CASE REPORTS

PRIMARY CUTANEOUS ADENOID CYSTIC CARCINOMA OF THE CHEST WALL WITH AXILLARY LYMPH NODE METASTASES AND ITS MANAGEMENT: A CASE REPORT

Klein Dantis*[1], Radhakrishna Ramchandani[2], Shamendra Anand Sahu[3], Vandita Yogendra Singh[4], Subhajit Dasgupta[5]

[1] Assistant Professor, Department of Cardiothoracic Surgery, All India Institute of Medical Sciences, Raipur, India
[2] Associate Professor, Department of General Surgery, All India Institute of Medical Sciences, Raipur, India
[3] Assistant Professor, Department of Burns and Plastic Surgery, All India Institute of Medical Sciences, Raipur, India
[4] Associate Professor, Department of Pathology, All India Institute of Medical Sciences, Raipur, India
[5] Junior resident, Department of Nuclear Medicine, All India Institute of Medical Sciences, Raipur, India

* **Corresponding author:** drkleindantis@yahoo.com

Abstract

Primary cutaneous adenoid cystic carcinoma (PCACC) is a rare form of adenoid cystic carcinoma (ACC) arising commonly from the salivary gland. Less often they originate outside the head and neck region, with the scalp being the commonest cutaneous site in 40% of the cases. The presentation on the chest wall is rare, with no reports to date on axillary lymph node metastases. Here we report a case of a 65-year-old female with previously operated PCACC of the chest wall at a different center, showing uptake on positron emission tomography imaging at the site of surgical scar that w as inconclusive on needle biopsy metastasized to the axillary lymph node confirmed by needle biopsy managed with wide local excision, axillary lymph node dissection, and chest wall reconstruction with keystone island flap. The postoperative outcome was uneventful with no recurrence or axillary complications at one year's follow-up. She was advised to receive adjuvant radiotherapy; however, she refused. To conclude, though PCACC is rare, they can have an aggressive presentation, and a multidisciplinary approach is necessary for a better outcome.

Keywords: *Primary tumor; adenoid cystic; lymph node; chest wall; reconstruction*

INTRODUCTION

Primary cutaneous adenoid cystic carcinoma (PCACC) is a rare form of adenoid cystic carcinoma (ACC) arising commonly from the salivary gland[1]. ACC rarely originates outside the head and neck region with commonly reported sites including breast, skin, cervix, and lung[2]. Among the cutaneous site, the scalp is the most common site occurring in 40% of the cases[3]. The presentation on the chest wall is rare with no reports to date on lymph node metastases[3]. Here we present a rare case of previously operated PCACC of the chest wall at a different center, showing uptake on positron emission tomography (PET) imaging at the site of surgical scar that was inconclusive on needle biopsy, metastasized to the axillary lymph node confirmed by needle biopsy managed with wide local excision, axillary lymph node dissection, and chest wall reconstruction with keystone island flap.

CLINICAL CASE

A 65-year-old female presented with a surgically operated scar over the right posterior chest wall for two months duration. She gave a history of the right posterior chest wall swelling excision at a different center diagnosed to have adenoid cystic carcinoma two months back. On clinical examination, an oblique surgical scar measuring 5 x 1 cm on the previously operated site over the right posterior chest wall and ipsilateral single palpable axillary lymph node was seen. She was hemodynamically stable with a regular blood profile, and normal liver and renal function tests. Her chest x-ray posterior-anterior view was unremarkable, while computed tomography detected an irregular lesion measuring 0.9 x5.2x2.3cms over the posterior chest wall with an enlarged right axillary lymph node. Her PET revealed irregular soft tissue density lesion measuring 0.9 x5.2x2.3cms with a maximum standardized uptake value (SUVmax) uptake of 2.6 at the site of previous surgery and 18 F-fluorodeoxyglucose (FDG) avid single right axillary lymph node measuring 3.5x 2 cm with SUVmax uptake of 5.6 suggestive of metastatic node disease (figure 1a, 1b). Fine needle biopsy from the right axillary lymph node revealed cutaneous appendageal tumor-ACC, while that from the scar site was inconclusive. Her bronchoscopy was normal with negative bronchoalveolar lavage findings. She underwent wide local excision of the lesion with a 2cm margin, right axillary lymph node dissection followed by reconstruction of the defect by keystone island flap under general anesthesia (figure 2a, 2b, 2c, 2d). Histopathology of the gross specimen did not detect any abnormality (figure 3a). Single enlarged lymph node lesion revealed metastatic adenoid cystic carcinoma (figure 3b). Tumor cells were positive for S-100 (figure 3c) and carcinoembryonic antigen was not contributory with no detected mutation. The postoperative course was uneventful with the healing of the flap site by primary intention (figure 3d). She was to receive adjuvant radiotherapy, but she refused to receive it. The one-year follow-up did not detect any surgical site recurrence or axillary complications.

DISCUSSION

Boggio first described PCACC in 1975[1]. They are apocrine in origin, affecting the elderly population with equal incidence in both sexes, as seen in our case[3]. They have an indolent course with a high frequency of local recurrence and rarely metastasize to the lung and lymph nodes, resulting in a poor prognosis 4. The characteristic histological feature is the basaloid cells with angulated hyperchromatic nuclei and scanty cytoplasm arranged into three patterns: cribriform, tubular, and solid[2]. Excessive secretion of mixed lipids and proteins, including proteoglycans, by secretory cells has led to cellular differentiation and metaplasia[2]. Perineural invasion, a characteristic finding in ACC in non-cutaneous variants, is rarely seen in PCACC[3].

Recently, c-Kit mutation association studies have shown poor prognosis, while epidermal growth factor expressions are associated with better prognosis[4]. Abnormal lymphatic communication explains the mechanism for lymph node metastasis; hence, lymph node status is the most important predictor of survival[5]. Surgery followed by radiotherapy provides the best

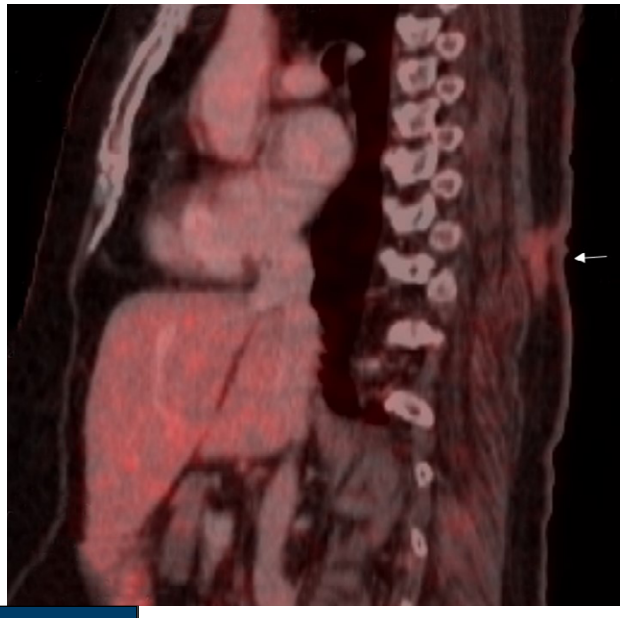

Figure 1a *Positron emission tomography (PET) scan image showing uptake at the surgical site (marked with an arrow).*

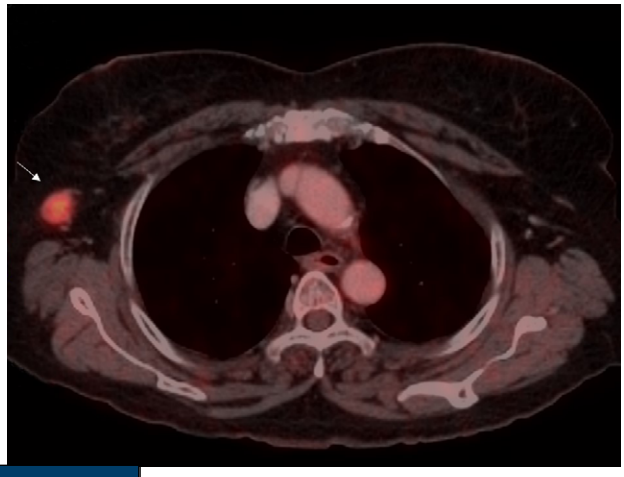

Figure 1b *PET scan image showing right axillary lymph node uptake (marked with an arrow).*

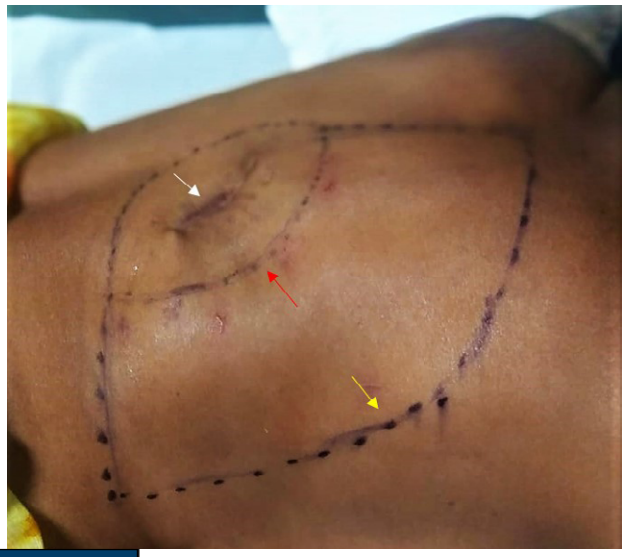

Figure 2a *Right posterior chest wall showing surgical scar (white arrow), wide local excision margin marking (red arrow) and keystone island flap marking (yellow arrow) preoperatively*

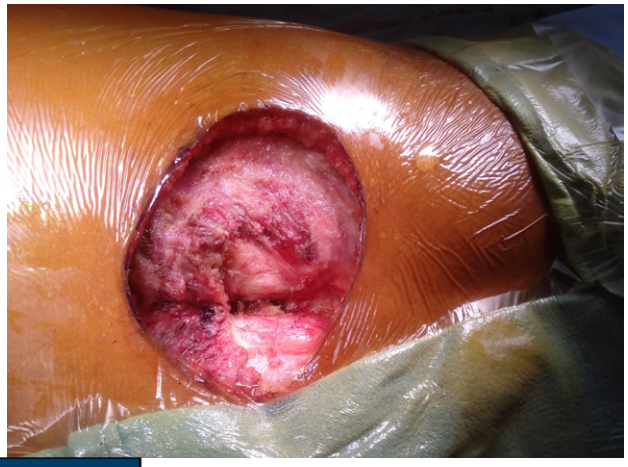

Figure 2b *Surgical site following wide local excision*

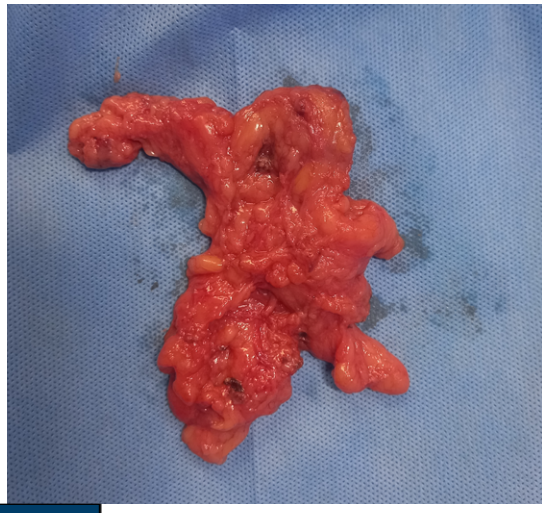

Figure 2c *Axillary lymph node dissection specimen*

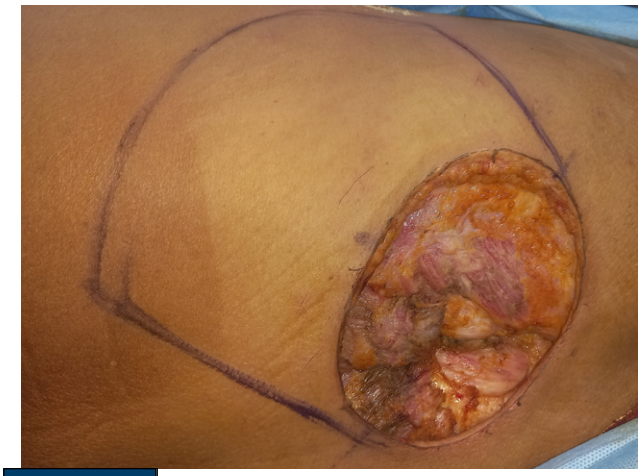

Figure 2d *Keystone island flap marking following wide local excision for chest wall reconstruction*

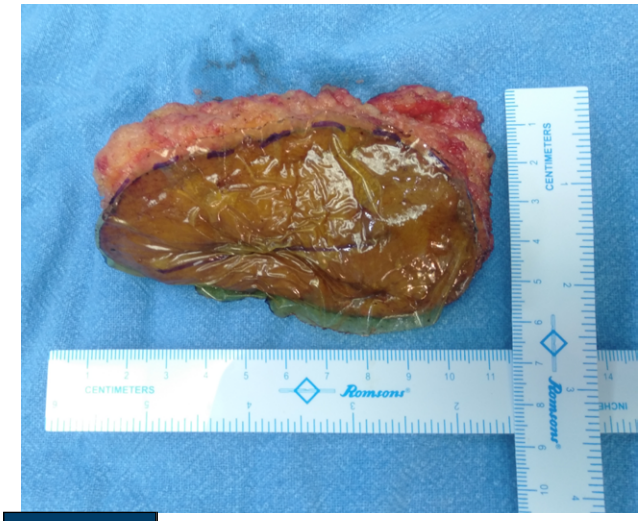

Figure 3a *Gross specimen of the lesion with wide local excision*

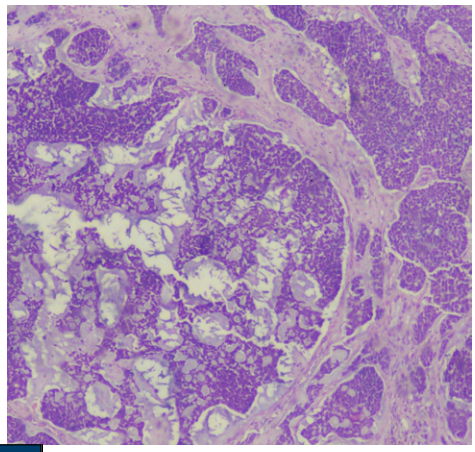

Figure 3b *Histopathology showing tumor cells arranged in nest and cribriform pattern with hyalinised globules and myxoid change having hyperchromaticmoulded medium sized nuclei with indistinct cytoplasmic borders (Hematoxylin and Eosin staining, 100X magnification)*

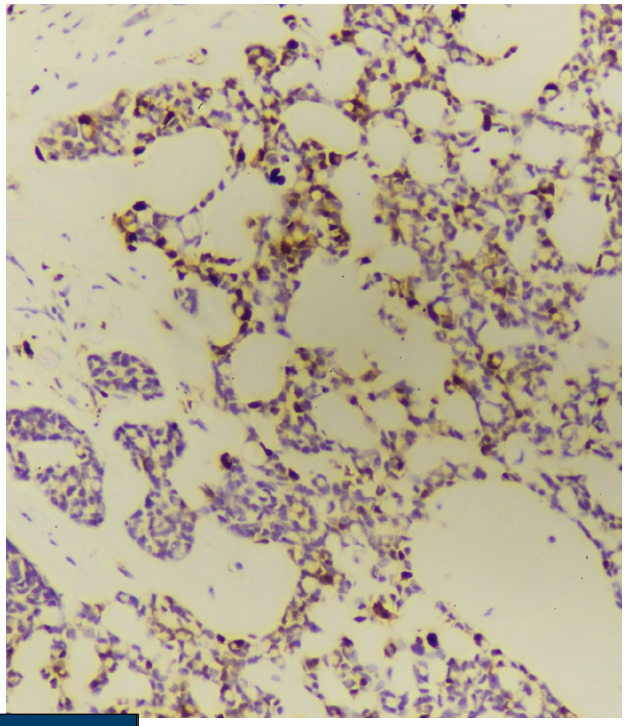

Figure 3c *S 100 protein positivity of myoepithelial cells on immunohistochemistry (100X magnification)*

3d

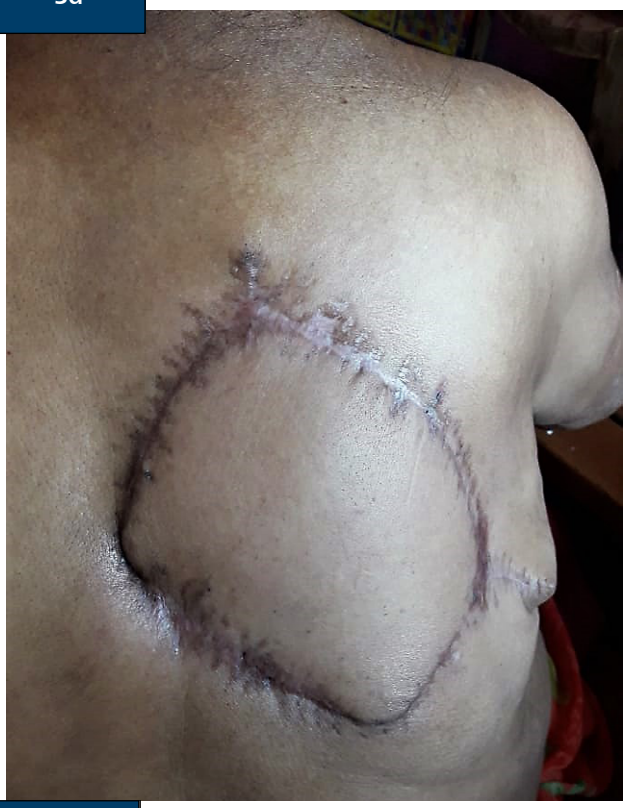

Figure 3c *Postoperative surgical site, healed by primary intention*

survival of treatment with ACC, including chest wall[4]. Surgical excision included a wide local excision with a 2 cm margin[3]. However, axillary lymph node dissection, previously not reported for PCACC of the chest wall, is mandatory for axillary lymph node metastasis. The role of adjuvant chemotherapy therapy is still unclear. However, adjuvant radiation therapy is recommended to prevent a locoregional recurrence while chemotherapy is not offered as it tends to be resistant[1, 4]. Various reconstructive techniques for chest wall reconstruction have been described in the literature. We opted for the keystone island procedure, a rarely cited solution that grants a considerable amount of cutaneous and muscular tissue with low morbidity.

CONCLUSION

PCACC, though rare, can have an aggressive presentation and requires a multidisciplinary approach for a better outcome.

No funding received.
Authors declare no conflicts of interest.
Inform consent obtained from the patient for the purpose of publication.

REFERENCES

1. 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 AcadDermatol. 2010;63(1):71-8. doi: 10.1016/j.jaad.2009.07.027.
2. Moskaluk CA. Adenoid cystic carcinoma: clinical and molecular features. Head Neck Pathol. 2013 ;7(1):17-22. doi: 10.1007/s12105-013-0426-3.
3. Raychaudhuri S, Santosh KV, SatishBabu HV. Primary cutaneous adenoid cystic
4. carcinoma of the chest wall: a rare entity. J Cancer Res Ther. 2012;8(4):633-5. doi: 10.4103/0973-1482.106583.
5. Tirelli G, Capriotti V, Sartori G, Tofanelli M, Marcuzzo AV. Primary adenoid cystic carcinoma of the frontal sinus: case description of a previously unreported entity and literature review. Ear Nose Throat J. 2019;98(4):E8-E12. doi: 10.1177/0145561319837881
6. Sekine C, Kawase K, Yoshida K. Sentinel lymph node biopsy of primary apocrine sweat gland carcinoma of the axilla: A case report and review of the literature. Int J Surg Case Rep. 2020;77:122-125. doi: 10.1016/j.ijscr.2020.10.067.