

Primary cutaneous adenoid cystic carcinoma of the scalp: dermatosurgical approach with favourable outcome

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Abstract

Although described as early as 1975 as a distinct, rare form of cancer with diverse localization, primary cutaneous adenoid cystic carcinoma (PCACC) remains a mystery and challenge for both clinicians and pathologists. The clinical presentation cannot be clearly distinguished from amelanotic melanoma or intradermal nevus, Merkel cell carcinoma, trichofolliculoma, trichoepithelioma or other rare tumors of the adnexa, or dermatofibrosarcoma protuberans. The histopathological diagnosis requires not only careful evaluation of standard hematoxylin/eosin preparations, but also immunohistochemical staining with a number of markers such as epithelial membrane antigen (EMA), S-100, SOX-10, Ki-67, CD-117 (c-kit), Vimentin, carcinoembryonic antigen (CEA), Ber-EP4 and many others. The surgical approach should consist of excision with margins between 1 and 2 cm, with the choice of margins depending upon the histopathological findings in the primary excisional specimen. We present a 31-year-old patient with an enlarging, amelanotic, plaque-like tumor of the scalp with a duration of no more than 18-24 months. Surgical treatment was performed within two surgical sessions with a total resection field of 1.3 cm. A good cosmetic result was achieved.

Introduction

Primary cutaneous adenoid-cystic carcinoma (PCACC) of the scalp is a rare tumor affecting middle-aged to elderly individuals.¹ Characteristic of the clinical man-

ifestation of the tumor is its slow growth in the form of a nodule, with dimensions ranging between 0.5 and 9 cm.^{2,3} The tumor was first described by Boggio in 1975, and to date (45 years later), fewer than 100 cases have been reported.⁴ Characteristic of the neoplasm is the tendency for local, relatively slow growth, but also a tendency for local postoperative recurrence and - very rarely - the development of distant metastases.⁵ According to a number of authors, the tendency for local recurrence is mainly associated with certain histopathological characteristics of the neoplasm, namely, infiltrative growth in the reticular dermis and subcutaneous tissue, often accompanied by perineural invasion.^{6,7}

The pathogenesis of PCACC remains a mystery, and the presence or absence of certain predisposing factors is also unclear or controversial.

Case Report

A 31-year-old, otherwise healthy, female reported to the dermatology department with primary complaints of pain and discharge on the scalp. She first noticed swelling at the site 1-2 years prior, with gradual increase in size and with occasional episodes of pain. Dermatological examination revealed a well-circumscribed nodular, oval-shaped cutaneous lesion measuring 3 cm in greatest diameter and having a grey-tan appearance (Figure 1a-b). The nodule had a soft, cystic feel on palpation and appeared to be endophytic in growth. A complete blood work up was conducted and was without abnormalities. A native-head CT with IV contrast showed an atypical lesion extending to the bone under the scalp. There were no signs of infiltration or destruction of bony structures within the vicinity of the lesion. All other parameters appeared normal on the imaging study. The clinical differential diagnosis included amelanotic nevus/ melanoma, Merkel cell carcinoma, or cyst. The patient underwent a narrow surgical excision with 2 mm margins in all directions (Figure 1a-c). The defect was closed with nylon sutures (Figure 1d-f). The histopathology consisted of neoplastic cells in the form of nests with cribriform arrangements consisting of multiple mucin-filled cystic spaces (Figure 2a). These tumor cells were seen within the dermis and subcutaneous tissue in infiltrative patterns and composed of monomorphous round, basoid and polygonal cells localized to the centers of the islands (Figure 2b-d, Figure 3a-d). The dept of invasion was 5.0 mm

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with no perineural or lymphovascular invasion identified and staged at T1Nx. Based on the clinicopathological findings, a diagnosis of adenoid cystic carcinoma was considered and a re-excision with 1.0 cm margins in all directions was performed, without neoadjuvant radiation (Figure 4a-4b). Further samples were sent for immunohistochemical (IHC) analysis; with tumor cells appearing positive for SOX10 and Ki67 (10 % of tumor cells positive), supporting the diagnosis. S-100 staining was negative. Re-excisional samples analyzed by histology and IHC showed no tumor cells at the surgical margins. A 2-week follow-up following the reexcision showed good evidence for wound healing (Figure 4a), and the patient showed no signs of clinical or radiological recurrence at week 8 follow-up (Figure 4b).

Discussion

Adenoid cystic carcinoma is a tumor of the major and minor salivary glands.⁸ It can also occur in the area of the lacrimal glands, the external ear canal, the respiratory tract, the breast, the cervix and other locations.⁹ Among primary cutaneous examples of the tumor, the scalp has been reported to be a relatively common site of origin, wherein tumor infiltration mainly involves the dermis and subcutaneous adipose tissue.^{8,9} The tumour is also seen to grow exophytically, with the calvarium acting as a natural barrier that impedes tumour infiltration downward into deeper tissue.^{8,9} On the other

hand, it should be noted that the limited skin mobility in the scalp area may lead to difficulties with surgical intervention; as a result, the postoperative results when treating large tumor formations or tumor recurrences may be less than satisfactory from an aesthetic standpoint.

Characteristic of the histopathological presentation in PCACC is the presence of epithelial cells in the form of multiple lobules, many of which show cystic characteristics as well as cribriform arrangements within the tumor.⁹ Basophilic mucinous material is found in the cystic lobules, and they are separated from each other by a fibrous stroma.⁹

There is evidence that CEA, EMA, and S-100 immunostains can be useful in distinguishing PCACC (often positive for these markers) from a potential histologic mimic, adenoid basal cell carcinoma (typically negative).¹⁰ The utility of EMA and S-100 staining in this scenario has been confirmed by other authors, though results with CEA and S-100 have been variable in PCACC.¹¹

SOX-10 is one of the modern immunohistochemical markers widely used in the diagnosis and differential diagnosis of salivary gland tumors.¹² Positive expression has been observed in adenoid cystic carcinomas, epithelial-myoeplithelial carcinomas, myoeplithelial carcinomas, and pleomorphic



Figure 1. a-b) Oval, well-circumscribed, erythematous nodule of between 2.8-3 cm in diameter. Clinical image of the lesion with additional inscription of 2mm surgical margins in the shape of an elliptical incision. c-e) Intraoperative image with excised lesion and closure of defect. f) Complete closure of defect with single nylon sutures.

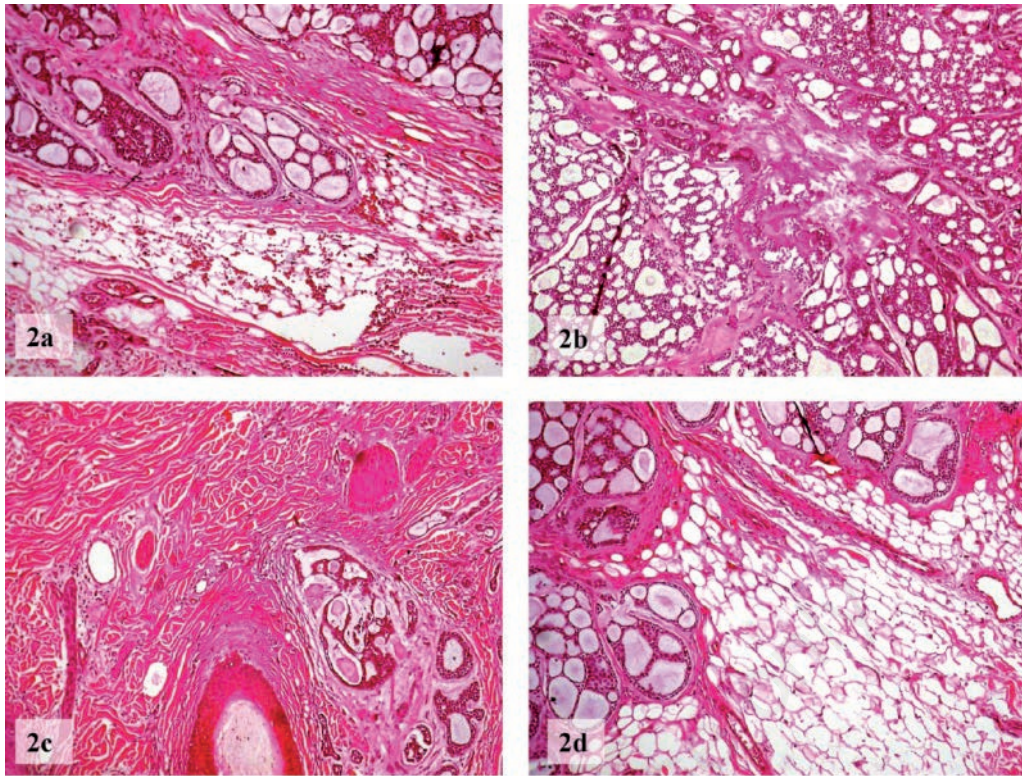


Figure 2. a) Epithelial neoplasm composed of nodules and nests with cribriform pattern due to the presence of multiple small mucin-filled cystic spaces. b) Infiltrative pattern with invasion of the subcutaneous tissue. c) Epithelial cords and nodules surrounding a hair follicle and arrector pili muscle. d) Detail showing small cribriform nodules composed of monomorphous round epithelial cells without significant cytological atypia.

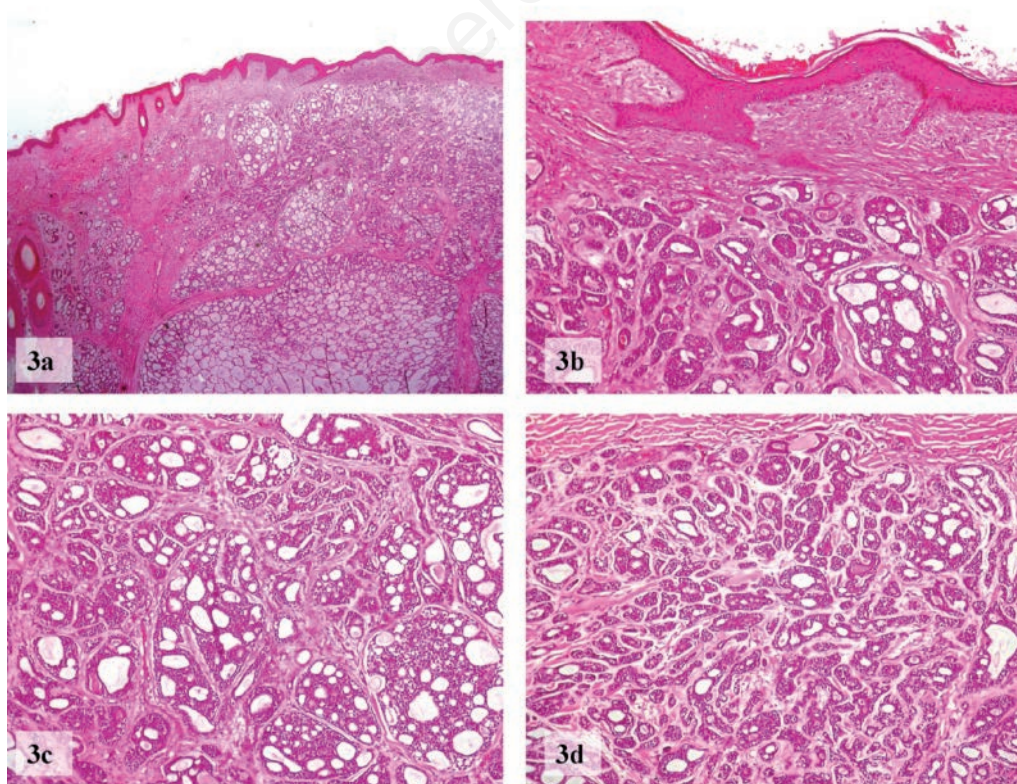


Figure 3. a,b) Subcutaneously-located tumor formation of adenoid cystic carcinoma of the scalp. The tumor lacks contact with the epidermis. There is infiltration of the reticular dermis by multiple tumor complexes. c,d) The typical histological picture of the tumor with the characteristic multiple lobules with a cribriform arrangement, separated by a fibrous stroma.

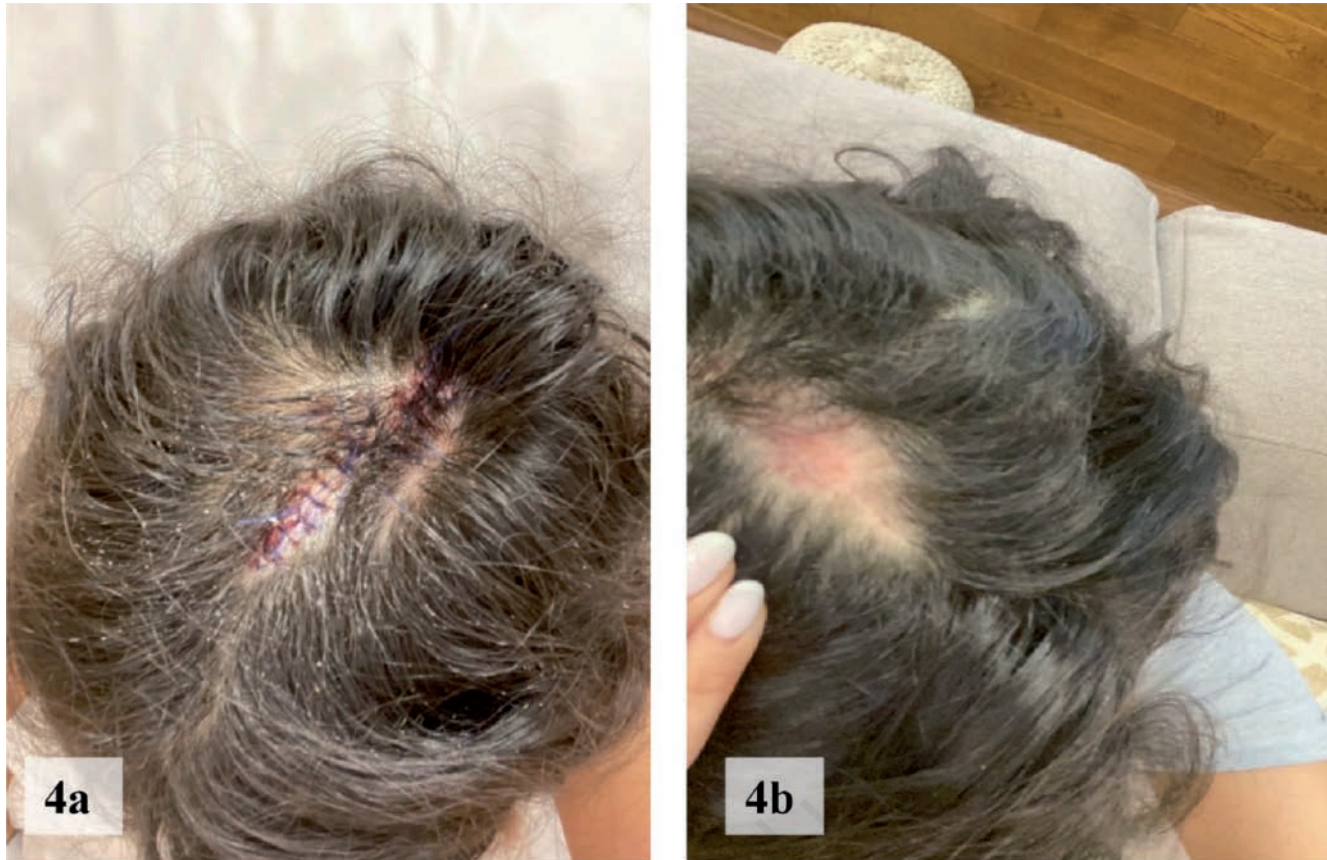


Figure 4. a) Postoperative 4-week follow up showing favorable signs of wound healing. b) Follow-up 8 weeks post-surgery with complete wound healing.

adenomas, including the pleomorphic adenoma component of carcinoma.¹²

Its positivity has also been documented in PCACC.¹³ SOX-10 is also a specific and sensitive marker for cutaneous melanoma.¹⁴ SOX-10 is a nuclear transcription factor with an important role in melanocytic differentiation.¹⁴

The negative finding for S-100 in combination with the positive for SOX-10 in the patient described by us argues against a diagnosis of melanoma and at the same time supports the diagnosis of PCACC of the scalp. There is also literature that supports the negative expression of S-100 in adenoid cystic carcinomas localized in the oral cavity.¹⁵ Comprehensive histopathological evaluation in these cases should be a priority and should be mainly supported by standard hematoxylin/eosin staining.

The standard treatment for adenoid cystic carcinoma consists of complete surgical excision.¹⁶ The recommended excision margins according to the literature vary between 1 and 2 cm, and the histopathological characteristics of the tumor and, in particular, the presence or absence of perineural invasion are decisive for the choice of the

surgical field. Given that the diagnosis is virtually impossible to make clinically, patients with PCACC almost always require at least two surgical sessions in order to achieve a total resection margin between 1 and 2 cm. The choice of a smaller resection margin is a very common cause of local recurrences.¹⁶ The use of postoperative radiotherapy in order to minimize the risk of locoregional recurrences is controversial.^{16,17}

Conclusions

In conclusion, we present a patient with a rare PCACC of the scalp, successfully treated surgically within two surgical sessions with a total resection field of 1.2 cm. Due to the lack of perineural invasion and the double establishment of clean resection margins, a consensus decision was made not to employ radiation therapy in this case.

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