American Journal of Case Reports

e-ISSN 1941-5923
© Am J Case Rep, 2019; 20: 1896-1901
DOI: 10.12659/AJCR.919444

Received: 2019.08.15
Accepted: 2019.10.01
Published: 2019.12.19

Syringoid Eccrine Carcinoma in the Abdominal Wall: A Rare Case Report and Literature Review

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABCDEF 1 Miguel Augusto Pereira
AF 2 Luciana Pantaleão
AF 2 Mayra Rochael

1 Medical School of Fluminense Federal University, Niterói, Rio de Janeiro, Brazil
2 Department of Pathology, Fluminense Federal University, Niterói, Rio de Janeiro, Brazil

Corresponding Author: Miguel Augusto Pereira, e-mail: mappereira@icloud.com.br
Conflict of interest: None declared

Patient: Male, 58-year-old
Final Diagnosis: Syriongoid eccrine carcinoma
Symptoms: Burning sensation
Medication: —
Clinical Procedure: Surgical excision
Specialty: Surgery

Objective: Rare disease

Background: Syringoid eccrine carcinoma (SEC) is an extremely rare malignant adnexal neoplasm derived from eccrine sweat glands, of unknown pathogenesis. We report a case of this rare entity presenting in the abdomen, which is the only one reported in this area and the only case of SEC in a patient with so many comorbidities.

Case Report: A 58-year-old black male from Brazil reported a nodular lesion in the abdomen with a progressive increase in size and pain and local burning sensation. The histopathological examination showed a syringoid eccrine carcinoma.

Conclusions: We present a rare case of SEC and did an extensive literature review in order to describe the clinical characteristics, histopathological findings, immunohistochemical profile, treatments, and difficulties found in the diagnosis of this tumor. To avoid misdiagnosis, we gave special attention to biopsy quality.

MeSH Keywords: Carcinoembryonic Antigen • Diagnosis, Differential • Immunohistochemistry • Sweat Gland Neoplasms

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/919444

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

Background

Syringoid eccrine carcinoma (SEC) is an extremely rare malignant adnexal neoplasm derived from eccrine sweat glands, whose pathogenesis has not yet been well defined. It equally affects males and females, is most often present in the fourth to seventh decade of life, and represents less than 0.01% of all skin cancers [1–3]. This tumor is considered extremely invasive, locally destructive, slow growing and high recurrence, but difficult to metastasize.

This type of carcinoma has been referred to by many names throughout history, probably due to the different stages of differentiation found in the lesion [1]. It was first described in 1969 by Freeman and Winklemann as a basal cell tumor with eccrine differentiation [4]. Thus, from the 1970s onwards, other names emerged, including malignant syringoma [5], sweat gland carcinoma with syringomatous characteristics [6], eccrine epithelioma [4,7] and sclerosing carcinoma (syringomatosis) of sweat duct [8]. However, the term SEC is more appropriate because of cytological and enzymatic features that differ from basal cell carcinoma [9]. In this article, we report a case of a 58-year-old man with this rare malignant neoplasia in an unusual location.

Case Report

A 58-year-old black male, a bricklayer from Brazil, with no family history of cancer, was examined for a cutaneous lesion in the left abdominal region. The patient reported the appearance of a nodular lesion in the abdomen 3 years ago, with a progressive increase in size and local burning sensation. The patient is a former smoker and former consumer of alcoholic beverages already treated in the hospital due to schizophrenia, asthma, and arterial hypertension, and medical history of 5 episodes of tuberculosis (last day of treatment in 2011), portal hypertensive gastropathy, and HCV cirrhosis (treated in 2016).

The dermatological examination revealed a hardened nodule measuring about 2 cm in diameter, painful to palpation. The overlying skin presented a brownish color, erythematous, with hardened papules of the same color, coalescing, around the nodular lesion (Figure 1).

Pathological findings

At the macroscopic examination, a skin fragment measuring 7.0×5.0×1.5 cm was observed, with dark brown epidermis showing a slightly delimited paracentral lesion slightly elevated measuring 1.5×1.5 cm. A 2.0-cm central white linear scar was

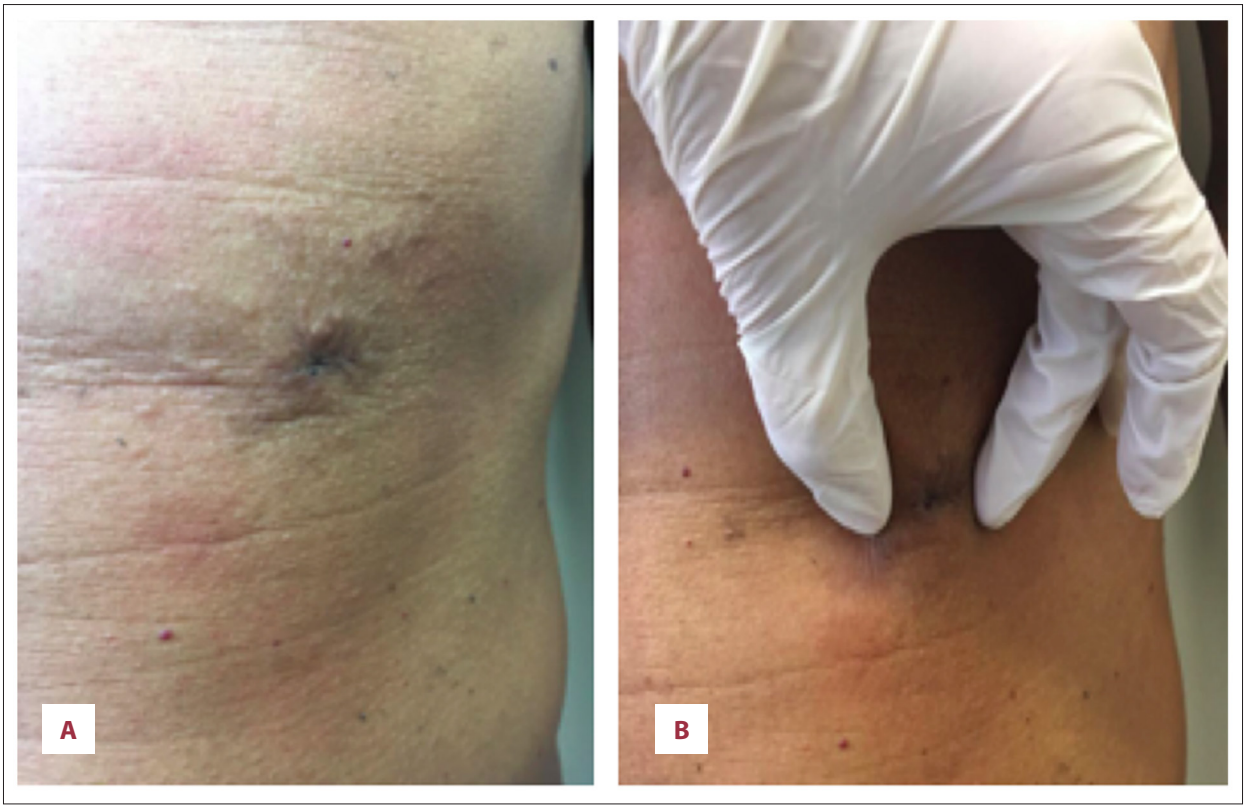

Figure 1. (**A, B**) Atrophic lesion of poorly defined margins located in the upper left quadrant of the abdomen. The overlying skin presented a dark and brownish color with hardened papules of the same color, coalescing, around the central lesion.

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

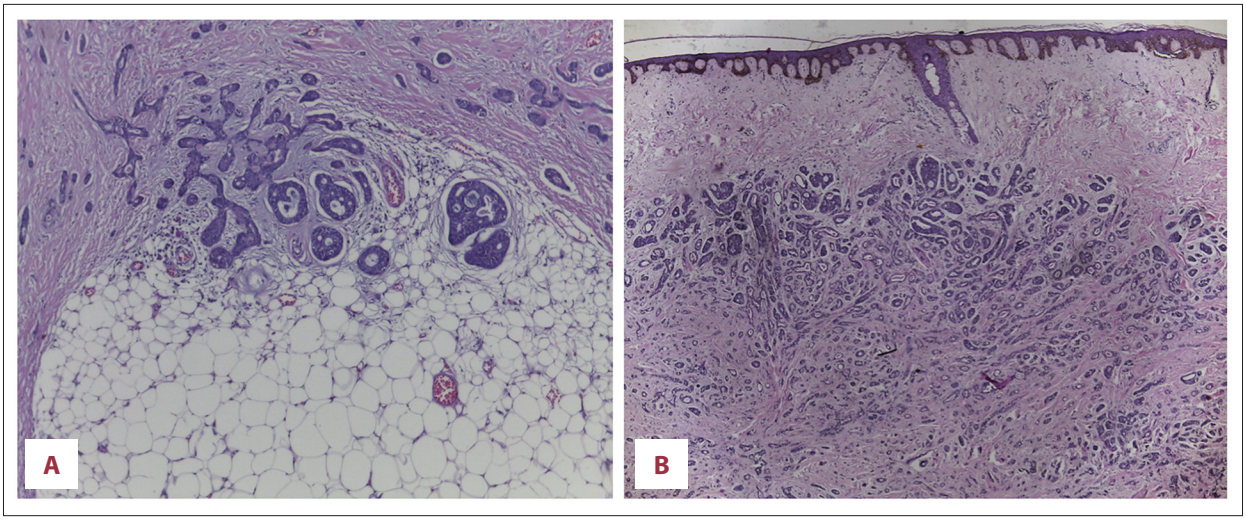

Figure 2. (**A**) Syringoid eccrine carcinoma consisting of atypical epithelial cells forming multiple architectures such as cords and tubules in the midst of a compact dense collagen matrix (desmoplastic reaction). (H&E, original magnification ×40). (**B**) Infiltration of subcutaneous tissue by tubules and cords and some with tadpole morphology. (H&E, original magnification ×100).

observed, and the lesion was located 2.0 cm from the nearest margin. The cut surface was white and elastic, with a thickened area in the area of the described lesion.

The histopathological examination revealed a syringoid eccrine carcinoma, which consisted of atypical epithelial cells forming cords and tubules in the midst of a compact dense collagen matrix, infiltrating even the hypodermis (Figure 2). There were regular acanthosis of the epidermis, multiple foci of perineural invasion, and absence of vascular invasion (Figures 2, 3). No usual skin adnexal were observed. There were circumferential and deep compromised surgical limits. The material was submitted to immunohistochemical study, showing reactivity for S100, CK7, EMA (Epithelial Membrane Antigen), p-CEA (CD66e), and SMA (alpha smooth muscle actin), as well as negativity for CK20 (Figure 4).

The treatment used by the medical team was surgical excision under local anesthesia, and no flap was required. There was no recurrence of the lesion so far (follow-up of 15 months); however, surgery is being scheduled to increase the surgical margin.

Discussion

SEC usually manifests as a subcutaneous plaque or nodule with slow growth and poorly delimited margins, arising between the fifth and sixth decade of life, which was compatible with the age of the patient presented. However, the location in which the lesion was found is somewhat unusual, since SEC predominates in the area of the head (scalp) and neck. However, in the literature, there are cases reported in the trunk, extremities, nipples, vulva, and ungual complex, as well as in the scapular region, plantar region, and even in the auditory canal [1,9–15].

The histology of SEC can range from a well-differentiated to a more anaplastic lesion, with little evidence of eccrine differentiation. It is characterized by numerous infiltrative structures, connected and covered by squamous or basaloid cells of hyperchromatic nuclei, in a fibrocolagenous stroma (desmoplastic reaction), extending through the dermis, underlying subcutaneous tissue or skeletal muscle, with perineural infiltration. Tumor cells can organize into nests, cords, or tubules, some of which exhibit a morphology resembling tadpole-like syringoma due to the cords of epithelial tissue. Cytonuclear atypia is not very evident, as is the presence of mitotic activity, which may even be absent [1,11,14,16]. The epithelial component of the tumor may show a cribriform pattern in focus (Figure 3), which makes it difficult to distinguish from other tumors [3].

The immunohistochemical profile of SEC is quite varied, with the carcinoembryonic antigen (CEA) being the most consistent marker [17]; other markers generally present are high and low molecular weight cytokeratins, the epithelial membrane antigen (EMA), PS100, and Leu M1. In addition, the immunohistochemical study may exclude the possibility of breast cancer in the differential diagnosis.

Despite the apparent poor prognosis, when considering only some characteristics of the tumor, such as infiltration of disfiguring character (especially in the face) or high recurrence, and it is difficult to observe regional metastases, distant metastases or involvement of the lymphatic chain [1,18–20].

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

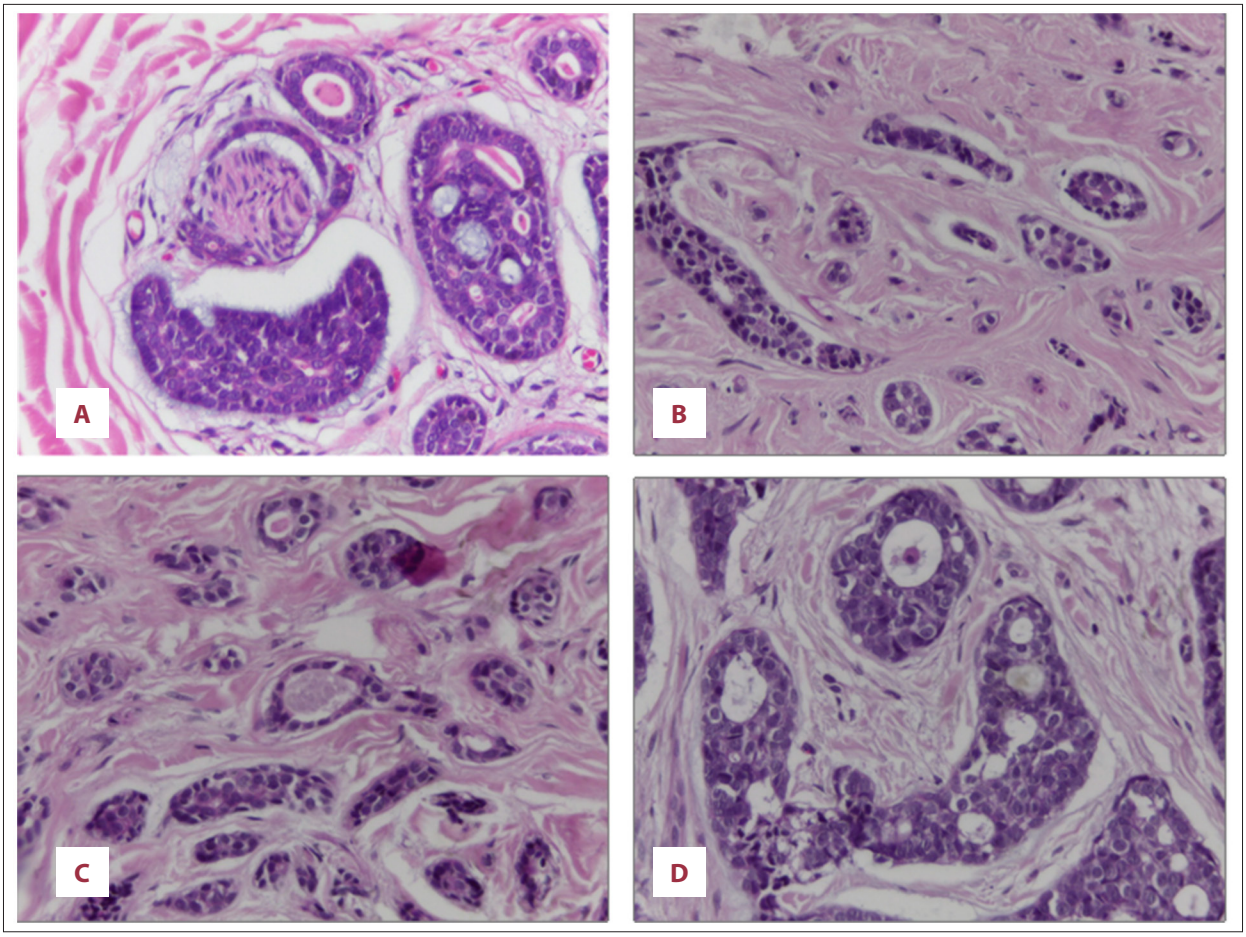

Figure 3. (**A**) The tumor displays perineural invasion. We also observed cystic spaces containing mucin and amorphous eosinophilic material. (**B, C**) Infiltration of the dermis by tubules and cords with (syringoma-like) tadpole morphology. (**D**) Cribriform architecture may be present in this type of tumor. (H&E, original magnification ×100).

The present case was a diagnostic challenge since its initial evaluation. At the time, the clinical diagnostic hypotheses raised were adnexal carcinoma, cutaneous calcinosis, dermatofibrosarcoma protuberans, or panniculitis. The malignant characteristics of the lesion, observed under microscopy, also suggested cutaneous metastasis of adenocarcinoma of unknown primary site, adenoid cystic carcinoma (ADC), and SEC. In the differential diagnosis with ADC, perineural infiltration and histological presentation might resemble ADC with a cribriform morphological pattern; however, the positivity for CEA and S100 in our patient would be more consistent with the tubular pattern [21]. Thus, in addition to the immunohistochemical pattern disagreeing with the morphological pattern, there is no accumulation of mucin, as occurs in ADC, favoring the diagnosis of SEC [3,14,19,17]. Imaging studies have been reported to be important but for delimiting the lesion, but not for diagnosis of SEC [11], or searching for metastases [9,10,15].

Every physician should pay attention to the biopsy quality, which should be of adequate size and depth. In some cases, a superficial biopsy (e.g., shaved off) may not adequately represent the architectural and depth of infiltration patterns, and are thus erroneously interpreted as being benign syringomas, trichoepitheliomas, trichoadenomas, or squamous cell carcinomas due to the microscopic appearance. Thus, malignant lesions such as SEC can be mistaken for benign lesions such as benign syringomas. If ductal differentiation is poorly represented, an erroneous diagnosis of basal cell carcinoma can occur [22]. The importance of biopsy quality is demonstrated by a study involving microcystic adnexal carcinoma of the skin conducted by Leboit, in which 9 of the 17 lesions studied were initially misdiagnosed due to the small size of the biopsy specimen [23], which is also a mistake made in the diagnosis of SEC. It should be noted that this is an extremely infiltrating and disfiguring lesion, having serious consequences for patients.

In this case, the criterion standard treatment was used, so surgical resection of the lesion was performed. This method is considered to be optimal and the main treatment choice, since the other option, radiation, is rarely used since the tumor

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

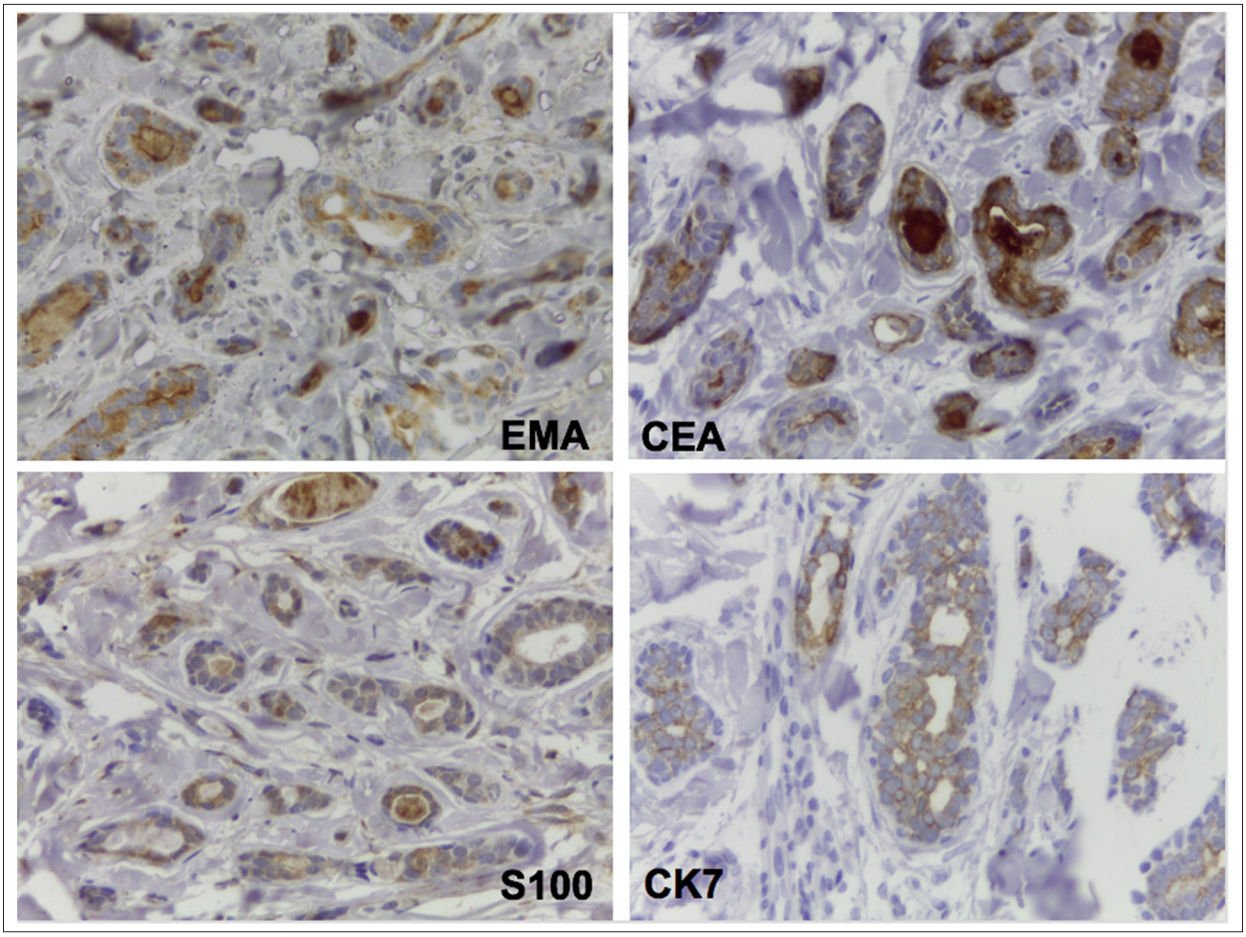

Figure 4. Carcinoembryonic antigen (CEA) is very reactive on cytoplasmic membranes. Cytokeratin (CK)7, protein S-100, and epithelial membrane antigen (EMA) are positive. (Original magnification ×100).

is considered resistant, but it has been reported in the literature as adjuvant therapy [11].

Follow-up is necessary to evaluate recurrence, since this type of tumor is frequently recurrent, with about 40% to 60% of patients presenting within 6 months to 30 years after excision [22].

Conclusions

In conclusion, we report a rare case of SEC presenting in the upper left quadrant of the abdomen. To the best of our knowledge, is the only case reported in this area and the only case of SEC in a patient with so many comorbidities. We present another well-illustrated case of this rare entity in order to guide dermatologists and pathologists in this difficult diagnosis, especially regarding the importance of a biopsy that adequately considers the lesion size and depth, thus avoiding serious diagnostic errors.

Conflict of interests

None.

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

References:

1. El khannoussi B, Hechlaf H, Lalya I et al: Syringomatous carcinoma: Case report of a rare tumor entity. Pan Afr Med J, 2012; 12: 76
2. Tulenko JF, Conway H: An analysis of sweat gland tumors. Surg Gynecol Obstet, 1965; 121: 343–48
3. Moy RL, Rivkin JE, Lee H et al: Syringoid eccrine carcinoma. J Am Acad Dermatol, 1991; 24(5): 857–60
4. Freeman RG, Winkelmann RK: Basal cell tumor with eccrine differentiation (eccrine epithelioma). Arch Dermatol, 1969; 100: 234–42
5. Glatt HJ, Proia AD, Tsoy EA et al: Malignant syringoma of the eyelid. Opthalmology, 1984; 91: 987–90
6. Lipper S, Peiper SC. Sweat gland carcinoma with syringomatous features: A light microscopic and ultrastructural study. Cancer, 1979; 44: 157–63
7. Sanchez NP, Winkelmann RK: Basal cell tumor with eccrine differentiation (eccrine epithelioma). J Am Acad Dermatol, 1982; 6: 514–18
8. Cooper PH, Mills SE, Leonard DD et al: Sclerosing sweat duct (syringomatous) carcinoma. Am J Surg Pathol, 1985; 9: 422–33
9. Gregurek-Novak T, Talan-Hranilović J, Troskot N et al: Syringoid eccrine carcinoma. J Eur Acad Dermatol Venereol, 2001; 15: 143–46
10. Won YY, Suh DW, Lew BL, Sim WY: Syringoid eccrine carcinoma of the thigh. Ann Dermatol, 2017; 29: 6
11. Ballardini P, Margutti G, Pedriali M, Querzoli P: Metastatic syringoid eccrine carcinoma of the nipple. Int Med Case Rep J, 2012; 5: 45–48
12. Piovano E, Ferrero A, Ravarino N et al: Syringoid eccrine carcinoma: Case report of a rare tumor occasionally detected in the vulva. Gynecol Oncol Case Rep, 2011; 1: 17–19
13. Grady JF, Boumendjel Y, Tahniyath MS: Subungual syringoid eccrine carcinoma of the great toe nail complex: A case report. J Am Podiatr Med Assoc, 2014; 104: 504–7
14. Cho WC, Wagner B, Gulosh M, Elaba Z: Syringoid eccrine carcinoma of the foot: Report of a rare cutaneous adnexal neoplasm. Int J Surg Pathol, 2017; 25: 659–64
15. Ahmed MK, Ishino T, Hirakawa K, Arihiro K: Syringoid eccrine carcinoma of external auditory canal: A case report. Auris Nasus Larynx, 2010; 37: 519–21
16. Terushkin E, Leffell DJ, Futoryan T et al: Squamoid eccrine ductal carcinoma: A case report and review of the literature. Am J Dermatopathol, 2010; 32: 287–92
17. Sidiropoulos M, Sade S, Al-Habeeb A, Ghazarian D: Syringoid eccrine carcinoma: A clinicopathological and immunohistochemical study of four cases. J Clin Pathol, 2011; 64: 788–92
18. Werbrouck A, Wechsler J, Blin H, Gontier MF: [A palpebral tumor]. Ann Pathol, 2006; 26: 135–37 [in French]
19. Malmusi M, Collina G: Syringoid eccrine carcinoma: A case report. Am J Dermatopathol, 1997; 19: 533–35
20. Goto M, Sonoda T, Shibuya H et al: Digital syringomatous carcinoma mimicking basal cell carcinoma. Br J Dermatol, 2001; 144: 438–39
21. Bradley PJ: Adenoid cystic carcinoma of the head and neck: A review. Curr Opin Otolaryngol Head Neck Surg, 2004; 12: 127–32
22. Hoppenreijs VP, Reuser TT, Mooy CM et al: Syringomatous carcinoma of the eyelid and orbit: A clinical and histopathological challenge. Br J Ophthalmol, 1997; 81: 668–72
23. Leboit PE, Sexton M: Microcystic adnexal carcinoma of the skin. J Am Acad Dermatol, 1993; 29: 609–18

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]