Giant Primary Cutaneous Adenoid Cystic Carcinoma of the Perineum: Histological and Radiological Correlations

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Sir,

Adenoid cystic carcinoma (ACC) is widely known as a malignant neoplasm of the head and neck region, especially of the salivary glands. It may also occur in the lacrimal glands, the mucosal glands of the upper respiratory tract, the oesophagus, the breast, the uterine cervix, and at other sites (1). Primary cutaneous adenoid cystic carcinoma (PCACC) is a particularly rare variant of ACC and was first described by Boggio (2) in 1975. As most PCACCs reported previously have been located in the head and were very small, their radiological features have not been documented. There is only one previous report of a PCACC in the perineum, but it contains no radiological and histological information. In this report, we present a case of a giant perineal PCACC that had haemorrhagic and multilocular characteristics, and we discuss the relationship between its magnetic resonance imaging (MRI) characteristics and its histology.

CASE REPORT

A 79-year-old woman with a 6-year history of a subcutaneous nodule of the perineum was referred to our hospital. The patient mentioned that the nodule had enlarged rapidly during the previous 3 months and that she had dysuria. Physical examination revealed a protruding, brownish, elastic, soft nodule of the perineum measuring about 15×11×10 cm (Fig. 1a). The patient had a history of hypertension, premature ventricular contraction, and was a hepatitis B e-antigen carrier. T1-weighted (T1W) MRI revealed 3 subcutaneous encapsulated lesions of high signal intensity in the perineum and buttock. Each capsule comprised many small cysts with septa, separated by fluid. T2-weighted (T2W) imaging demonstrated that the multilobular cysts had high intensities in the ventral aspect and low intensities in the dorsal aspect (Fig. 1b and c). The nodule image was homogeneously enhanced with contrast medium.

The results of blood analyses, including levels of tumour markers such as carcinoembryonic antigen (CEA) and cancer antigen-125, were within normal limits. Fine-needle aspiration samples consisted mainly of erythrocytes and neutrophils. The excision was performed under general anaesthesia. The tumour did not adhere to the vagina or rectum. The blood loss was about 1100 g, and the postoperative course was uneventful.

Histopathological analysis showed well-demarcated tumour clusters in the dermis and subcutaneous fat tissue. The tumour clusters consisted of alveoli of various sizes surrounded by fibrous interstitium (Fig. 1 d and e). The tumours were mainly of cribriform appearance and consisted of basaloid cells and small cysts with myxoid or hyalinized matrixes. Partially luminal lesions were also observed. Invasion of peripheral nerves

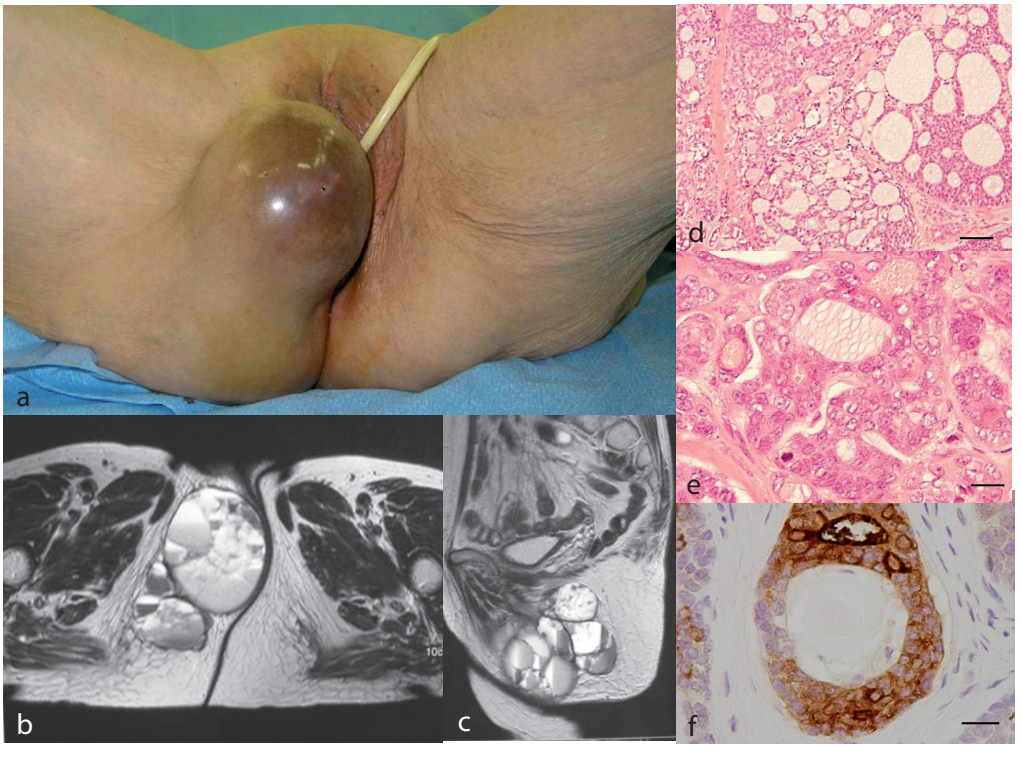

Fig. 1. (a) A protruding, brownish, elastic, soft nodule, measuring about 15×11×10 cm was present in the perineum. The patient complained of dysuria. (b) T2-weighted magnetic resonance images showing 3 subcutaneous encapsulated lesions of various signal intensities in the perineum and buttock, and (c) high signal intensities in the ventral aspect and low signal intensities in the dorsal aspect. (d) The tumour extended from the dermis through to the perineal adipose tissue and was composed mainly of alveoli of various sizes that were surrounded by fibrous interstitium in a cribriform pattern (haematoxylin and eosin (H&E) stain; scale bar=250 μm). (e) Small cysts containing myxoid or hyalinized matrixes are evident (H&E; scale bar=50 μm). (f) Luminal tumour cells reacted positively to epithelial membrane antigen (scale bar=50 μm).

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was not observed. The intraluminal material was positive for periodic acid-Schiff (PAS) staining and Alcian blue staining. Luminal tumour cells were positive for epithelial membrane antigen (EMA; Fig. 1f) but negative for CEA, S100 protein, and α smooth muscle actin. Thus, the diagnosis was PCACC. We recommended subsequent chemoradiotherapy as a precaution against recurrence, but the patient declined this treatment because of her advanced age. No clinical or radiological recurrence was evident at a 4-month follow-up.

DISCUSSION

ACC is an uncommon but well-recognized neoplasm of the major and minor salivary glands. It also occurs in the lacrimal gland, the respiratory tract, the external auditory canal, the breast, the cervix, and at other sites (1, 3). PCACC, a rare variant of ACC, was first described in the scalp (2). Only 20 cases have been reported subsequently. PCACC occurs mainly in the scalp, where it is usually located in the dermis and the subcutaneous fat. PCACC tumours are composed of epithelial cells arranged in multiple lobules, many of which show cystic characteristics and therefore impart a cribriform appearance to the tumour. Basophilic mucinous material is present within the cystic lobules, and the stroma between the lobules is fibrous. In addition to cribriform areas, tubular foci may be present (4). Although Wick & Swanson (5) reported that PCACCs are mostly positive for CEA, EMA, and S100 protein antigens, the reactivity of tumours to specific antigens is controversial (4).

The report of Seab & Graham (3) constitutes the only previous documented case of perineal PCACC. The report mentioned tumour recurrence within 8 months of excision, but provided little information. In our patient, the tumour was an exophytic giant nodule with a smooth, brownish surface. Palpation of the tumour under the dermis, which had thinned, indicated that several nodules were present in a botryoidal (resembling a bunch of grapes) pattern.

The radiological characteristics of PCACC have not been described previously. In MRI scans of lesions of the head and neck, ACCs generally show low signal intensities on T1W images. On T2W images, the masses may show high or low signal intensities (6–8). Tumours that show high intensities on T2W images have low cellularity and luminar or cribriform patterns, and are associated with a good prognosis (6). In contrast, tumours that show low intensities on T2W images have high cellularity, are solid, and are associated with a poor prognosis. In our patient, the dorsal part of the mass showed low intensity on the T2W image and had high cellularity, but the ventral part showed high intensity on the T2W image and had a tubular and cribriform histological pattern. Hemorrhagic areas were mainly confined to the ventral part of the mass. Thus, this tumour displayed the characteristics of both types of ACC, possibly because it contained several large multilocular lesions and consisted of a large amount of tissue.

The material within the lumen consisted mainly of haemorrhaged blood cells and a mucin-like substance. Although the sizes of the lumens varied somewhat, most were very large. These findings differ from those of previous reports of scalp PCACCs. Although perineural invasion is common in scalp PCACCs, vascular invasion is uncommon (5, 9, 10). Tumours of the scalp show exophytic progress because the hard calvarian bone impedes downward proliferation, but perineal masses can progress in an upward or downward direction because perineal and buttock tissue normally contains abundant soft fat tissue. Because perineal tissue has a rich vascular supply, rupture of the walls of the cysts and consequent haemorrhaging may have been caused by pressure exerted by the patient when sitting. Although the skin appendages, hair, and glands of the perineum are well developed (11), these organs were squashed by the mass, and the skin had thinned to the extent that the normal characteristics of perineal skin were absent.

The standard treatment for ACC is wide local excision with tumour-free margins established by permanent section (1). Simple excision may result in frequent recurrence. Although ACCs of the salivary glands grow slowly, the local recurrence rate is very high (50%) (3, 12). In contrast, the rate of metastasis of PCACC is 10%, which is much lower than that of extracutaneous ACC (3). However, there are too few reported PCACC cases and their follow-up times are too short to determine whether this difference is caused by differences in the biological behaviours of the tumours (3). There is currently no consensus regarding the optimal treatment for PCACC because of its low frequency. We consider that minimal surgery is the best option for elderly patients because morbidity is associated with wide excision, and the rate of metastasis of PCACC is less than that of extracutaneous ACC.

REFERENCES

1. Krunic AL, Kim S, Medenica M, Laumann AE, Soltani K, Shaw JC. Recurrent adenoid cystic carcinoma of the scalp treated with mohs micrographic surgery. Dermatol Surg 2003; 29: 647–649.
2. Boggio R. Letter: Adenoid cystic carcinoma of scalp. Arch Dermatol 1975; 111: 793–794.
3. Seab JA, Graham JH. Primary cutaneous adenoid cystic carcinoma. J Am Acad Dermatol 1987; 17: 113–118.
4. Fueston JC, Gloster HM, Mutasim DF. Primary cutaneous adenoid cystic carcinoma: a case report and literature review. Cutis 2006; 77: 157–160.
5. Wick MR, Swanson PE. Primary adenoid cystic carcinoma of the skin. A clinical, histological, and immunocytochemical comparison with adenoid cystic carcinoma of salivary glands and adenoid basal cell carcinoma. Am J Dermatopathol 1986; 8: 2–13.
6. Sigal R, Monnet O, de Baere T, Micheau C, Shapeero LG, Julieron M, et al. Adenoid cystic carcinoma of the head and

neck: evaluation with MR imaging and clinical-pathologic correlation in 27 patients. Radiology 1992; 184: 95–101.
7. Aikawa H, Tomonari K, Okino Y, Hori F, Ueyama T, Suenaga S, et al. Adenoid cystic carcinoma of the external auditory canal: correlation between histological features and MRI appearances. Br J Radiol 1997; 70: 530–532.
8. Kaku A, Kiyosue H, Matsumoto S, Mori H, Kashima K, Miyakawa I. Radiological features of adenoid cystic carcinoma of the uterine cervix. Eur Radiol 2005; 15: 975–977.
9. Chang SE, Ahn SJ, Choi JH, Sung KJ, Moon KC, Koh JK. Primary adenoid cystic carcinoma of skin with lung metastasis. J Am Acad Dermatol 1999; 40: 640–642.
10. Cooper PH, Adelson GL, Holthaus WH. Primary cutaneous adenoid cystic carcinoma. Arch Dermatol 1984; 120: 774–777.
11. McKee PH, editor. Pathology of the skin. 2nd edn. London: Mosby-Wolfe; 1996: p. 1–21.
12. Matsuba HM, Spector GJ, Thawley SE, Simpson JR, Mauney M, Pikul FJ. Adenoid cystic salivary gland carcinoma. A histopathologic review of treatment failure patterns. Cancer 1986; 57: 519–524.