Case Report

Ductal Eccrine Carcinoma with Bony Metastasis: A Case Report

Pichaya Thanindratarn MD[1], Puwadol Veerapun MD[2], Thipachart Punyaratabandhu MD[2], Phutsapong Srisawat MD[2]

[1] Department of Orthopedics, Chulabhorn Hospital, Bangkok, Thailand

[2] Musculoskeletal Oncology and Bone Metabolism Unit, Department of Orthopedics, Phramongkutklao Hospital and College of Medicine, Bangkok, Thailand

Ductal eccrine carcinoma is a rare malignant eccrine sweat gland tumor with ductal differentiation. The tumor may grow de novo or in the setting of underlying poroma. It has been found more commonly in lower extremity (44%) and trunk (24%). A few have been reported in the upper extremities (11%). One third of ductal eccrine carcinomas is fatal from distant metastasis. The authors present a case of ductal eccrine carcinoma in a 46-year-old male who developed a painful nodular lump at his right leg and painless bone lesion at left thigh. Seven months ago, he had a painless 2.5 cm nodular lump at his left axilla and the histopathology from marginal excision confirmed ductal eccrine carcinoma. The authors performed prophylaxis fixation and cementation at right tibia and left femur.

Keywords: Ductal eccrine carcinoma, Sweat gland carcinoma

J Med Assoc Thai 2018; 101 (5): 695-8
Website: http://www.jmatonline.com

Case Report

A 46-year-old Thai male presented to orthopedic department with a painful 5 cm mass at right leg for the past three months (Figure 1). Seven months ago, he developed painless 2.5 cm nodular lump at his left axilla and had an excisional biopsy that revealed malignant epithelium neoplasm. Immunohistochemical stains showed positive for cytokeratin [CK] 7, CK 5/6, CK (AE1/AE3), epithelial membrane antigen [EMA], and carcinoembryonic antigen [CEA] but negative for CK 20, GCDFP 15, TTF-1, estrogen and progesterone receptor. The diagnosis of ductal eccrine carcinoma was made but he did not return for follow-up.

At this admission, we performed the preoperative imaging including bone scan, which demonstrated an increased uptake at right tibial shaft and left femoral shaft. Plain radiographs showed osteolytic lesions at shaft of right tibia and left femur with more than fifty percent cortical involvement (Figure 2A, 3A). The computed tomography [CT] of the chest found a 1.6x1.5x1.2 cm spiculate nodular lesion at right upper lobe with pleural attachment. He refused to undergo any procedures for his lung lesion but agreed to pursue operations for his lower extremities.

Correspondence to:

Thanindratarn P. Department of Orthopedics, Chulabhorn Hospital, Bangkok 10210, Thailand.
Phone: +66-2-5766000
Email: pichaya.tha@pccms.ac.th

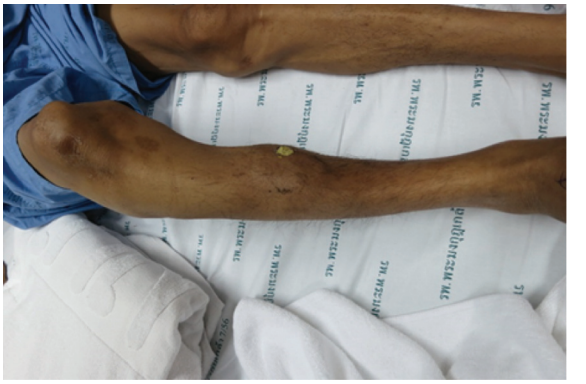

Figure 1. Clinical picture showed a painful bulging mass of 5 cm on the anterior right leg.

Core needle biopsy of his right leg showed similar pathological findings as his left axilla lump. Tumor debulking and prophylactic intramedullary nailing with cementation at right tibia and left femur was performed successfully (Figure 2B, 3B, 3C). Palliative radiation with concurrent chemotherapy was scheduled soon.

Discussion

Ductal eccrine carcinoma is a rare malignancy of the eccrine sweat glands that usually affects both genders equally in their sixth or seventh decade[1]. It may arise de novo as a primary sweat gland tumor or a malignant transformation of a longstanding

How to cite this article: Thanindratarn P, Veerapun P, Punyaratabandhu T, Srisawat P. Ductal eccrine carcinoma with bony metastasis: a case report. J Med Assoc Thai 2018;101:695-8.

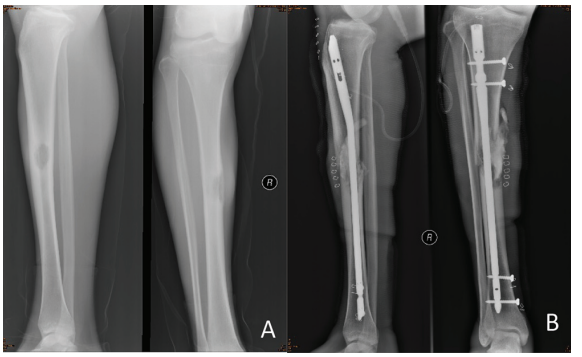

Figure 2. A) AP and lateral radiograph shows a metastatic lesion of the tibial shaft with significant bone destruction. B) Postoperative AP and lateral radiograph following surgical management with intramedullary tibial nailing and cementation.

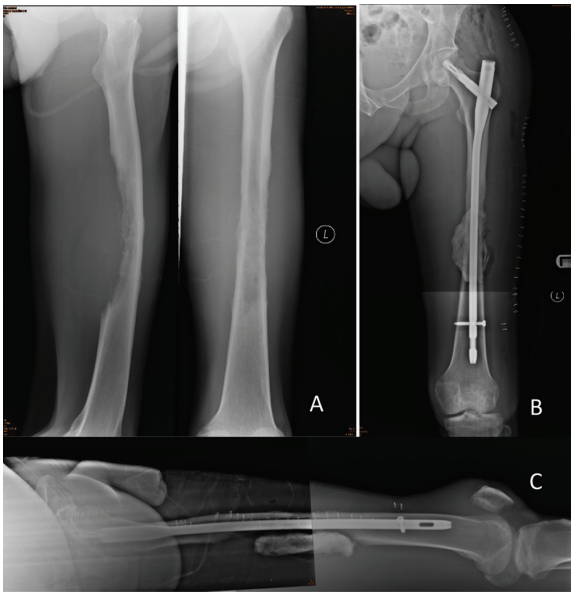

Figure 3. A) AP and lateral radiograph shows a metastatic lesion of the femoral shaft with significant bone destruction. B) Postoperative AP and C) lateral radiographs following surgical management with intramedullary femoral nailing and cementation.

eccrine poroma, a benign sweat gland tumor[2,3]. The term ductal eccrine carcinoma has been used to describe tumors that may appear in the dermis as an infiltrative poroma (porocarcinoma) or as a moderately differentiated adenocarcinoma[3].

Since it was first described by Pinkus and Mehregan[4], fewer than 300 cases have been reported[5]. Given the rarity of eccrine neoplasms (0.005% of all cutaneous carcinomas)[6], there are no prospective studies to define the natural history or to guide

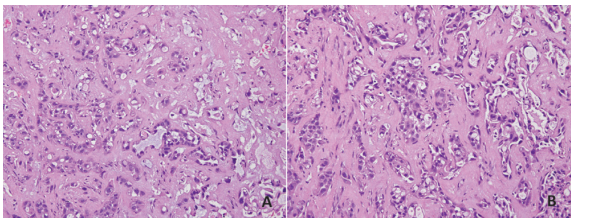

Figure 4. A, B) Low-power photomicrograph (x40) of metastatic lesion from the tibia with standard heatoxylin-eosin stain, showing a well-defined tumor nests composed of polygonal or cuboidal cells.

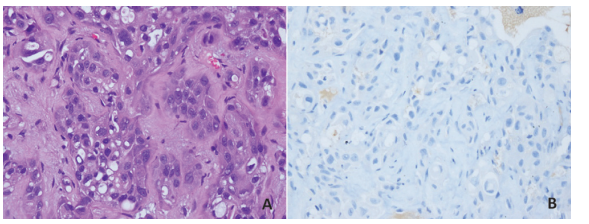

Figure 5. A) Higher magnification revealing nests of epithelial tumor cells with a significant degree of cytologic atypia and mitotic activity (hematoxylin and eosin stain, x60). B) Immunohistochemical study showing non-reactivity for thyroid transcription factor-1 [TTF-1].

the optimal management. Treatment decisions are primarily based on few retrospective case series. The tumor is most commonly involved in extremities, particularly legs and feet. The ductal eccrine carcinoma could be localized, manifesting itself as a nodule, plaque, ulcerated tumor, or polypoid/verrucous lesion that is clinically similar to squamous cell carcinoma, pyogenic granuloma, or amelanotic melanoma[3,7].

Ductal eccrine carcinoma may be associated with the underlying eccrine poroma or hidroacanthoma simplex. In such cases, we could see the benign-appearing eccrine poroma cells lying adjacent to the areas of anaplastic cells. The malignant cells generally have large, hyperchromatic, irregularly shaped nuclei, which could be multi-nucleated, with features of atypia (frequent mitoses and necrosis). Lesional cells are rich in glycogen and cystic lumina may be seen. In the primary tumor, the malignant cells may be confined to the epidermis or down into the dermis in asymmetric cribriform pattern of cords and lobules of polygonal malignant cells.

Useful clues to help identify areas with eccrine differentiation include spiraling ductal structure, ducts lined by cuticular material, zones of cytoplasmic glycogenation, as well as intraepidermal cells in discrete aggregates. The stroma may be fibrotic, hyalinized, highly myxoid, or frankly mucinous[3].

However, this histopathologic finding alone is usually insufficient to establish the diagnosis because of its low specificity[8]. Furthermore, other conditions may share similar histologic findings as well. These include squamous cell carcinoma, sebaceous carcinoma, Paget's disease, and metastatic adenocarcinoma, especially of the breast and lungs[9,10].

Ductal eccrine carcinoma has three subtypes: infiltrative, pushing and pagetoid. Robson et al found that infiltrative subtypes tend to have higher rate of recurrence. Other predictive factors of death are tumor depth greater than 7 mm, more than 14 mitoses per high-power field, and lymphovascular invasion[11].

The best management of ductal eccrine carcinoma remains controversial[12]. Most patients would undergo surgical treatment with wide excision. Using Surveillance, Epidemiology, and End Results data from the National Cancer Institute, a recent study by Martinez et al[13] examined 4,032 malignant cutaneous adnexal tumors [MCATs], including 180 malignant eccrine poromas and 210 eccrine adenocarcinomas. The authors found that wide surgical excision resulted in improved overall survival [OS] compared with alternative surgical approaches such as narrow-margin excision (OS: hazard ratio 0.78, 95% CI 0.68 to 0.89, $p = 0.001$). However, no significant difference in disease-specific survival [DSS] was noted[13]. Despite negative-margin primary excision, many patients would develop regional nodal involvement (20%) or distant metastases (10%)[7,14,15]. Nodal metastases were associated with worse DSS, whereas distant metastases predicted both worse OS and DSS[13].

Mortality rate among these patients with nodal metastases was high at 67%[8]. The authors concluded that surgical treatment should ideally include a wide resection rather than a less aggressive approach and the sentinel lymph node biopsy should be considered[13].

The adjuvant therapy for ductal eccrine carcinoma is also debatable[9]. Adjuvant radiotherapy, single- or multi-agent chemotherapy with isotretinoin, interferon alfa, methotrexate, cisplatin, bleomycin, adriamycin, paclitaxel or tegafur, and electrochemotherapy have been reported with varying responses[1,4,16-19].

To our knowledge, this was the first case of ductal eccrine carcinoma treated with negative-margin wide excision, which later developed multiple distant bone metastases. The authors reported this case to remind oncologists, orthopedists, and dermatologists of the aggressiveness of this rare cutaneous malignancy. Although the short clinical course and follow-up was the limitation in our case, we elected to pursue a therapeutic approach with surgery and radiation due to its aggressive potential for loco-regional spreading. Additional studies designed specifically for treatments are warranted to help manage this rare entity.

What is already known on this topic?

Ductal eccrine carcinoma is a rare malignant eccrine sweat gland tumor. It has been found more often in lower extremity and trunk. Few have been reported in the upper extremities. One third of ductal eccrine carcinomas is fatal from distant metastasis.

Because of the rarity of eccrine neoplasms, there are no prospective studies to define the natural history or to guide the optimal management. Treatment decisions are primarily based on a few retrospective case series. Most patients would undergo surgical treatment with wide excision and adjuvant therapies such as radiotherapy, chemotherapy, and electrochemotherapy with varying response.

What this study adds?

To our knowledge, this is the first case of eccrine porocarcinoma treated with negative-margin wide excision, which later developed to multiple distant bone metastases. We report this case to remind oncologists, orthopedists, and dermatologists of the aggressiveness of this rare cutaneous malignancy. Although the short clinical course and follow-up is the limitation in the present case, we elected to pursue a therapeutic approach with surgery and radiation due to its aggressive potential for loco-regional spreading. Additional studies designed specifically for treatments are warranted to help manage this rare entity.

Potential conflicts of interest

The authors declare no conflict of interest.

References

1. Chang O, Elnawawi A, Rimpel B, Asarian A, Chaudhry N. Eccrine porocarcinoma of the lower extremity: a case report and review of literature. World J Surg Oncol 2011;9:94.
2. Wick MR, Goellner JR, Wolfe JT 3rd, Su WP. Adnexal carcinomas of the skin. I. Eccrine carcinomas. Cancer 1985;56:1147-62.
3. Ahmed TSS, Del Priore J, Seykora JT. Tumors of the epidermal appendages. In: Elder DE, Elenitsas R, Johnson BL Jr, Murphy GF, Xu X, editors. Lever's histopathology of the skin. 10th ed. Philadelphia: Wolters Kluwer, Lippincott Williams & Wilkins; 2009:851-910.

4. Pinkus H, Mehregan AH. Epidermotropic eccrine carcinoma. a case combining features of eccrine poroma and paget's dermatosis. Arch Dermatol 1963;88:597-606.
5. Marone U, Caraco C, Anniciello AM, Di Monta G, Chiofalo MG, Di Cecilia ML, et al. Metastatic eccrine porocarcinoma: report of a case and review of the literature. World J Surg Oncol 2011;9:32.
6. Mehregan AH, Hashimoto K, Rahbari H. Eccrine adenocarcinoma. A clinicopathologic study of 35 cases. Arch Dermatol 1983;119:104-14.
7. Shaw M, McKee PH, Lowe D, Black MM. Malignant eccrine poroma: a study of twenty-seven cases. Br J Dermatol 1982;107:675-80.
8. McCalmont TH. Batman and adnexal tumors. J Cutan Pathol 2010;37:401-2.
9. Vleugels FR, Girouard SD, Schmults CD, Ng AK, Russell SE, Wang LC, et al. Metastatic eccrine porocarcinoma after Mohs micrographic surgery: a case report. J Clin Oncol 2012;30:e188-91.
10. Idrissi SK, Harmouch T, Chbani L, El Fatemi H, Sekal M, Hammas N, et al. Eccrine carcinoma: a rare cutaneous neoplasm. Diagn Pathol 2013;8:15.
11. Robson A, Greene J, Ansari N, Kim B, Seed PT, McKee PH, et al. Eccrine porocarcinoma (malignant eccrine poroma): a clinicopathologic study of 69 cases. Am J Surg Pathol 2001;25: 710-20.
12. Belin E, Ezzedine K, Stanislas S, Lalanne N, Beylot-Barry M, Taieb A, et al. Factors in the surgical management of primary eccrine porocarcinoma: prognostic histological factors can guide the surgical procedure. Br J Dermatol 2011; 165:985-9.
13. Martinez SR, Barr KL, Canter RJ. Rare tumors through the looking glass: an examination of malignant cutaneous adnexal tumors. Arch Dermatol 2011;147:1058-62.
14. Luz MA, Ogata DC, Montenegro MF, Biasi LJ, Ribeiro LC. Eccrine porocarcinoma (malignant eccrine poroma): a series of eight challenging cases. Clinics (Sao Paulo) 2010;65:739-42.
15. Shiohara J, Koga H, Uhara H, Takata M, Saida T. Eccrine porocarcinoma: clinical and pathological studies of 12 cases. J Dermatol 2007;34:516-22.
16. Barzi AS, Ruggeri S, Recchia F, Bertoldi I. Malignant metastatic eccrine poroma. Proposal for a new therapeutic protocol. Dermatol Surg 1997; 23:267-72.
17. González-López MA, Vázquez-López F, Soler T, Gómez-Diéz S, Garcia YH, Manjón JA, et al. Metastatic eccrine porocarcinoma: a 5.6-year follow-up study of a patient treated with a combined therapeutic protocol. Dermatol Surg 2003;29: 1227-32.
18. Gutermuth J, Audring H, Voit C, Trefzer U, Haas N. Antitumour activity of paclitaxel and interferon-alpha in a case of metastatic eccrine porocarcinoma. J Eur Acad Dermatol Venereol 2004;18:477-9.
19. de Bree E, Volalakis E, Tsetis D, Varthalitis Y, Panagiotidis J, Romanos J, et al. Treatment of advanced malignant eccrine poroma with loco-regional chemotherapy. Br J Dermatol 2005;152: 1051-5.