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Case Report

Pathology Section

# Primary Cutaneous Adenoid Cystic Carcinoma: A Rare Entity

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## ABSTRACT

Primary Cutaneous Adenoid Cystic Carcinoma (PCACC) is an uncommon skin adnexal tumour that mostly affects people over 50 years and has no known gender preference. It has no apparent predilection and can appear in a variety of anatomical locations. Less than 200 instances have been reported in the literature so far. Here, we present a case report of cutaneous adenoid cystic carcinoma with unusual location. A 45-year-old male presented with synchronous painful flesh-coloured swellings on the chest and back in a local hospital. These were clinically misdiagnosed as sebaceous cysts. Wide local excision of both swellings was performed, and the samples were sent for histopathology examination. Histology of the swellings revealed an uncommon basaloid adnexal tumour, and the patient was diagnosed with PCACC. PCACC is an important histological diagnosis due to its aggressive tendency to metastasise and recur. Diagnosis of PCACC is primarily based on histopathological examinations, as there are no particular clinical distinguishing features. This case highlights the significance of histological assessment of the mass lesion for proper diagnosis and treatment, as well as thorough examination for PCACC diagnosis post-initial surgery. Here, we report a rare tumour, probably the only case in the literature showing synchronous/bifocal atypical presentation sites.

**Keywords:** Adnexal tumour, Fibrocystic swelling, Histopathology, Synchronous

## CASE REPORT

A 45-year-old male presented at a local hospital with painful swellings over his back and chest for three months. These swellings were associated with a gradual increase in size. There was no significant history of trauma, bleeding, discharge, or fever. His medical history was significant for diabetes mellitus diagnosed five months ago, and he was on oral hypoglycaemics. The patient was also a chronic tobacco chewer for 12 years. There was no history of other comorbidities.

The physical examination done at the local hospital revealed a 5×5 cm flesh-coloured cystic swelling located near the right scapular region. This swelling was hard in consistency, non-mobile and no local rise of temperature or discharge was noted. The chest showed a 3×3 cm fibrocystic swelling. This was firm in consistency, and non-mobile without local rise of temperature or discharge. The clinical diagnosis was suggestive of a 'sebaceous cyst' or 'carbuncle'. Following this, local excision of both swellings was done and the specimen was sent to HBTMC and Dr RN Cooper Municipal General Hospital for further histopathology.

On gross examination, the chest and back swellings were single, globular masses measuring 3×2×1.5 cm and 4×3×2 cm. On the cut section of the chest lesion, grey, white, and haemorrhagic firm areas were observed, while the skin-covered lesion on the back showed a cystic space filled with haemorrhagic material [Table/Fig-1].

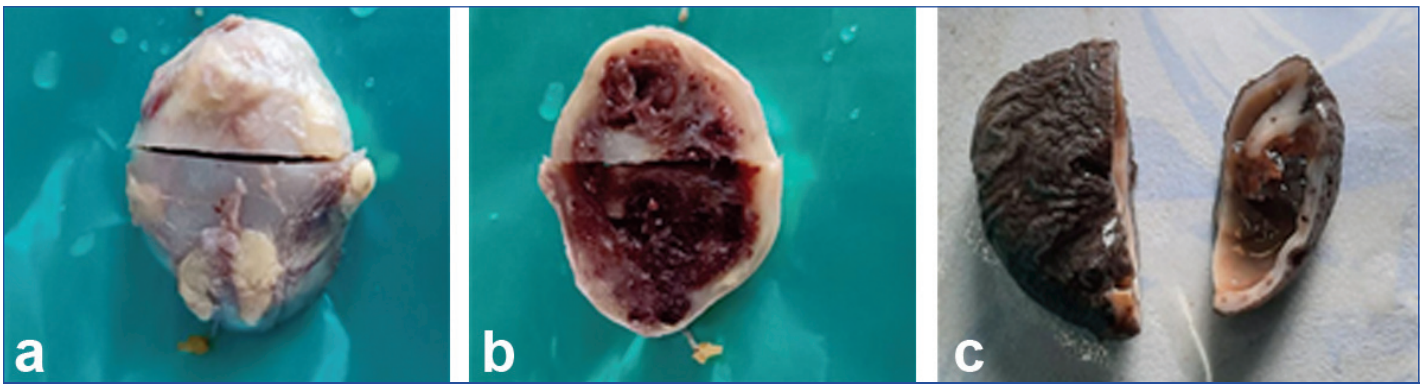
On microscopy, both the swellings showed similar histology and showed a dermal-based basaloid neoplasm arranged in cribriform, trabecular, glandular, pseudocyst and nested pattern [Table/Fig-2,3]. These glandular spaces were surrounded by basaloid cells with intraluminal basement membrane-like material and the extracellular matrix [Table/Fig-4]. The tumour seemed to invade the overlying subcutaneous tissue and muscles [Table/Fig-5]. Large areas of hyalinisation were seen. The base was free of tumour and perineural invasion was absent.

The final histopathology was suggestive of PCACC.

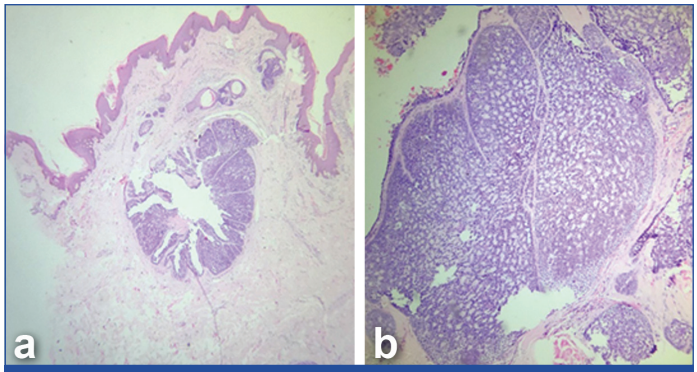
No further chemotherapy or radiation was given to the patient. The patient was followed up for 3, 6, and 9 months post-excision and has not developed any more similar lesions or recurrence.

## DISCUSSION

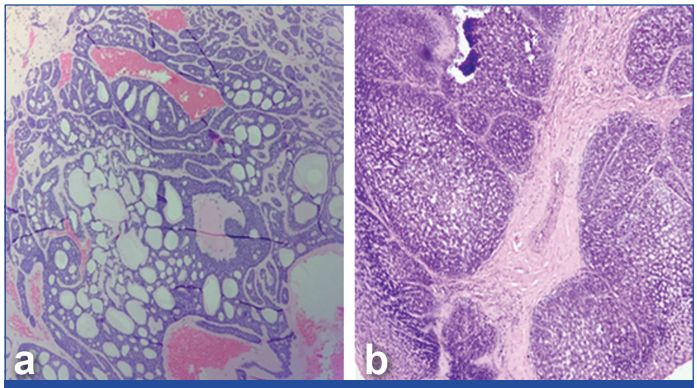
The PCACC is an uncommon tumour with only 450 cases reported in the literature, first described by Boggio in 1975, with an unclear aetiology. It most frequently involves the head and neck region, particularly the scalp (34%). Other infrequent locations include abdomen (11%), upper and lower limbs (10%), back (7%) and genitalia (5%) [1-3]. This case, most probably to the best of our knowledge, is the only case of PCACC showing infrequent location and synchronous primary lesions, i.e., the back and the chest.



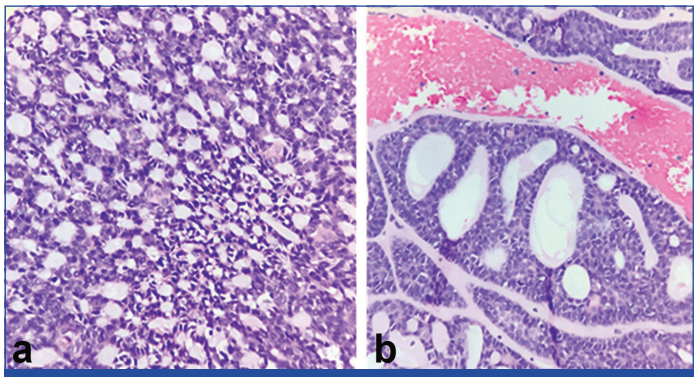

**[Table/Fig-1]:** a,b) Grey white globular mass from the chest. Cut-section shows firm, haemorrhagic, grey, white areas; c) Skin-covered lesion on the back. Cut-section shows a cystic space filled with haemorrhagic material.




**[Table/Fig-2]:** a,b) Epidermis with dermal based basaloid neoplasm (H&E, 10x).




**[Table/Fig-3]:** a,b) Basaloid tumour arranged in cribriform, trabecular, glandular, pseudocyst and nested pattern with hyalinised, paucicellular stroma (H&E, 20x).



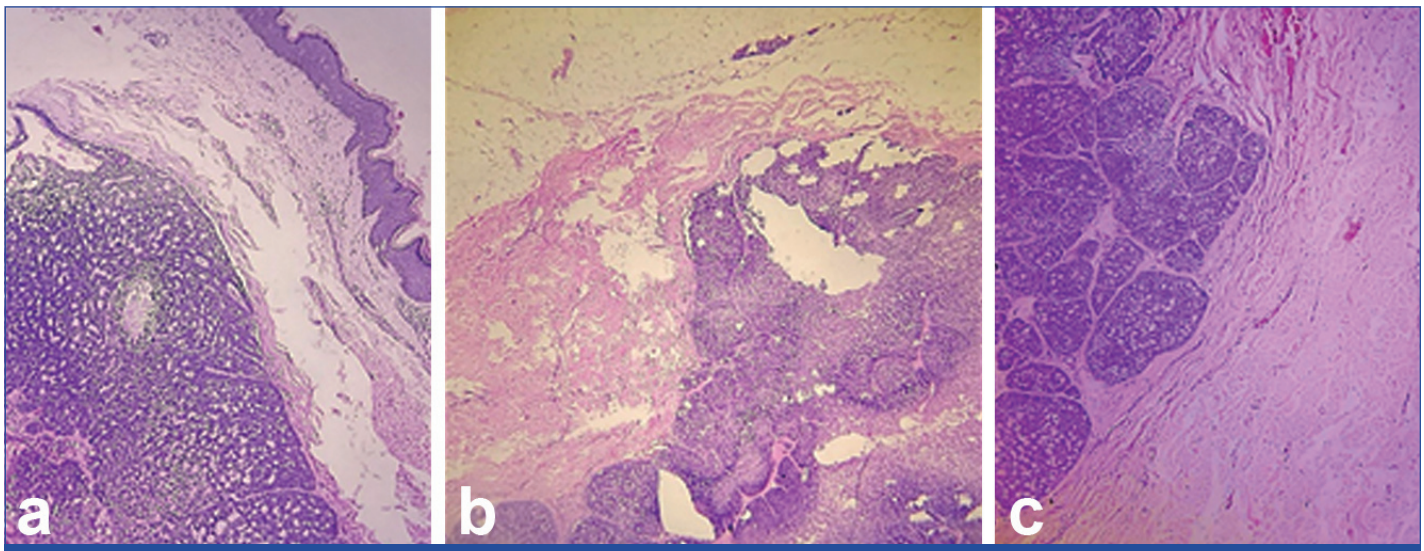

**[Table/Fig-4]:** a,b) These glandular spaces are lined by bilayered inner epithelial and outer basal cells with intraluminal basement membrane-like material and the extracellular matrix (H&E, 40x).

PCACC usually affects the seventh decade of life, with the mean common age reported to be 59 years and has equal male and female distribution, though women have a slightly higher predominance (nearly 53%). Most of the patients remain asymptomatic and others may exhibit localised hair loss, discomfort, or, in rare cases, pruritus [4-6]. The most common treatment involves wide local excision. Recurrence is noted in 44% of cases with an average follow-up period of 58 months, which complicates the clinical course [2].

The current case showed a median age group of 45 years, like other studies. While Soughi M et al., Mokhtar J et al., and Themnithikul B et al., discovered the erythematous nodule on the common location on the scalp, Oualla K et al., and Lee SJ et al., found the erythematous lesion at an unusual location on the trunk [7-11]. Similarly, the current case study showed the presence of flesh-coloured lesions on the trunk synchronously at two uncommon locations. The histology of the excised tumour in all the studies showed the presence of basaloid tumour arranged in cribriform, trabecular, glandular, pseudocyst and nested pattern, intraluminal basement membrane-like material and mucin. Perineural invasion was seen in most of the studies in contrast to the current study. Immunohistochemical (IHC) studies by Soughi M et al., confirmed the diagnosis of PCACC utilising PS100, CD117, CK7, SOX10 antibodies in endoluminal and basaloid cells, like usage of Epithelial Membrane Antigen (EMA) and Carcinoembryonic Antigen (CEA) by Mokhtar J et al., and Themnithikul B et al., [8,9]. However, the current study confirmed PCACC on the basis of histology without using IHC studies. All the above studies on follow-up up including the current case study, showed no recurrence or new lesions. This contrasted with the case reported by Oualla K et al., which showed recurrence of axillary nodule post prior surgical wide local excision. This outlined the importance of routine follow-up of these tumours post resection as they have a tendency for late recurrence [7-10].

The origin of PCACC is highly debatable. While some authors favour an eccrine origin, an apocrine origin has also been proposed because of reports of tumours that are identical to those of the apocrine ceruminous glands of the external ear meatus, which do not have eccrine glands. However, the histologic appearance of tubular and cribriform structures, IHC staining, and lack of continuity with the epidermis all support an eccrine origin [3].

Certainly, the clinical diagnosis remains challenging due to its numerous differential diagnoses. Histologically, as well, PCACC may mimic a lot of pathological entities. The differential diagnosis includes Cylindroma, Adenoid Basal Cell Carcinoma (ABCC), Mucinous Apocrine Carcinoma (MC), Primary Cutaneous Cribriform Apocrine Carcinoma (PCCAC) and metastatic cutaneous adenoid cystic carcinoma (ACC) [3,5]. Although all the lesions closely resemble PCACC on microscopy, histological examination remains the gold standard for its diagnosis.




**[Table/Fig-5]:** a,b) The basaloid neoplasm involves the dermis with no epidermal connection and is seen to invade the underlying subcutaneous tissue (5b) (H&E, 10x); c): Tumour arranged in cribriform, pseudocyst and tubular pattern, invading the muscular layer. (H&E, 20x).

PCACC usually involves the dermis and has no connection with the overlying epidermis. On histology, it appears as a poorly circumscribed tumour composed of predominantly basaloid cells arranged in glandular, cystic, cribriform patterns with pseudocysts filled with mucin and tubular pattern [3]. The background stroma is loose fibromucinous. The nuclei of the tumor cells are hyperchromatic, but there is no obvious nuclear atypia. Modified myoepithelial cells, frequently with noticeable basement membrane material, encircle true lumina. The tumour displays an infiltrating pattern invading the entire dermis and often involving the underlying subcutaneous tissue [5,10,11]. The epithelial cells have scant cytoplasm with basophilic and small cells. Mitosis is rarely found in the tumour. Perineural invasion is often present in nearly 50% of the cases and vascular invasion is rare. This perineural invasion is associated with increased recurrence risk [5,10,11]. IHC of PCACC reveals strong EMA positivity along with S-100 and CEA [5,8,11]. This is especially helpful in ruling out the other differentials of PCACC.

Metastasis of PCACC is rare; however, the common sites of metastasis involve the lung, brain, and lymph nodes. While diagnosing PCACC, it Is crucial to rule out the possibility of metastatic disease to the skin from other primary locations, and several primary skin tumours, including the adenoid-cystic variant of basal cell carcinoma, mucinous carcinoma, dermal cylindroma, and the PCCAC [Table/Fig-6] [6].

| S. No. | Differential diagnosis | Microscopy |
|---|---|---|
| 1 | Adenoid Basal Cell Carcinoma (ABCC) | • ABCC is composed of large, irregular islands with palisading edges and adenoid spaces showing sulphated mucin.<br>• Arranged in cribriform, tubules, cord, and nodular patterns composed of basaloid cells. associated with an epidermal connection<br>• Lack of dual cell (epithelial and basaloid) type morphology and absence of perineural invasion<br>• Immunoperoxidase stains positive |
| 2 | Mucinous Apocrine Carcinoma (MC) | • Presence of vast pools of basophilic mucin with thin fibrovascular septa surrounding islands of eosinophilic tumour cells with cuboidal nuclei.<br>• The mucin in MC is sialomucin |
| 3 | Cylindroma | • Islands consist of an inner group of giant cells with localised ductal differentiation and vesicular nuclei, and an outside palisading cell type<br>• Basaloid cells form a jigsaw puzzle appearance with a thick basement membrane<br>• No Lymphovascular Invasion (LVI) /Perineural Invasion (PNI) seen |
| 4 | Metastatic Adenoid Cystic Carcinoma (ACC) | • Clinical history present: Salivary ACC is highly metastatic with frequent local recurrence |

**[Table/Fig-6]:** Differential diagnosis of PCACC.

ABCC and PCACC share similar morphology, having similar cribriform, tubules, cord, and nodular patterns composed of basaloid cells. However, ABCC is composed of somewhat large, irregular islands with palisading edges and adenoid spaces showing sulfated mucin [1,2,11]. ABCC is usually associated with an epidermal connection unlike PCACC. However, key histological characteristics that aid in distinguishing between the two include the lack of dual cell (epithelial and basaloid) type morphology, peripheral palisading, retraction artifact, origin from hair follicle or overlying epidermis in adenoid BCC, and usually the absence of perineural invasion [1]. Immunoperoxidase stains may be useful in differentiating between ABCC and ACC. EMA, S-l00 protein, and CEA are found in ACC but are typically lacking in ABCC [1]. Another close differential of PCACC is MC differentiated from it by the presence of vast pools of basophilic mucin with thin fibrovascular septa surrounding islands of eosinophilic tumour cells with cuboidal nuclei. The mucin in MC is sialomucin, unlike hyalomucin of PCACC [2,5]. Histologically, cylindromas are different from PCACC since they do not have an invasive growth pattern or perineural invasion. Cylindroma tumour is composed of similar cells as PCACC but lacks the cribriform pattern and the perineural invasion. Cylindroma islands consist of an inner group of giant cells with localised ductal differentiation and vesicular nuclei, and an outside palisading cell type [1]. In this context, primary cutaneous cribriform apocrine carcinoma, a rare low-grade tumour, is also significant. It is a non-encapsulated dermal tumour formed by multiple interconnected basophilic epithelial cells. In contrast to cutaneous ACC, true elongated tubules are observed, but no basement membrane material deposition is seen, and neoplastic cells have pleomorphic rather than monomorphous nuclei. Basophilic aggregations and the spaces within are frequently more varied in size and shape. There is no sign of intravascular or perineural invasion. Additionally, there is no perineural invasion or basement membrane material present [2]. ACC has to be differentiated from metastasis or direct extension of salivary gland ACC. Consequently, the diagnosis of cutaneous ACC can only be confirmed by the absence of any history or present evidence of ACC from an extracutaneous source, and the distinction between cutaneous and extracutaneous ACC can only be made based on clinical grounds. This is extremely important since salivary gland ACC is an aggressive tumour that usually results in death from both local recurrence and extensive metastases, but cutaneous ACC usually progresses slowly despite a high risk of local recurrence.

The prognosis of PCACC is generally good, with a 5-year survival rate of around 96%. Factors affecting prognosis include male gender, age less than 50 years, and location involving the head and neck region compared to the trunk.

Long-term follow-up for recurrence is indicated, as is wide surgical excision with at least 2 cm tumour-free margins to lower the chance of local recurrence, in which perineural invasion plays a role following resection.

Numerous examples have involved the use of Mohs micrographic surgery. Radiotherapy and chemotherapy can be used as adjuvant treatments, but no studies have demonstrated that radiation decreases the risk of local recurrence. To treat patients with distant metastases, chemotherapy has been employed.

## CONCLUSION(S)

In conclusion, PCACC is a rare tumour with synchronous lesions being an unusual presentation of this entity, not previously reported. Our investigation into PCACC demonstrates the undeniable benefit of careful histological examination in determining the true kind of cutaneous tumour. In dermatological oncology, Primary ACC is a rare but challenging condition. Because of its propensity for misdiagnosis, a multidisciplinary approach combining dermatologists, dermatopathologists, and oncologists is crucial. It is essential to make an accurate diagnosis using histological analysis, IHC, and thorough investigations to exclude out secondary localisations, histopathology being the mainstay for diagnosing these tumours. Long-term follow-up is still necessary because of the risk of late recurrences, even though the favourable prognosis is achieved through effective therapy.

## REFERENCES

[1] Seab JA, Graham JH. Primary cutaneous adenoid cystic carcinoma. J Am Acad Dermatol. 1987;17(1):113-18. Doi: 10.1016/s0190-9622(87)70182-0.
[2] Tiwari R, Agarwal S, Sharma M, Gaba S. Primary cutaneous adenoid cystic carcinoma: A clinical and histopathological mimic: A case report. Oral Maxillofac Surg Cases. 2018;4(4):175-79. Doi: 10.1016/j.omsc.2018.09.002.

[3] Dores GM, Huycke MM, Devesa SS, Garcia CA. Primary cutaneous adenoid cystic carcinoma in the United States: Incidence, survival, and associated cancers, 1976 to 2005. J Am Acad Dermatol. 2010;63(1):71-78. Doi: 10.1016/j.jaad.2009.07.027.
[4] Ramakrishnan R, Chaudhry IH, Ramdial P, Lazar AJ, McMenamin ME, Kazakov D, et al. Primary cutaneous adenoid cystic carcinoma. Am J Surg Pathol. 2013;37(10):1603-11. Doi: 10.1097/PAS.0b013e318299fcac.
[5] Naylor E, Sarkar P, Perlis CS, Giri D, Gnepp DR, Robinson-Bostom L. Primary cutaneous adenoid cystic carcinoma. J Am Acad Dermatol. 2008;58(4):636-41. Doi: 10.1016/j.jaad.2007.12.005.
[6] Prieto-Granada CN, Zhang L, Antonescu CR, Henneberry JM, Messina JL. Primary cutaneous adenoid cystic carcinoma with MY Baberrations: Report of three cases and comprehensive review of the literature. J Cutan Pathol. 2016;44(2):201-09. Doi: 10.1111/cup.12856.
[7] Soughi M, Elloudi S, Ardigo M, Baybay H, Mernissi F. Primary cutaneous adenoid cystic carcinoma: A misdiagnosed tumour. Cureus. 2024;16(7):e64318.
[8] Mokhtar J, Akbarpoor F, Oghanna NG, Bennett NJ. Primary cutaneous adenoid cystic carcinoma arising in the scalp: A diagnostic challenge. BMJ Case Rep. 2025;18(3):e260654-4. Doi: 10.1136/bcr-2024-260654.
[9] Temnithikul B, Rungrunanghiranya S, Limtanyakul P, Jerasuthat S, Paige DG. Primary cutaneous adenoid cystic carcinoma of the scalp: A case report, immunohistochemistry and review of the literature. Skin Health Dis. 2022;2(2). Doi: 10.1002/ski2.118.
[10] Oualla K, Acharfi N, Benbrahim Z, Arifi S, Bouhafa T, Hassouni K NM. Primary cutaneous adenoid cystic carcinoma: A case report and literature review. Cutis. 2006;77(3):157-60. Doi: 10.24966/CRTS-310X/100008.
[11] Lee SJ, Yang WI, Kim SK. Primary cutaneous adenoid cystic carcinoma arising in umbilicus. J Pathol Transl Med. 2016;50(4):322-24. Doi: https://doi.org/10.4132/jptm.2015.11.24.

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