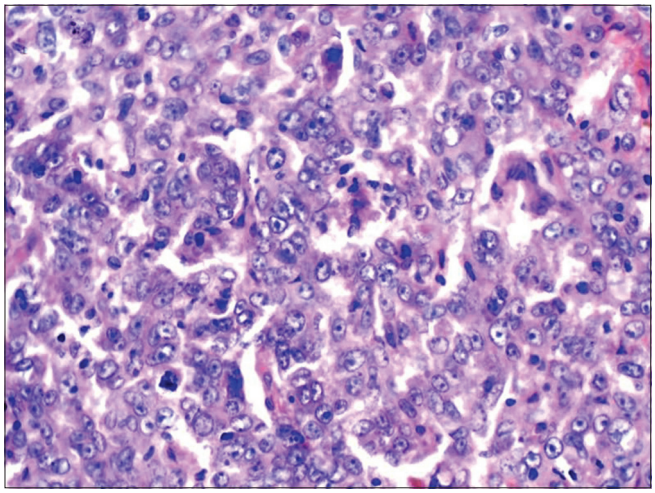

Figure 5: Nuclear atypia with abnormal mitoses

further to detect any metastases, which were absent. She had undergone a wide surgical excision without any adjuvant therapy.

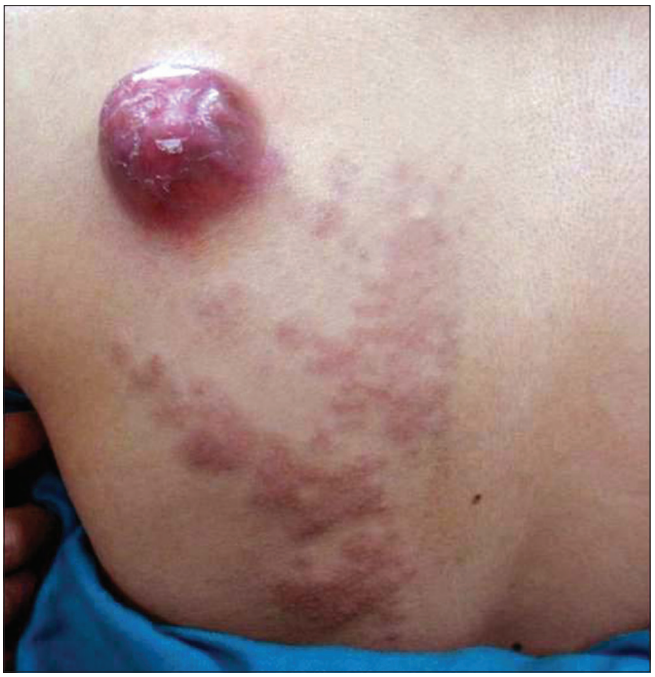

Figure 2: Large erythematous nodule on the left scapular region

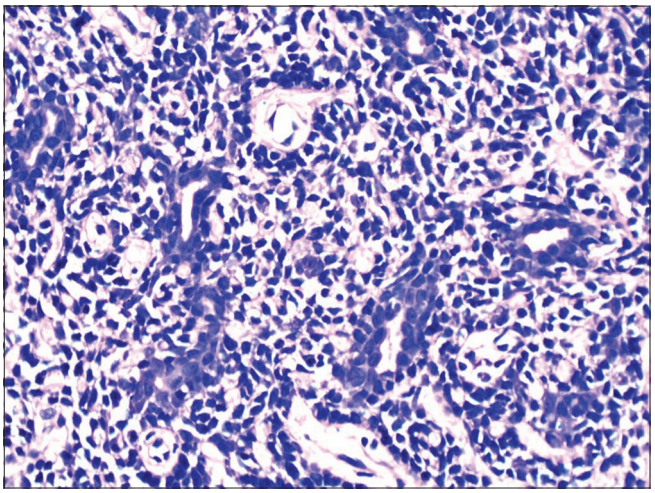

Figure 4: Two type of cells with tubules (H and E ×400)

DISCUSSION

ES is an uncommon well-differentiated benign tumor originating from the sweat glands.[1] It usually occurs as a single lesion in adults. Multiple ES in a linear or zosteriform distribution are rare.[2] ES is uncommon slowly growing well-differentiated benign adnexal neoplasm that has been historically designated as tumor of eccrine lineage, however some reviews indicate an apocrine origin.[3,4] It classically presents as single, firm, or soft skin-colored or bluish nodule, ranging in size from 0.5 to 5 cm in diameter. The most striking clinical feature of ES is the presence of pain or tenderness.[3] There is no male or

female preponderance.[5] ES may occur with other tumors such ascylindroma and trichoepithelioma as seen in Brooke-Spiegler syndrome.[6] Histopathologically, eccrine spiradenoma consist of one or more large, sharply defined basophilic nodules in the dermis without any connections to the epidermis and sometimes it may extend into the subcutis. They consist of groups of cells in cords or islands or sheets. Two types of cells are found in these nodules. These are small, dark, basaloid cells with hyperchromatic nuclei and cells with large pale vesicular and ovoid nuclei. Pale nuclei tend to be at the center of the lesions.[7]

Malignant eccrine spiradenomas (MES) is a rare tumor that was first described by Dabska in 1972.[8] MES generally arises as malignant transformation in long-standing benign ES and rarely can arise *de novo*.[9,10] Clinical features of MES reported in the literature include a previously stable lesion of more than 2 years of duration to more than 12 years duration.[11] There is no age, gender, or site predilection.[12,13] Majority of reported cases have occurred on the extremities (upper > lower) or trunk and very rare cases have been reported on the head and neck.[8] The tumor size ranges from 0.5-10 cm. All spiradenocarcinomas contain areas of benign precursor lesion. Apart from that, two different histologic patterns can be observed: (1) gradual transition from benign to malignant where the dual cell population of the spiradenoma imperceptibly merges with the monomorphous cell population of the carcinoma and poorly defined cell nests and cords replace the structural pattern of spiradenoma. (2) No transition from benign to malignant, the malignant areas are adjacent to and demarcated from spiradenoma.

Advanced stages of both patterns display necrosis, hemorrhage, and infiltrative growth pattern.[7] On immunohistochemistry tumor cells of MES express cytokeratin, epithelial membrane antigen, and p53.[7]

The clinical course of MES is aggressive. Multiple local recurrences and widespread metastases are common. Lymph nodes, bones, and lungs are the organs frequently affected.[14,15] Management is primarily wide local excision. The role of prophylactic lymph node dissection, post-operative radiotherapy, and chemotheraphy is uncertain. If there is an absence of distant metastasis and presence of regional lymph node metastasis, lymph node dissection should be undertaken.[16]

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Gupta S, Radotra BD, Kaur I, Handa S, Kumar B. Multiple linear eccrine spiradenomas with eyelid involvement. J Eur Acad Dermatol Venereol 2001;15:163-6.
2. Gupta S, Jain VK, Singh U, Gupta S. Multiple eccrine spiradenomas in zosteriform distribution in a child. Pediatr Dermatol 2000;17:384-6.
3. Englander L, Emer JJ, McClain D, Amin B, Turner RB. A rare case of multiple segmental eccrine spiradenomas. J Clin Aesthet Dermatol 2011;4:38-44.
4. Jin W, Kim GY, Lew BL, Yang DM, Kim HC, Ryu JK, *et al*. Sonographic findings of an eccrine spiradenoma: Case report and literature review. J Ultrasound Med 2008;27:813-8.
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. Weyers W, Nilles M, Eckert F, Schill WB. Spiradenomas in Brooke-Spiegler syndrome. Am J Dermatopathol 1993;15:156-61.
7. Lever WF, Schaumburg-Lever G. Histopathology of the Skin. Philadelphia: JB Lippincott; 1983. p. 555-7.
8. Beekley AC, Brown TA, Porter C. Malignant eccrine spiradenoma: A previously unreported presentation and review of the literature. Am Surg 1999;65:236-40.
9. Braun-Falco M, Bonel H, Ring J, Hein R. Linear spiradenoma with focal malignant transformation. J Eur Acad Dermatol Venereol 2003;17:308-12.
10. Leonard N, Smith D, McNamara P. Low-grade Malignant eccrine spiradenoma with systemic metastases. Am J Dermatopathol 2003;25:253-5.
11. Elder D, Elenitsas R, Jaworsky C. Lever's Histopathology of the Skin; 8-th ed. Philadelphia: Lippincott Raven; 1997. p. 785.
12. Ishikawa M, Nakanishi Y, Yamazaki N, Yamamoto A, Malignant eccrine spiradenoma: A case report and review of the literature. Dermatol Surg 2001;27:67-70.
13. Granter SR, Seeger K, Calonje E, Busam K, Mckee PH.Malignant eccrine spiradenoma (Spiradenocarcinoma): A clinicopathologic study of 12 cases. Am J Dermatopathol 2000;22:97-103.
14. Evans HL, Su D, Smith JL, Winkelmann RK.Carcinoma arising in eccrine spiradenoma. Cancer 1979;43:1881-4.
15. Zamboni AC, Zamboni WA, Ross DS. Malignant eccrine spiradenoma of the hand. J Surg Oncol 1990;34:131-3.
16. Jamshidi M, Novak MA, Chiu YT, Perry EA, Fatteh SM. Giant, malignant eccrine spiradenoma of the scalp. Dermatol Surg 1999;25:45-8.