Journal Homepage: -www.journalijar.com

INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI:10.21474/IJAR01/16462
DOI URL: http://dx.doi.org/10.21474/IJAR01/16462

RESEARCH ARTICLE

PRIMARY CUTANEOUS APOCRINE CARCINOMA IN AN ELDERLY MAN: A RARE ENTITY, ABOUT A CASE

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Manuscript Info

Manuscript History
Received: 19 January 2023
Final Accepted: 24 February 2023
Published: March 2023

Abstract

Primary cutaneous gland apocrine carcinoma, a subtype of sweat gland carcinoma, is an extremely rare malignancy and only a few cases have been reported in the literature. It involves the axillary region but can occur elsewhere on the skin. It is indolent and slow growing. Treatment remains surgical with wide local excision and healthy margins, with or without lymph node dissection. We report a case of a 79-year-old man who came to our hospital with a cauliflower-like mass in the right axilla measuring 1.5 × 1 cm. Histological evaluation showed features of apocrine gland carcinoma arising in an area of high apocrine gland density. The objective of this work is to highlight the diagnostic difficulty that this entity can present and the importance of anatomopathological examination to make the diagnosis and eliminate differential diagnoses.

Introduction:-

Primary cutaneous apocrine carcinoma of the apocrine glands (CACP), a subtype of carcinoma of the sweat glands, is an extremely rare malignancy [1].

CACP primarily involves the axillary region, but lesions can also occur elsewhere on the skin. Most often it is indolent and slowly growing, but cases with a rapidly progressive and extremely aggressive course have been reported in the literature [2,5].

Treatment remains surgical with wide local excision and healthy margins, with or without lymph node dissection [1].

Case Observation:-

We report the case of a 79-year-old man, with no personal or family history, who consulted for a budding cauliflower-shaped right axillary mass measuring 1.5 cm × 1 cm. The lump had been there for over 3 years but only increased in size in the past 7 months.

The complete breast, skin, and lymph node physical examination was unremarkable. Anatomical imaging examinations revealed no significant lesions. The mass was biopsied and the specimen was submitted for histopathological examination.

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Microscopic examination revealed a poorly differentiated tumor proliferation, made of nests, spans, and tubes with obvious apocrine characteristics {Figure 1}. The tumor cells were rounded, with abundant eosinophilic cytoplasm, with clarified nuclei with granular chromatin. Numerous apocrine glands were present near the carcinoma {Figure 2}.

The immunohistochemical study showed that the tumor cells expressed the CK7, EMA, and AE1/AE3 antibodies {Figure 3, 4, 5}. On the other hand, they were negative for CD34, CK5/6, P40, PS100, Melan A, CK20, and CD 30 antibodies. On the basis of the morphological and immunohistochemical results above, the diagnosis of CACP was made.

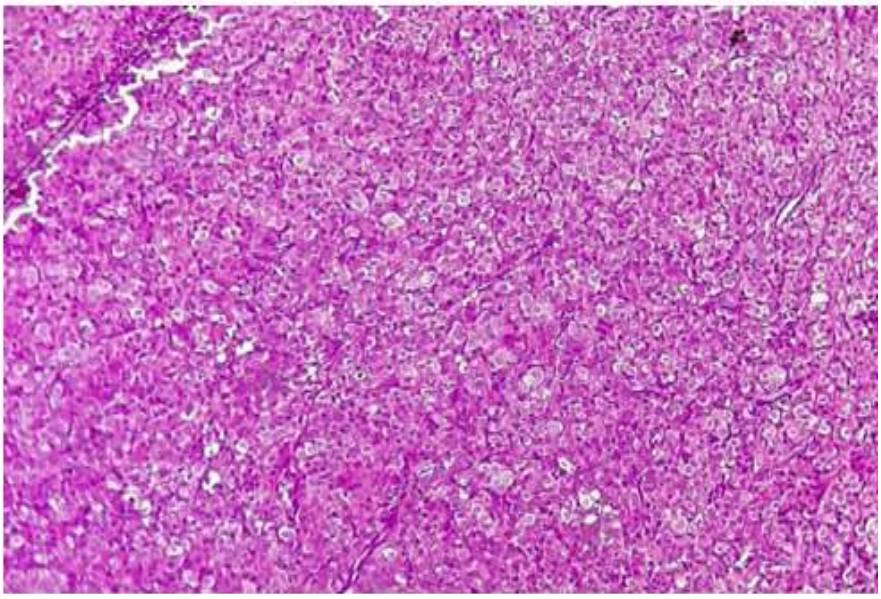

Figure 1:- Carcinoma apocrine x10.

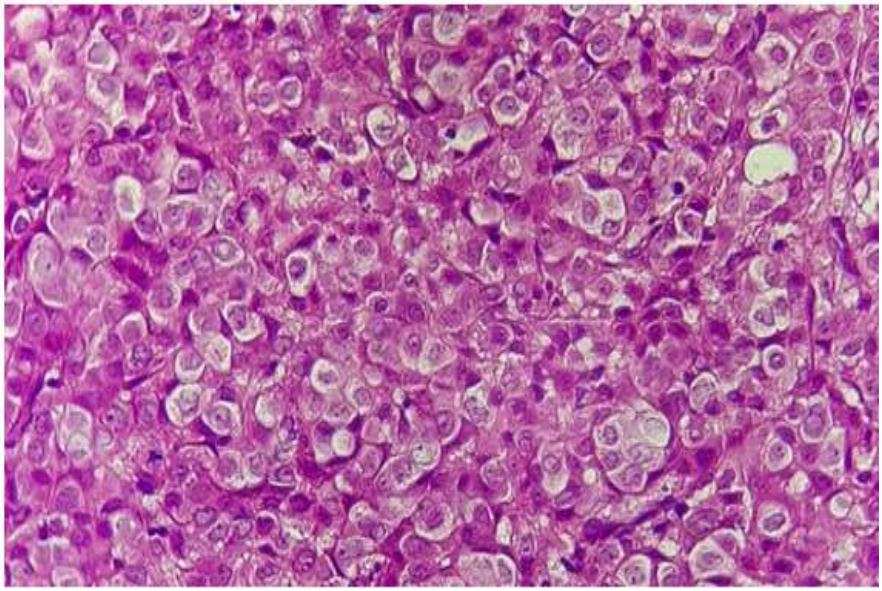

Figure 2:- Apocrinecarcinoma x40.

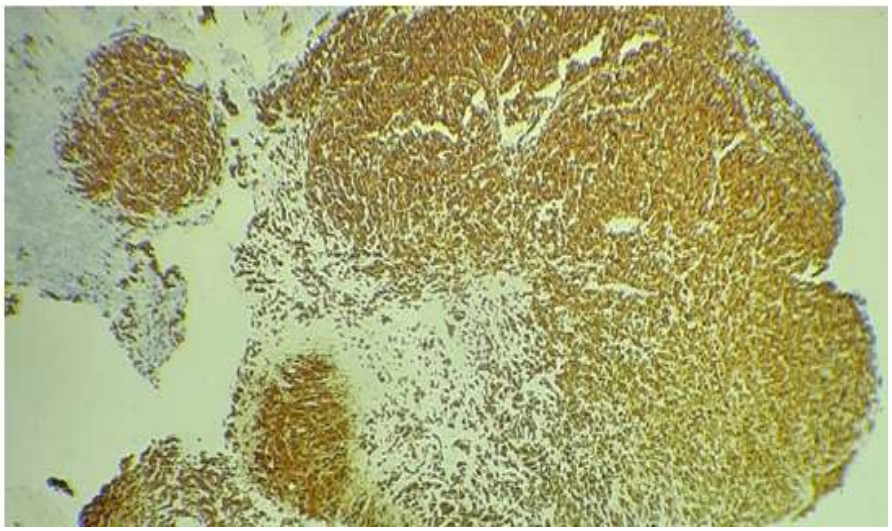

Figure 3:- Apocrinecarcinoma CK7 x10.

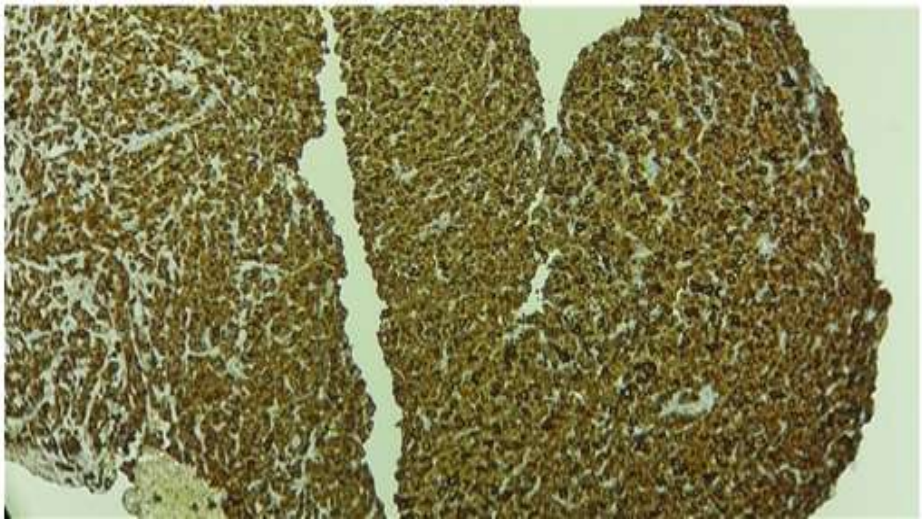

Figure 4:- Apocrinecarcinoma EMA x10.

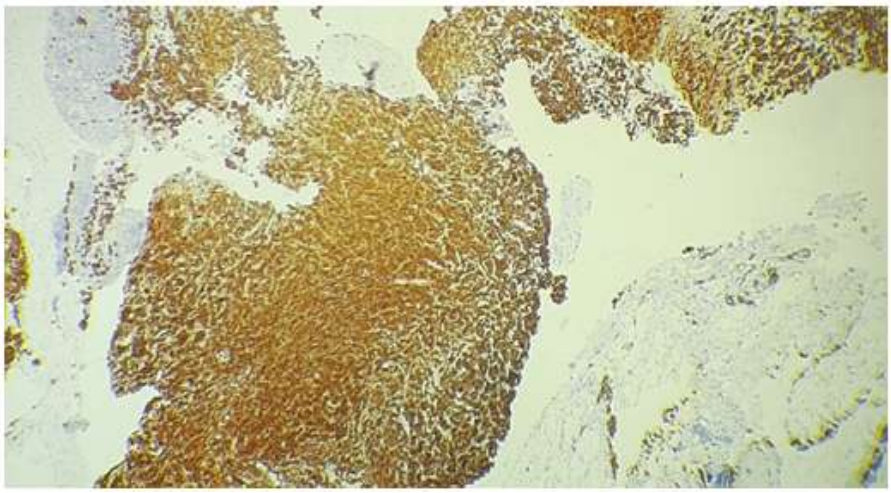

Figure 5:- ApocrinecarcinomaAE1/AE3 x10.

Discussion:-

Primary cutaneous apocrine carcinoma (CACP) is a rare tumor, of which only a hundred cases have been reported in the literature [4].

Demographically, these malignancies have a Caucasian predominance with an equal distribution in both sexes. They commonly show a peak around the 6th and 7th decades of age, with a median age of 67 years, which has been noted in the largest cohort studied to date [1].

CACP occurs primarily in regions with a high density of apocrine glands such as the axillary and anogenital areas, although it has also been reported in less typical locations such as the forehead, wrist, ear canal, eyelid, trunk, foot, toe, and finger [1,2,4].

It is mainly in the form of a single or multinodular mass, 2 to 3 cm, painless, indurated, without bleeding sometimes with ulcerations; with a slow and progressive increase extending over months or even years, but some tumors are rapidly progressive and extremely aggressive involving the loco-regional lymph nodes, with lung, liver, bone, brain and kidney metastases [8,9,10].

These lesions are vague in their presentation and in their appearance and are often misdiagnosed. It usually develops spontaneously, but the association with other benign tumors such as apocrine adenoma and apocrine hyperplasia has also been observed [6,7].

A thorough history and clinical evaluation accompanied by careful histopathological examination are necessary, as they are difficult to differentiate from metastatic skin lesions [3].

The characteristic morphological appearance of apocrine adenocarcinoma shows ductal or glandular structures with apocrine characteristics. The cytoplasm of tumor cells is abundant, eosinophilic with luminal decapitation secretion, and contains periodic acid–Schiff-positive, diastase-resistant, and often iron-positive granules [2].

The diagnosis of primary apocrine carcinoma is based histologically on the apocrine differentiation of the tumor and clinically on a usual anatomical location where the apocrine glands are numerous. The histological picture is similar to adenocarcinoma which can be well, moderately, or poorly differentiated [3, 5].

Histologically, axillary CACP must be differentiated from metastasis of a mammary adenocarcinoma [3].

However, for morphological and immunohistochemical reasons, the differential diagnosis of CACP and mammary adenocarcinoma metastases is not possible except for a few cases, because CACP expresses estrogen and progesterone receptors, and is therefore of limited use in differential diagnosis [8,9,11].

Features that favor the diagnosis of an axillary CACP are neoplastic glands high in the dermis, apocrine glands near the tumor, and intracytoplasmic iron granules [11].

The literature review supports high local recurrence and regional lymph node metastasis but little evidence of distant metastasis. Metastases have been reported in approximately 30% of cases. Regional nodal metastasis is considered an important prognostic factor.

Median overall survival and disease-specific survival at 5 years were 51.5 months and 88%, respectively [1].

The treatment of choice is wide local excision with healthy margins, with or without regional lymph node dissection. Radiation therapy and postoperative chemotherapy have been used as adjunctive treatments but have shown little benefit to mortality. The incidence of local recurrences is high and prophylactic lymph node dissection does not reduce the incidence of recurrences [5].

Conclusion:-

Primary cutaneous apocrine carcinoma remains a rare entity not to be ignored, whose slow, indolent, and progressive evolution causes delays in diagnosis. There is no guideline for the treatment of recurrent or metastatic disease. Therefore, patients with metastatic progression have a very poor prognosis.

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