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Case Report

Giant Pendulous Carcinosarcoma – Squamous Cell Carcinoma-Type - of the Leg – A Case Report and Review of the Literature

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Abstract

Cutaneous carcinosarcoma (CCS) is a rare non-melanoma skin cancer with a biphasic growth pattern. A tumour is composed of epithelial and mesenchymal cells that show clonality. In most cases, CCS develops in the head-and-neck region on the chronic sun-exposed skin of males. Here, we describe an 80-year-old female patient who developed a giant, pendulous CCS on the leg. A tumour was surgically removed. We found no evidence of metastatic spread.

Introduction

Cutaneous carcinosarcoma (CCS) also known as sarcomatoid carcinoma is a rare non-melanoma skin cancer occurring mainly on sun-exposed skin of elderly males. The tumour is biphasic, i.e. composed of epithelial and mesenchymal elements. Various subtypes have been described such as a basal cell, pilomatrical, squamous cell, and trichoblastic [1][2].

The neoplastic cells show coexpression of keratins and vimentin – in particular, the spindle cells. Also, coexpression of p53, p16 and p63 has been reported in epithelial and spindle cells [3]. CCS display multiple copy number variations (CNVs) and copy-neutral loss of heterozygosity (CN-LOH). Furthermore, epithelial and spindle cells share the same clonality [4][5].

Here we report a case of CSS – squamous cell type – of the leg.

Case report

An 80-year-old female patient was referred to our department. The primary reason for hospital admission was an edematous swelling of the right leg and slight increase of fibrinogen to 4.96 g/L (normal range: 1.8-4.5). Duplex sonography revealed a 3-storey deep venous thrombosis of the right leg. Since the bandages had to be removed for diagnostics, a giant exophytic, pendulous, malodorous tumour became apparent. Therefore, she was referred to our department.

Her medical history was remarkable for breast cancer 1995, renal cell carcinoma 2015, and chronic lymphatic leukaemia. She suffered from type II diabetes mellitus and arterial hypertension. She had secondary lymphedema of the arm after axillar dissection 1995.

On examination, we observed a 9 cm x 7 cm large, partially ulcerated, pendulous tumour on her upper right leg (Fig. 1).

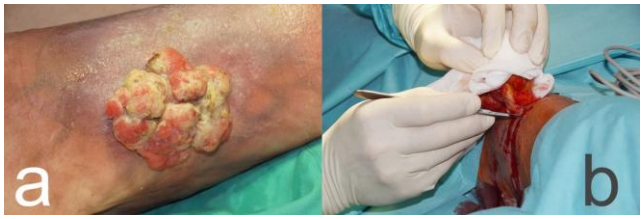

Figure 1: Clinical presentation of cutaneous carcinosarcoma of the leg (a). During surgery, the pendulous growth is apparent (b)

Laboratory findings: Leucocytes 11.71 Gpt/L (normal range: 3.8-11), erythrocytes 3.86 Tpt/L (4.2-5.4), hypochromic erythrocytes 14.6% (< 2.5%), microcytic erythrocytes 2.4% (< 1.5%), hemoglobin 6.4 mmol/L (7.4-10.7), hematocrit 0.336 (0.37-0.47), C-reactive protein 69.6 mg/L (< 5).

Imaging techniques did not reveal any metastatic spread.

Treatment was surgically excised with wide excision (2 cm safety margin) and primary closure by tissue expansion. Healing was uneventful.

Histopathologic examination of the specimen was performed. Histological examination showed a polypoid ulcerated tumour with structures of squamous cell carcinoma associated with the overlying epidermis, and beneath structures of a malignant spindle cell component in parts seeming one component transit into the other. The interlacing cords of epithelial cells extended from the epidermis and the ulcerated tumour surface to the intermediate dermis (Fig. 2a). Some of the deeper situated cords developed bulbar formations resembling glandular structures (Fig. 2b). However, ductal formations were completely missing.

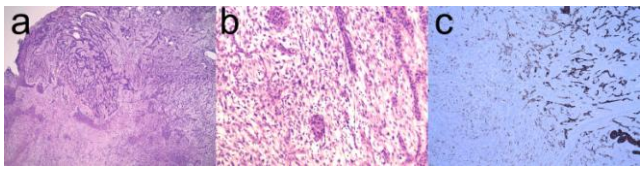

Figure 2: Histopathology of cutaneous carcinosarcoma (sarcomatoid carcinoma) of skin. (a) Overview, demonstrating the transition between epithelial and mesenchymal cells (Hematoxylin-eosin - HE x 20). (b) Detail (HE x 100). (c) Expression of CK5/6 (Immunoperoxidase x 40)

In both cellular components, immunohisto-chemistry demonstrated expression of cytokeratins (CK 5/6 and PanCK). In particular, in the spindle cell component, there was coexpression with vimentin, which was interpreted as clues to sarcomatoid dedifferentiated squamous cell carcinoma (CCS) (Fig. 2).

The patient also received low-molecular-weight heparin certoparin–sodium 8,000 U subcutaneously per day to treat the deep vein thrombosis.

Discussion

CCS is a rare tumour entity initially described by Dawson in 1972 [6]. We report a case of squamous cell type CCS on the leg of an elderly woman. The localisation on the leg is a rarely reported clinical feature since most of these tumours develop on the chronic sun-damaged skin of the head and neck region [1][2][3].

We could identify only three case reports with CCS of the leg – one in a 32-year-old female with a burn scar [7], another case of a 52-year-old female with a very rare myofibroblastic sarcomatous variant [8], and a last one of a 54-year-old male [9].

In the present case, we observed an ulcerated malodorous tumour that raised several differential diagnoses in a patient with multiple neoplastic disorders, including metastasis of breast or renal cancer, SCC, Merkel cell carcinoma, amelanotic melanoma, osteosarcoma, and rhabdomyosarcoma [10][11][12][13]. By histologic examination, a CCS of squamous cell subtype could be confirmed.

Cutaneous SCC can be associated with reactive fibroblastic proliferation. These spindle cells, however, do not co-express vimentin and keratin as seen in our case (Fig. 2c). In SCC epithelial-mesenchymal transition (EMT) is required for tumour invasion and dissemination. This is accompanied by overexpression of transcriptional factors Twist and ZEB1 [14].

Basosquamous carcinoma, also known as metatypical basal cell carcinoma (BCC), is a rare subtype of BCC. It occurs in two subtypes – mixed and intermediary. The mixed type shows focal keratinisation with a parakeratotic centre. The intermediary type is characterised by a network of narrow strands composed of an outer row of dark-staining basaloid cells and an inner layer of larger cells appearing lighter. Some of these tumours may express smooth muscle actin or myosin [15].

Cutaneous adenosquamous carcinoma is extremely rare neoplasia composed of malignant squamous and glandular cells without co-expression of keratin and vimentin. Luminal cells express cytokeratin 7. All tumour cells express cytokeratin 5/6

and p63. Cutaneous adenosquamous carcinoma is considered as a locally aggressive high-risk subtype of SCC [16].

BCC with ductal and glandular differentiation is very uncommon. The preferred tumour localisation is the eyelids. The glandular structures demonstrate an apocrine differentiation [17]. In contrast to our case, no co-expression of keratin and vimentin was reported. The present tumour had some glandular-like bulbar formations but no ductal parts.

The tumour was ulcerated, and ulcerated CCS of the hand had been described previously [18].

Our patient presented initially because of leg swelling caused by deep vein thrombosis. Tumors can alter the clotting system by various events including circulating tumour cells. Both ovarian and uterine carcinosarcoma-induced deep venous thrombosis have been reported [19][20].

The treatment of choice is surgery. Despite R0-resection, in one study, 27% of cases developed metastatic disease [21]. Negative prognostic factors are histologic subtype, age, tumour size > 2 cm, and nodal status. Patients with basal or squamous cell carcinoma-type CCS have a mean age of 72 years with clear male dominance. The 5-year disease-free survival is 70%. In contrast, adnexal CCS occurs in younger patients (mean age 58 years) and those have only a 25% 5-year disease-free survival [22].

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