Basal cell carcinosarcoma of the eyelid with osteosarcomatous transformation

John G. Heathcote[1], Phillip Moss[1], Noreen M. Walsh[1], Curtis W. Archibald[2]

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Abstract:

A carcinosarcoma is a neoplasm with malignant epithelial and mesenchymal components. It is thought to arise by mesenchymal transformation of the epithelial elements. The cutaneous form of carcinosarcoma is rare and is associated with sun exposure; most cases arise in the head and neck. The epithelial component may be a basal cell carcinoma, a squamous cell carcinoma, or an adnexal carcinoma. The mesenchymal component may be an osteosarcoma, a pleomorphic undifferentiated sarcoma, or another type of sarcoma. Only a few cases of cutaneous carcinosarcoma have been described in the periocular skin. We present a case of basal cell carcinosarcoma with osteosarcoma and pleomorphic undifferentiated sarcoma arising in the lower eyelid of an elderly man.

Keywords:

Basal cell carcinoma, cutaneous carcinosarcoma, eyelid neoplasms

Introduction

The concept of "carcinosarcoma" as a tumor of malignant epithelial elements combined with malignant mesenchymal elements was introduced by Virchow in 1864. These tumors are less common in the skin than in the viscera and are thought to arise when the epithelial component undergoes metaplastic transformation to malignant mesenchyme. They generally occur as exophytic, ulcerated nodules in the sun-exposed skin of older men, and there is often a history of recent rapid growth.

The epithelial component is usually a basal cell or squamous cell carcinoma, although rarely a malignant adnexal neoplasm may be present. A variety of sarcomas may form the mesenchymal component. Although basal cell carcinoma is the most common malignant tumor in the eyelid, there are few reports of the carcinosarcoma in the periocular skin.[1] In a review of 47 cases of cutaneous basal cell carcinosarcoma reported in the literature, only three were identified in this area.[2] In this report, we present of a case of a basal cell carcinosarcoma with osteosarcomatous differentiation arising in the lower eyelid of an elderly man.

Case Report

A 93-year-old male was referred to an oculoplastic surgeon with a 12–18 month history of a visible, fleshy mass on the left lower lid [Figure 1]. It had grown significantly and occasionally bled spontaneously. Six years previously, at a different hospital, the patient had had two incisional biopsies of a nodular basal cell carcinoma in the left lower lid. An excisional biopsy 4 months later confirmed the diagnosis, but despite positive margins, there was no further treatment.

On examination, the elevated lid mass measured 15 mm × 15 mm, was mobile, and was telangiectatic. It involved the lower canaliculus but was not adherent to the medial canthal tendon. Apart from a posterior chamber lens implant in the left eye, ocular examination was normal bilaterally. The lesion was thought to be a basal cell carcinoma, and the patient was scheduled for excision under frozen section control. The patient was taking warfarin because of a cardiac arrythmia, and this was stopped 1 week before surgery.

[1]Departments of Pathology and [2]Ophthalmology and Visual Sciences, Dalhousie University, Halifax, Nova Scotia, Canada

Address for correspondence: Dr. John G. Heathcote, 8 Sweet Grass Street, Bayfield, Ontario, N0M 1G0, Canada. E-mail: godfrey.heathcote@dal.ca

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The pathological specimen was a full-thickness excision of the eyelid measuring 22 mm horizontally (H) ×11 mm vertically (V). On the skin surface, there was a firm nodule measuring 17 mm (H) ×10 mm (V) ×7 mm in thickness. On the medial edge of the nodule, there was a black Eschar 7 mm across and some lashes were noted medially. The nodule was well circumscribed inferiorly but extended close to the inferior resection margin. Centrally, the tumor appeared to contain foci of calcification.

Microscopic examination revealed a basal cell carcinoma arising at the mucocutaneous junction that had caused expansion, distortion, and ulceration of the eyelid margin [Figure 2]. Part of the tumor was a conventional basal cell carcinoma with infiltrative sheets and nests, as well as adenoid and trabecular areas [Figure 3]. Adjacent to this, the basaloid cells showed more nuclear atypia and mitotic activity and had formed loosely cohesive sheets with small patches of necrosis. Within these sheets, there were islands of malignant osteoid, as well as osteoclastic giant cells, reflecting osteosarcomatous differentiation [Figures 4 and 5]. Some of the islands were partially calcified, and osteoid was also seen in the exudate on the ulcerated conjunctival surface. The immunophenotypes of the areas of conventional basal cell carcinoma and of the sarcomatoid areas were different [Table 1], with expression of EpCAM by the former and expression of vimentin by the latter [Figure 6a and b].

The full-thickness lid defect was reconstructed with a Hughes tarsoconjunctival flap and a cheek myocutaneous advancement flap. The final pathological diagnosis was basal cell carcinosarcoma with transition from undifferentiated sarcomatous to osteosarcomatous characteristics in the mesenchymal component of the tumor. Given the potentially aggressive nature of the tumor and its proximity to the inferior resection margin, further excision was considered, but the patient died shortly after the initial surgery.

Discussion

Cutaneous carcinosarcoma is an uncommon tumor with around 130 reported cases.[3] Approximately 55% arise in the head-and-neck region, but few have been reported in the periocular skin.[4] Mc Menamin *et al*. described a 3-mm basal cell carcinosarcoma "under right eye" in a 65-year-old female and a 6 mm basal cell carcinosarcoma "above left eye" in a 71-year-old female, but the precise locations of the tumors were not specified.[5] Both of these tumors contained areas of malignant osteoid. An inner canthal basal cell carcinosarcoma in a 71-year-old man was described as ulcerated, but the size was not given and the tumor did not contain malignant osteoid.[6] Patel *et al*. briefly reported an ulcerated squamous cell carcinosarcoma of the upper eyelid showing osteosarcomatous transformation in a 71-year-old male but again the size was not stated.[7] West and Srivastava described a squamous cell carcinosarcoma involving the medial canthus and lower eyelid of an 82-year-old male.[8]

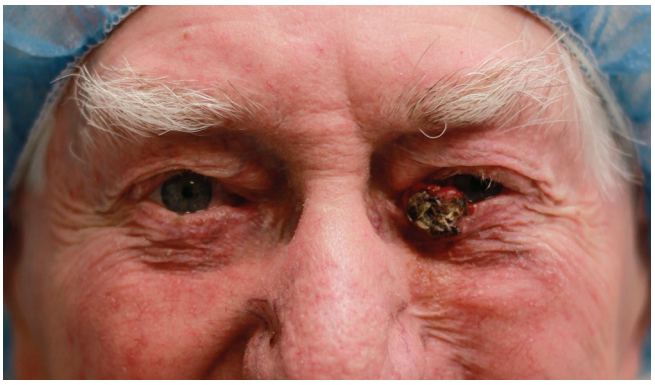

Figure 1: Ulcerated nodule on the medial left lower lid of a 93-year-old male

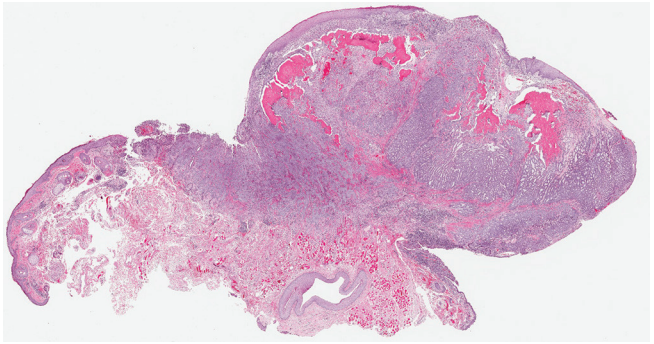

Figure 2: Full-thickness section of eyelid with basaloid tumor excrescence at lid margin. Both the skin (left) and conjunctiva (right) are ulcerated. The tumor mass contains irregular islands of eosinophilic and malignant osteoid. The inferior canaliculus is seen at the bottom of the picture (H and E; ×200)

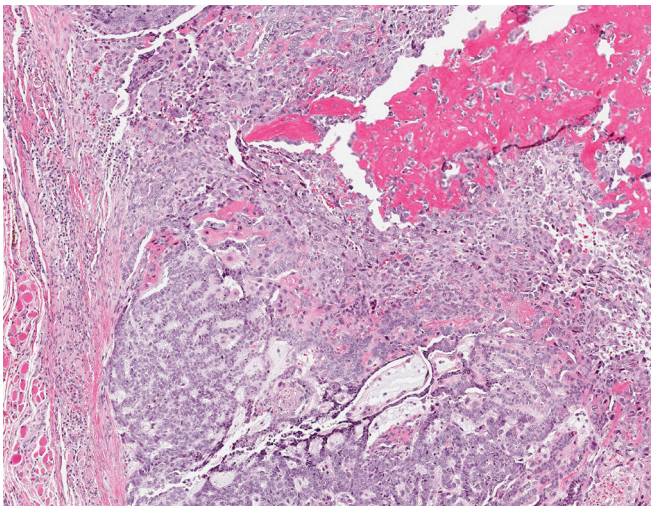

Figure 3: Conventional basal cell carcinoma with adenoid growth pattern is separated from malignant osteoid by pleomorphic basaloid cells (H and E; ×100)

This exophytic tumor appeared to develop over a period of 6 months and measured 31 mm × 21 mm.

The occurrence of epithelial–mesenchymal transformation in embryonic development is well established and it is

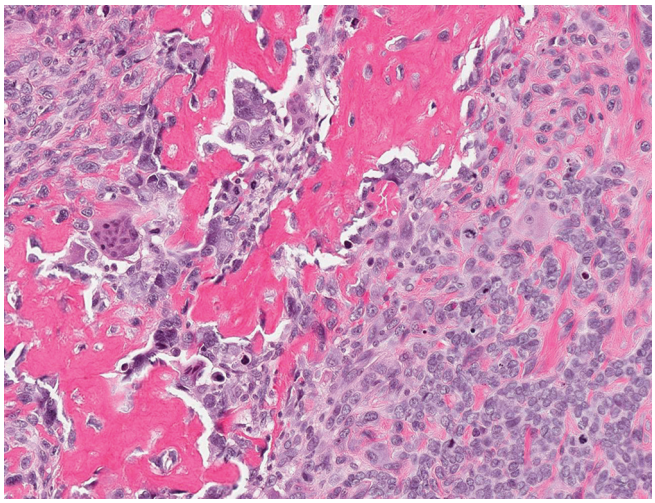

Figure 4: A sheet of pleomorphic basaloid cells contains islands of malignant osteoid and osteoclastic giant cells (H and E; ×200)

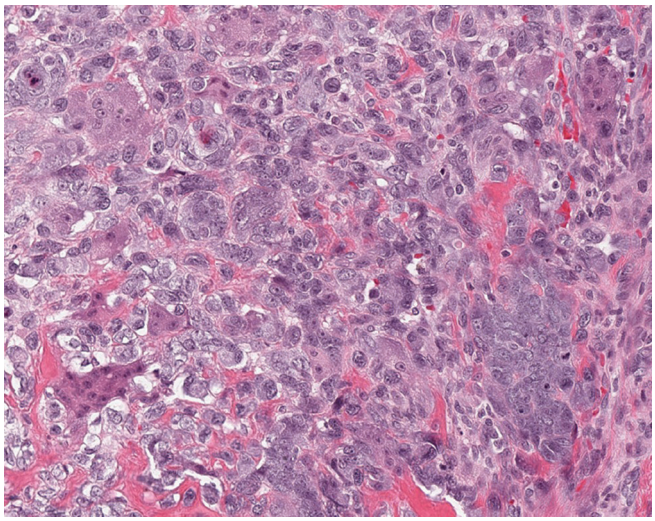

Figure 5: Pleomorphic sarcoma with osteoclastic giant cells (H and E; ×400)

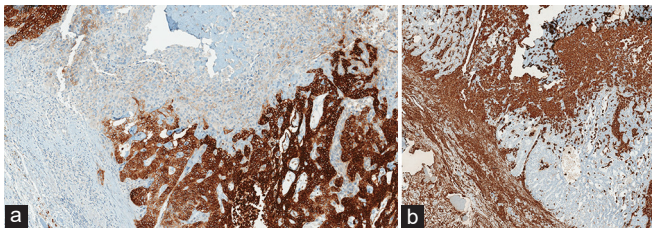

Figure 6: Expression of EpCAM in conventional basal cell carcinoma (a) contrasts with expression of vimentin in sarcomatoid area and desmoplastic stroma (b) (Immunoperoxidase; a, ×200; b, ×100)

not surprising that such divergent differentiation would happen occasionally in tumorigenesis. In some of the reported cases of cutaneous carcinosarcoma, there was a rapid growth of a preexisting lesion, suggesting that mesenchymal transformation was a late occurrence in the development of the tumor. Our patient had a previous history of nodular basal cell carcinoma of the left lower lid and it is likely that the carcinosarcoma resulted from metaplasia of residual/recurrent basal cell carcinoma over 12–18 months. Late occurrence of mesenchymal transformation may also help to explain the apparent rarity of basal cell carcinosarcoma in the periocular skin, despite the frequency of basal cell carcinoma in this area. When a basal cell carcinoma arises in this location, early recognition generally leads to prompt treatment before the lesion grows to a large size and perhaps before transformation can occur.

Table 1: Immunohistochemical profile of epithelial and mesenchymal components of basal cell carcinosarcoma of the eyelid

IHC stain	Conventional BCC	Sarcomatoid BCC
CK5/6	Strong	Weak/absent
EpCam	Strong	Weak/absent
Vimentin	Absent	Strong
p63	Present	Absent
BCL-2	Variable	Variable

IHC: Immunohistochemical, BCC: Basal cell carcinosarcoma

The case presented here illustrates the typical clinical picture of a cutaneous carcinosarcoma. The tumor cannot be distinguished from a conventional basal cell or squamous cell carcinoma and given the patient's history, a basal cell carcinoma was suspected. With this clinical diagnosis, the surgeon proceeded to full-thickness excision of the eyelid with frozen section examination of the resection margins. The prognosis of cutaneous carcinosarcoma is determined by the nature of the epithelial component rather than the mesenchymal component and is generally better with basal cell carcinosarcoma (4% recurrence or metastasis) than with squamous cell and adnexal carcinosarcoma (38%, recurrence or metastasis).[9] Despite its generally indolent course, it has been recommended that basal cell carcinosarcoma should be treated as a high-risk basal cell carcinoma with clear surgical margins around the tumor.[10] In our patient, the tumor was very close to the inferior resection margin, and re-excision was deemed prudent. The patient, however, died before this could be considered.

The mesenchymal component of cutaneous carcinosarcoma may be an osteosarcoma, chondrosarcoma, undifferentiated sarcoma and occasionally, leiomyosarcoma or rhabdomyosarcoma.[3] The epithelial and mesenchymal components may be intermingled or there may be a transition zone between the two.[5,11] Mc Menamin *et al*. described 11 cases of basal cell carcinosarcoma, ten of which contained osteosarcoma and undifferentiated sarcoma in varying proportions.[5] The 11th case did not contain malignant osteoid but had a component of malignant mesenchyme rich in osteoclastic giant cells, also seen in our case, that they interpreted as an early stage of osteosarcomatous transformation.

The pathological diagnosis of a cutaneous carcinosarcoma is more dependent on careful examination of hematoxylin and eosin-stained sections than immunohistochemistry and our specimen showed the characteristic histopathology of a

basal cell carcinosarcoma with transition to osteosarcoma.[7] Immunoperoxidase studies did reveal the expected pattern with the epithelial component expressing CK5/6, EpCAM (Ber EP4), and p63 but not vimentin. The latter marker was expressed by the sarcomatous cells [Table 1]. Although not examined in our case, both components have been shown to express p16 and p53, which are linked to sun exposure.[2]

In conclusion, our case draws attention to a rare development in the setting of nonmelanoma skin cancer and contributes to a scant literature on this topic. It serves to alert clinicians to the potential significance of a sudden change in a previously indolent tumor of the skin and advocates for prompt investigation and treatment of such lesions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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