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Extraocular sebaceous carcinoma: a series of three cases with varied presentation

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ABSTRACT

Background: Extraocular sebaceous carcinomas are rare cutaneous malignancies that account for only about 25% of all sebaceous carcinomas. The most common site of occurrence is the head/neck region. They are aggressive neoplasms that possess metastatic potential to regional or distant sites. Wide local excision of the lesion with removal of regional lymph nodes is the usual mode of therapy.

Methods: We present a series of three cases of sebaceous carcinomas occurring in extraocular sites outside the head and neck areas, like the axilla, chest wall, arm and thigh. One of these cases had an associated colonic carcinoma and constituted Muir-Torre syndrome (MTS). Another case presented with a skin nodule and regional nodal metastasis. The third case had a history of recurrent similar lesions at the same site.

Conclusion: Although rare, extraocular sebaceous carcinomas are seen at varied sites and frequently pose problems in diagnosis. A longterm follow-up of these cases is warranted due to their aggressive behaviour, risks of recurrence, metastasis and the possibility of development of visceral malignancies.

Introduction

Sebaceous carcinomas are most commonly encountered in the ocular region. Extraocular sites for these tumours account for only about a quarter of the cases and usually include the skin of the head and neck region. They can also occur on the trunk, extremities, genitalia and rare sites like lungs, salivary glands, breast [1]. The morphologic diversity of these tumours, especially in unusual sites, makes their distinction from more common primary tumours of those organs a difficult task. It is important to identify them accurately, as they have a high incidence of recurrence and association with regional nodal metastasis.

We report a series of sebaceous carcinomas occurring at extraocular, extra-head and neck sites. These cases are presented for their rarity, histopathologic spectrum and difficulty encountered in diagnosis.

Case Reports

Case 1

A 44-year-old man presented with skin nodules on the arm and anterior chest wall three years ago. The lesions ranged in size from 1 to 2.5 cm in diameter. On excision, the lesions were cystic and filled with purulent material, justifying the clinical impression of epidermal cyst. Histology revealed a cystic-solid tumour in the dermis (Figure 1A). The tumour cells were arranged in nests within the cyst wall. Necrosis was present (Figure 1A). The cells were crowded with scant cytoplasm and admixed with cells showing multivacuolated appearance and scalloped nuclei, characteristic of sebocytic differentiation (Figure 2B). A diagnosis of sebaceous carcinoma was made on both lesions. The patient had multiple recurrences of similar lesions on the thigh and back during a two-year follow-up. There was no regional node involvement. The patient gave a history of surgery for adenocarcinoma of the colon. Thus, it constituted Muir-Torre syndrome(MTS). The patient is currently asymptomatic.

Case 2

A 48-year-old man presented to the FNAC (fine needle aspiration cytology) clinic of our hospital one year ago with a nodular axillary mass measuring around 5 x 4 cm. The patient had no palpable or radiologically detected breast lesions. The axillary skin was puckered and the mass was palpable (Figures 2A, 2B). Fine needle aspiration of the mass showed a neoplasm with plasmacytoid morphology (Figure 2C). Biopsy revealed poorly differentiated tumour cells with a nodular architecture in the dermis. A few cells showed cytoplasmic vacuoles (Figure 2D). Immunohistochemistry was done for cytokeratin (CK) 7, CK 20, HMB-45, S-100, PSA (prostate specific antigen) and TTF-1 (thyroid transcription factor), all of which were negative. The tumour cells were positive for CK, EMA (epithelial membrane antigen) and focally for CD 15. EMA staining showed a bubbly, vacuolated cytoplasm in many cells, highlighting their sebocytic nature (Figure 3D). A final diagnosis of sebaceous carcinoma was made.

This was followed by a wide excision of the axillary mass with regional lymph node dissection. There were varying architectural patterns, such as solid nests (Figure 3A), areas with comedo-necrosis (Figure 3B), and thick cords with hyalinisation (Figure 3C). Plasmacytoid appearance, which dominated the FNAC smears, was seen only in foci. Sebocytic differentiation was also focal. Regional axillary nodal metastasis was present. The postoperative period was uneventful and the patient is currently free of disease.

Case 3

A 78-year-old lady presented to the FNAC clinic of our hospital with history of erythematous nodular and plaque-like lesions measuring 6 x 4 cm on the right anterior chest wall

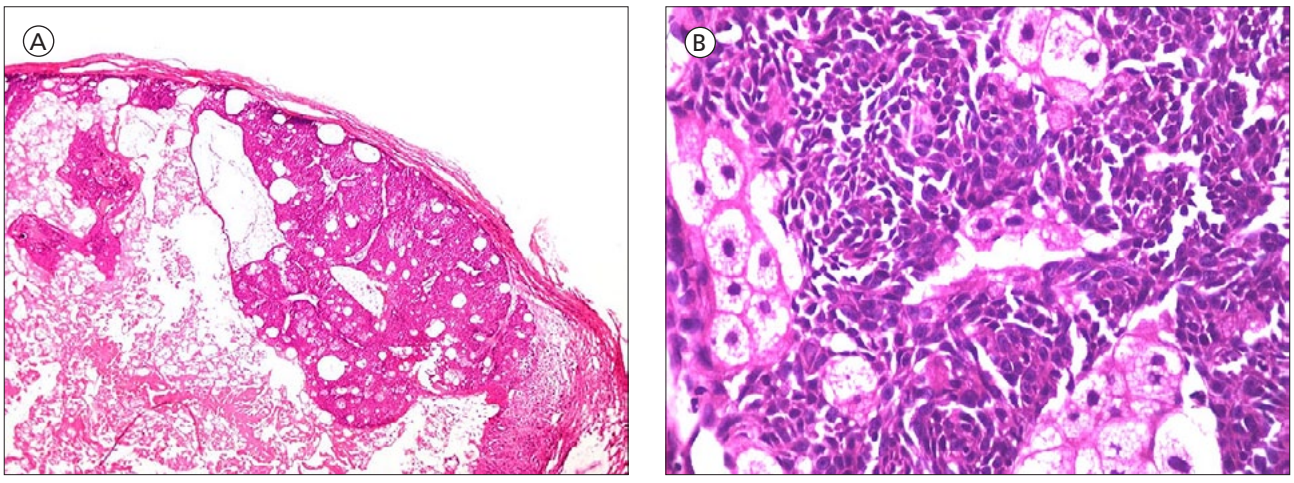


Figure 1. Cystic-solid tumour in dermis with areas of necrosis (A) (H&E, x100). Vacuolated bubbly cytoplasm of tumour cells (B) (H&E, x400). [Copyright: ©2012 Panjwani et al.]

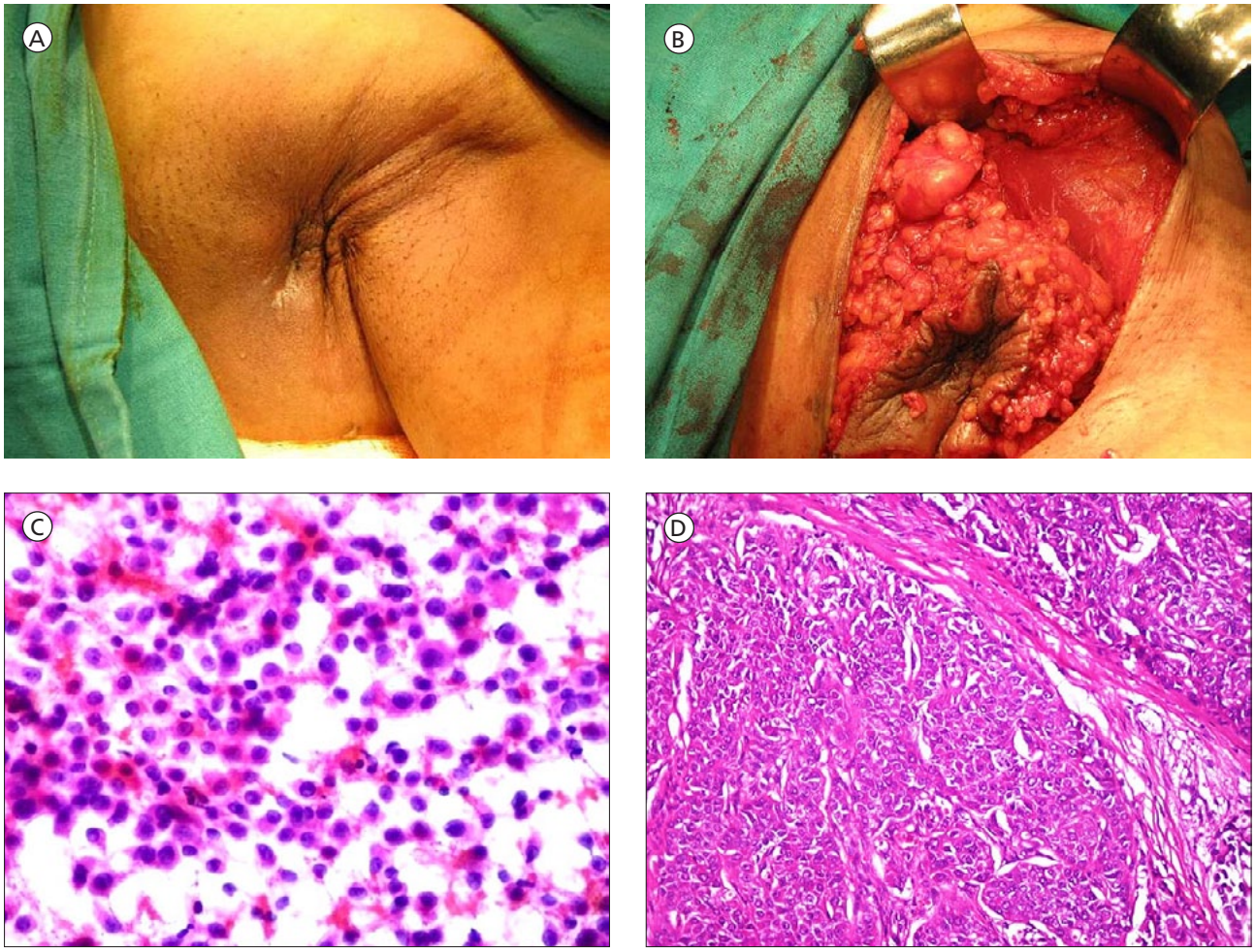


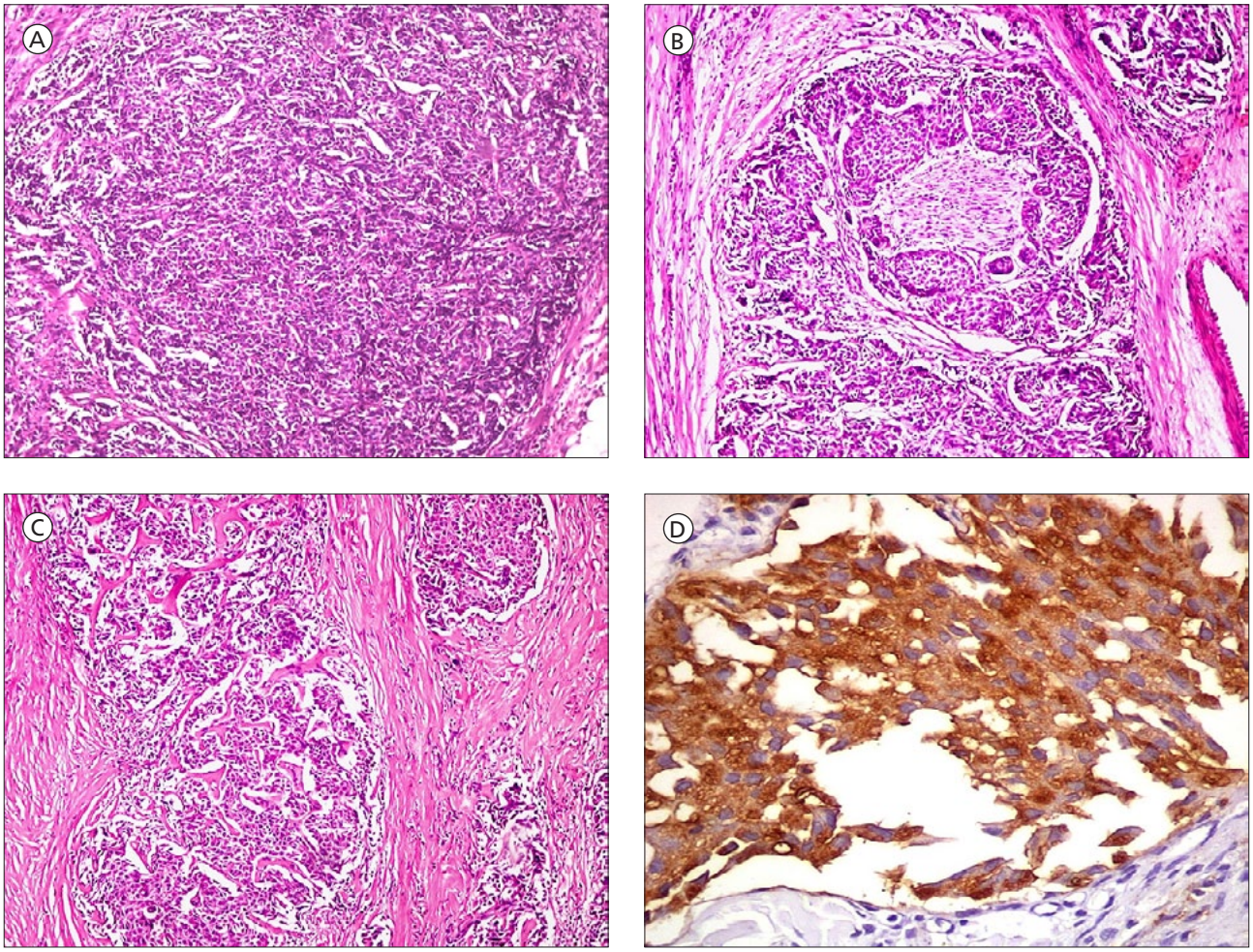
Figure 2. Nodular mass in the axilla with puckering of the skin (A+B). FNAC showing tumour cells with plasmacytoid morphology (C) (H&E, x400). Nests of vacuolated cells separated by fibrous septae (D) (H&E, x200). [Copyright: ©2012 Panjwani et al.]

(Figure 4A). She was operated on 10 years ago for similar lesions at this site. There were no palpable lesions in both breasts and mammography was negative. Fine needle aspiration from the nodule showed a poorly differentiated carcinoma (Figure 4B). A wide local excision was performed. No breast or axillary lesions were detected per operatively. A tumour was seen in the dermis with the characteristic morphology of sebaceous carcinoma (Figure 4C) and a prominent pagetoid spread (Figure 4D). Sebocytic vacuolated cells were highlighted by the EMA stain (Figure 4E).

Discussion

Extraocular sebaceous carcinomas are rare. They are common in the elderly with a slight female preponderance. They carry a high risk for local recurrence. Wide local excision with removal of regional nodes is the mode of treatment. Around 20–25% of cases show distant metastasis. The tumour mortality is around 20% [1].

The hallmark of sebocytic differentiation is the presence of multivesicular and vacuolated cytoplasm [1,2]. These variably atypical polygonal tumour cells are usually arranged as dermal nodules with a fibrovascular stroma that doesn't exhibit much desmoplasia. However, these classical features may be subtle or even absent in a few cases. A varied morphologic spectrum has been described for these tumours, like basaloid, squamoid, organoid, pseudo-neuroendocrine, etc [3,4]. This diversity may be explained by the common embryologic origin of the folliculo-sebaceous-apocrine unit being recapitulated in their neoplasms, as well. These tumours are known to be great mimics of different types of neoplasms, which often makes the right diagnosis a challenge [5]. This was demonstrated in our second case in which the tumour showed a range of cytoarchitectural features and rendered the diagnosis difficult. Immunohistochemistry confirmed the diagnosis. The vacuolated cytoplasm accentuated by the EMA stain is a typical feature of sebaceous carcinomas, along with positivity for pancytokeratin, CD 15 and CA 15.3 [1].



Figures 3. Varied morphology and patterns of the tumour cells: solid nests (A), comedonecrosis (B) and cords with hyalinisation (C) (H&E x10). IHC for EMA showing characteristic vacuolated cytoplasm in tumour cells (D) (EMA, x400). [Copyright: ©2012 Panjwani et al.]

The site of the tumour in the second case was also more in favour of a breast primary than other neoplasms. Breast carcinoma needs to be ruled out in axillary masses. Ductal carcinoma of the breast shows a positive stain for CK-7, unlike our case. It should be noted that primary sebaceous carcinoma of the breast is extremely rare in comparison to an extraocular sebaceous carcinoma arising from the skin. A few cases of a sebaceous carcinoma of the breast have been reported in patients having MTS [6,7]. Very rarely, a myoepithelial neoplasm of the breast may also show varied patterns like clear cells and a plasmacytoid morphology. However, one needs to consider the overall morphology, presence of spindle cells, mitotic activity and myoepithelial markers, if necessary, to make the distinction [8]. A metastatic tumour from other sites also needs to be ruled out in such cases, for which immunostains are very useful.

Our first case was a prototype of MTS. This is a rare autosomal dominant genodermatosis. It is characterised by the occurrence of sebaceous gland neoplasms and/or keratoacanthomas in association with a visceral malignancy, commonly of the gastrointestinal or the genitourinary tract [9–12]. Genetically, these tumours are characteristed by mutations in the mismatch repair gene products, especially MSH-2 [13]. Cystic differentiation is an important histologic clue to its association with MTS, as seen in our case. This cystic appearance is very rare in sebaceous tumours not associated with the MTS [14]. Such patients need to be screened for a concomitant GI malignancy and followed up closely. Screening of family members is desirable.

The third case that we encountered showed a pagetoid spread of the tumour. This is considered to be a "field effect" rather than being direct precursor of or extensions from an underlying sebaceous carcinoma [1]. It is particularly common in eyelid tumours. An exclusively in-situ component may rarely occur and differentiating this from an in-situ squamous carcinoma may be extremely difficult, especially if sebocytic differentiation is sparse. A thorough histologic examination is crucial.

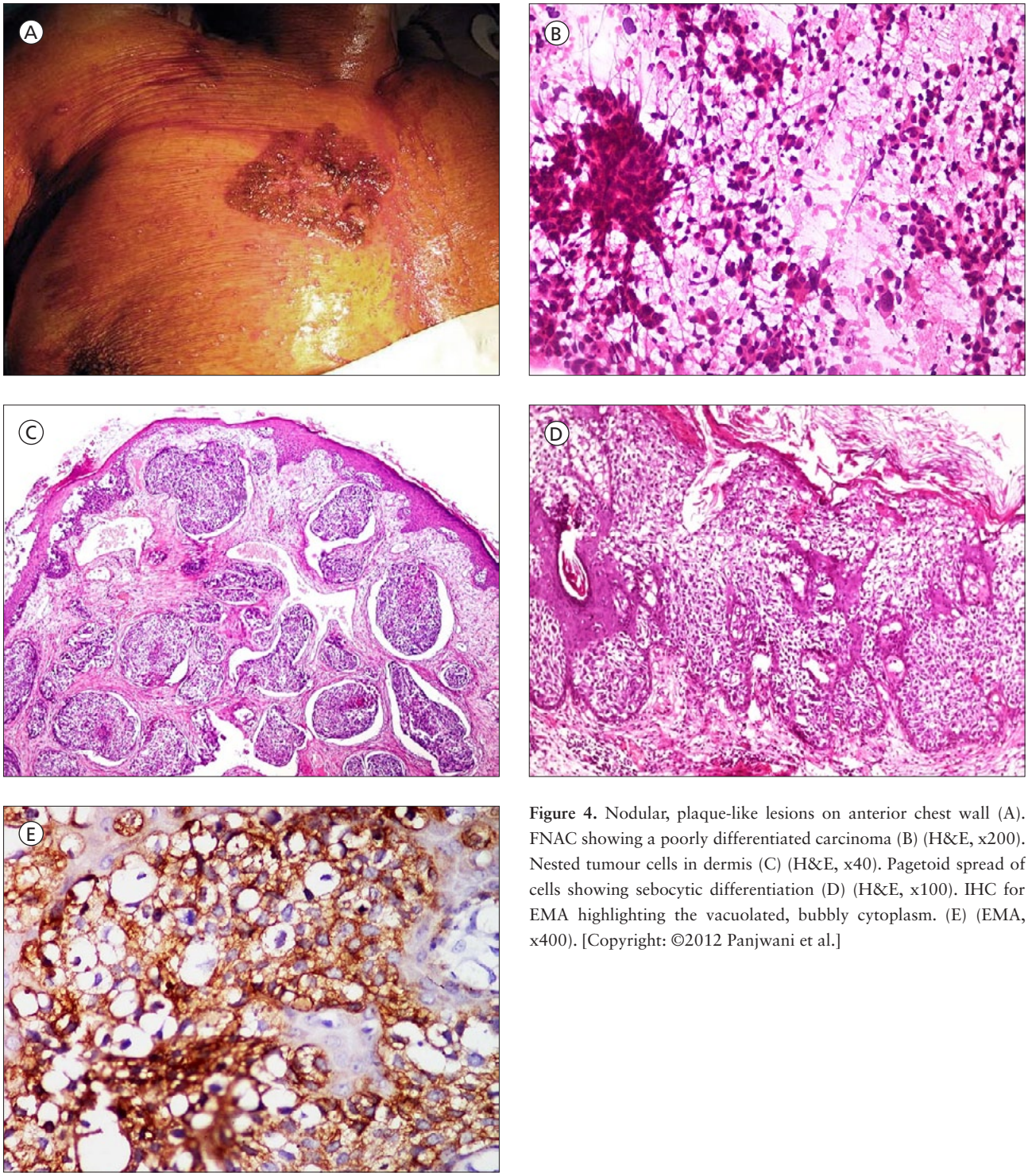


Figure 4. Nodular, plaque-like lesions on anterior chest wall (A). FNAC showing a poorly differentiated carcinoma (B) (H&E, x200). Nested tumour cells in dermis (C) (H&E, x40). Pagetoid spread of cells showing sebocytic differentiation (D) (H&E, x100). IHC for EMA highlighting the vacuolated, bubbly cytoplasm. (E) (EMA, x400). [Copyright: ©2012 Panjwani et al.]

Conclusion

The morphologic variations that can be encountered along with occurrence at unusual sites can render extraocular sebaceous carcinomas elusive to the pathologist. A high index of suspicion and a careful search for sebocytic differentiation are the key features. EMA immunostain is a useful adjunct in doubtful cases, as it highlights the multivacuolated cytoplasm. The pathologist needs to be cognisant of the histopathologic gamut of sebaceous carcinomas to avoid misdiagnosis.

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