Spiradenocarcinoma of the eyelid: a case report

Introduction

Spiradenocarcinoma (SPC) is an uncommon cutaneous neoplasm[1-10] that usually derives from the malignant transformation of a spiroadenoma (SPA) [1,4-10]. The cell of origin is eccrine in most pathological specimens[1-10] but appocrine markers have also been identified in some cases[2,4]. Although these tumours can be found virtually anywhere on the skin, only 7 cases of SPA and none of SPC of the eyelid have been reported so far[2,3], being this case the first reported such tumour in this location.

Case report

A 84 year-old woman was referred to our clinic for evaluation of a painless mass in her upper right eyelid of several months duration. She had advanced primary open angle glaucoma treated with travoprost and dorzolamide. Her medical history was unremarkable except for a severe osteoporosis that had a considerable effect on her quality of life.

On examination, a firm non-infiltrating bilobuled mass that involved both puncta and induced mechanical ptosis was observed (Figure 1). The right eye had a visual acuity reduced to light perception and glaucomatous optic disc atrophy whereas the left eye retained a visual acuity of 20/40 and a moderately excavated papilla. The rest of the ophthalmological examination was not contributory.

A biopsy of the lesion was performed under local anaesthesia. Gross examination showed a mass of yellowish tissue with necrotic and hemorrhagic areas and elastic consistency. Microscopic examination revealed groups of basaloid cells arranged in nodules and pseudoglandular structures suggestive of eccrine spiradenoma (figure 2),

with areas of focal necrosis and cell atypia (Figure 3) and muscle invasion (figure 4). Inmunohistochemistry showed positiveness to CAM 5.2.

Computed tomography (figure 5) showed a well circumscribed 24 x 20 x 25 mm mass in the medial aspect of the right orbit without apparent muscle or globe involvement that extended up to the lamina papyracea without bone destruction. Enhancement after contrast administration (Xenetix 350, 90 cc) demonstrated a low density heterogeneous mass consistent with multifocal areas of necrosis and hemorrhage.

With an histological diagnosis of SPC, the patient was informed about the nature of her disease and the potential risks of radiotherapy, surgery and of not having surgery. However she refused any kind of treatment. The patient remains free of metastatic disease one year after the diagnosis.

Comment

SPC is an uncommon tumour that usually arises from a previous long standing SPA[1,4-10]. They both affect patients older than 50 years with no sex predilection[1,4-10]. Around a hundred cases of SPA[6,8-10], most in the trunk and extremities,[14-6,8,10] and some of them associated with Brooke-Spiegler syndrome[7], have been reported since its first description in 1972, but only a few of SPC, this being the first reported case in the eyelid to our knowledge.

Although several signs such as change in colour or ulceration and symptoms such as pain can alert of a possible malignant transformation of a spiradenoma[1,4-10], in our case, we do not know if the actual lesion arised de novo because the patient did not refer any previous disorder in the eyelid.

Histopathological examination of SPC reveals areas of typical spiradenoma along with areas with atypia, necrosis and loss of lobular pattern [1,4-10]. It can also have the dual cell population and the vascular proliferation that characterize spiradenomas[4,6,7]. Inmunohistochemistry shows positiveness to CAM 5.2, epithelial membrane antigen (explains its epithelial nature)[5-8], carcinoembryonic antigen and S100 among others[1,4-9] but we did not use it because haematoxylin-eosin left no doubt about the histological diagnosis[7]. A mutation in p53 has also been described[4,6-9].

Metastases, usually found in bone, lung, liver and brain[1,4-6, 8-10] but also llymphatics infiltration, worsens the prognosis dramatically[1]. The probability of having metastases, which were not detected, reported in 21-57% of cases of SPC[1,6,9], increases if sarcomatous changes are found in the histopathological analysis[5,8,10]. No sarcomatous foci or syringoma were found in this case, either.

Wide surgical excision is the treatment of choice (with 100% survival rate if there are no metastases[9]) but the use of radiotherapy is controversial [1,4-6,8-10]. Chemotherapy is reserved for cases in which SPC has spread to other organs[4-6,8,10].

We can conclude that SPC must be included in the differential diagnosis of the tumours of the eyelid and also that observation instead of aggressive surgical intervention may be an option in a subgroup of patients with a low risk of metastases and/or short life expectancy.

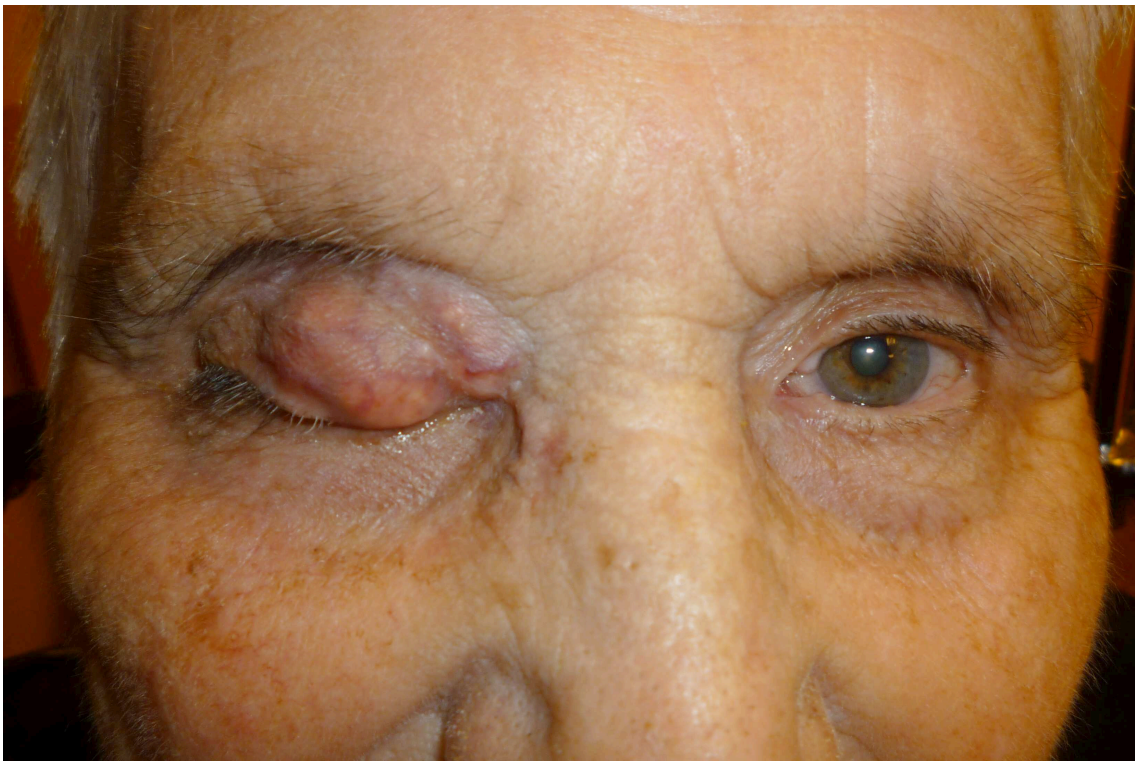

Figure 1. Eyelid mass inducing mechanical ptosis.

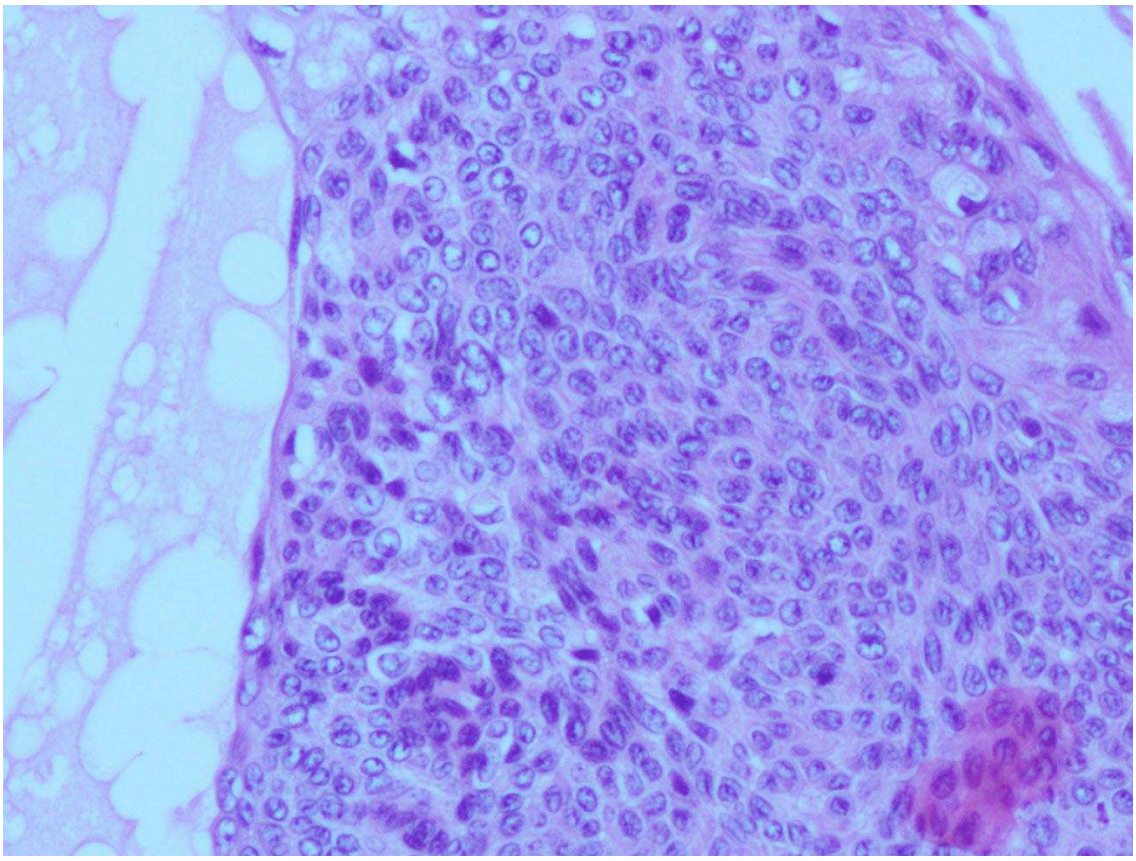

Figure 2. Areas of spiradenoma with its typical nodular pattern and without atypia. Haematoxylin-eosin, 20X.

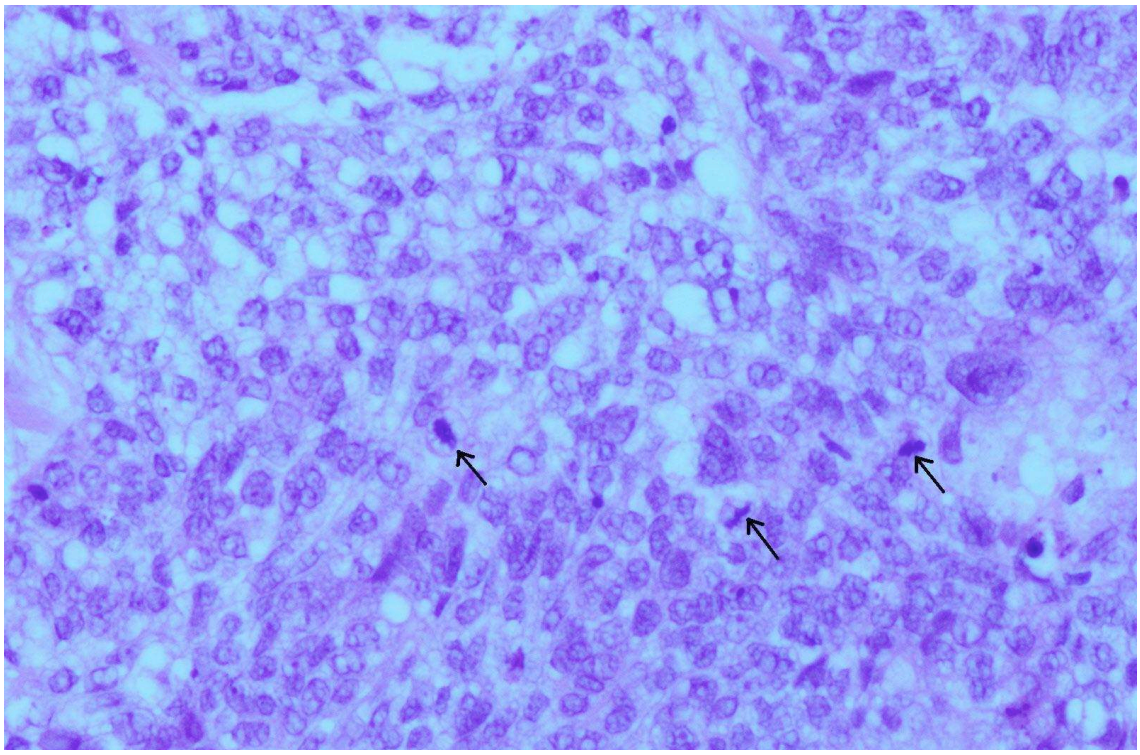

Figure 3. Areas with atypia showing malignant transformation and loss of lobular pattern. Mitosis are also evident (arrows). Haematoxylin-eosin, 20X.

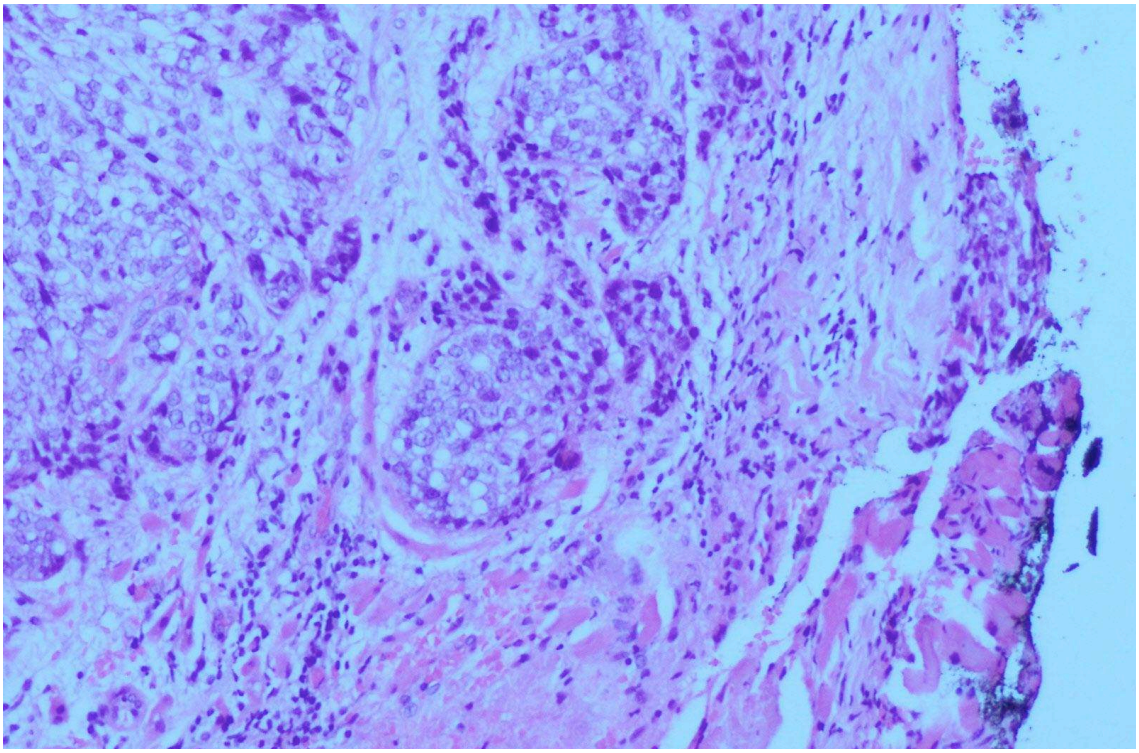

Figure 4. Invasion of medial rectus in its anterior insertion. Haematoxylin-eosin, 20X.

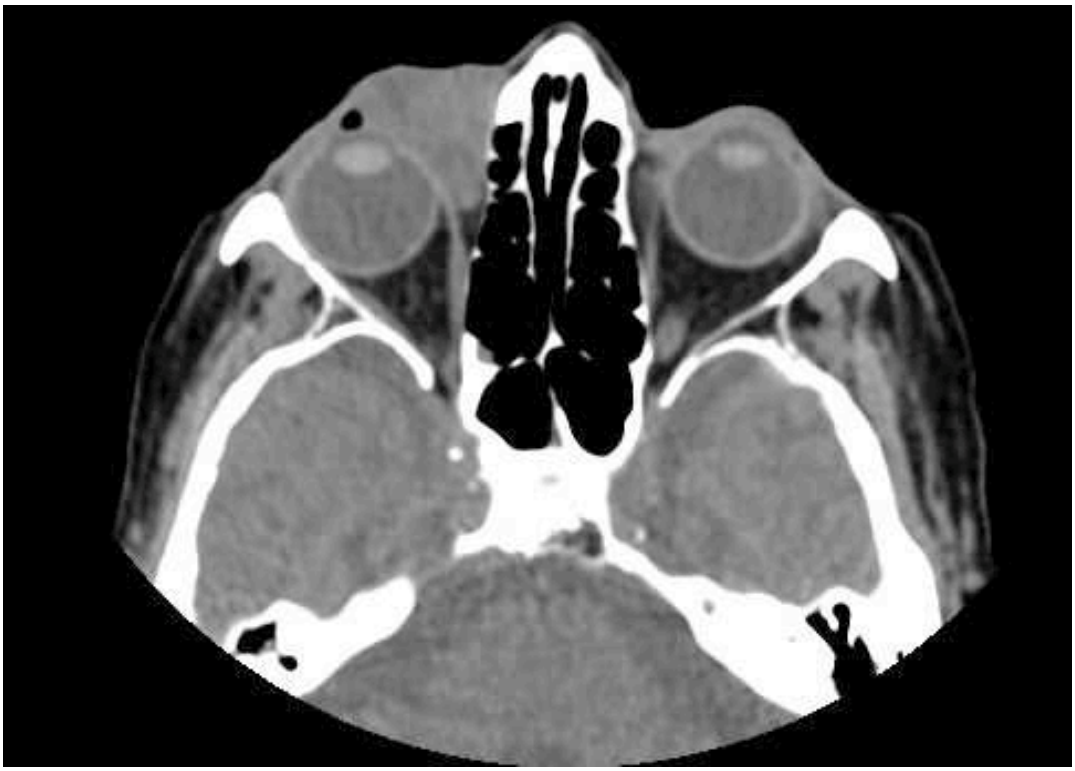

Figure 5. In the CT scan the tumor appears as a well delimited mass without aparent invasion of adjacent structures.

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