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Hidradenocarcinoma eccrinale syringomatodes of the eyelid – case of a rare malignancy

Hidradenocarcinoma eccrinale syringomatodes powieki – przypadek rzadkiego nowotworu złośliwego

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Summary: Purpose: Presentation of a case of very rare malignant tumor of eccrine sweat glands in the eyelid.
Material and methods: A 44 years old man with a tumor of the upper left eyelid is presented.
Conclusions: Sweat gland carcinomas are rare malignant tumors of the skin adnexa. Diagnosis of these carcinomas is difficult due to their infrequency.

Słowa kluczowe: guzy powiek, rak gruczołów potowych, gruczolakorak ekrynowy.
Key words: eyelid tumors, sweat gland carcinoma, eccrine adenocarcinoma.

Neoplastic lesions arising from the sweat glands are divided, according to their potential for distinctive local tissue infiltration, recurrence and metastasis, into benign and malignant tumors (cancers). The second main division based on the type of the glands is into "eccrine" and "apocrine" categories (1). Carcinomas of the eccrine sweat glands represent a very rare group of tumors, particularly in the eyelid location. In this article we report the case of eccrine hidradenocarcinoma of the upper eyelid that does not tend to local recurrence and metastasis to the regional lymph nodes.

Case report

A 44 years old man presented in December 2006 with an eight year history of a nodular lesion in the left upper eyelid, which had considerably increased during that time. Ophthalmic examination revealed the best corrected visual acuity of 1.0. The intraocular pressure in both eyes was 16 mmHg. He has been treated for general hypertension for 14 years.

A dome-shaped, firm mass, 9 mm x 3 mm in size, with a discrete net of intrinsic vessels was seen in the left upper eyelid (Fig. 1a). There was no loss of integrity of the skin or ulceration over the tumor. The margin of the eyelid was involved in the tumor mass (Fig. 1b). There was no obvious enlargement of regional lymph nodes. Ocular examination, including slit-lamp and fundus examination, was unremarkable. Surgical excision of the tumor was performed with retention of the appropriate margins of the macroscopically healthy tissues. Massive cryotherapy of the remaining, underlying tissue and plastic, reconstructive surgery were performed.

The tumor was histopathologically diagnosed as hidradenocarcinoma eccrinale syringomatodes. The excision was complete with narrow margin clearance.

In the follow-up the patient's visual acuity has not changed, the wound healed correctly with no evidence of local recurrence or lymph node or distant metastasis.

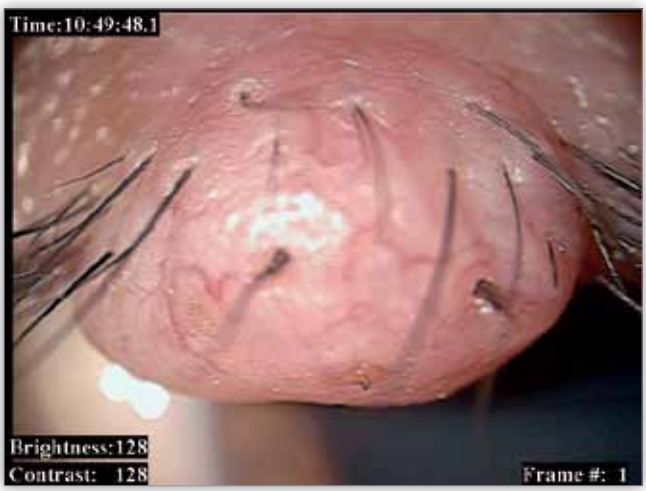

Fig. 1a. Dome-shaped tumor in the upper eyelid.
Ryc. 1a. Kopulasty guz powieki górnej.

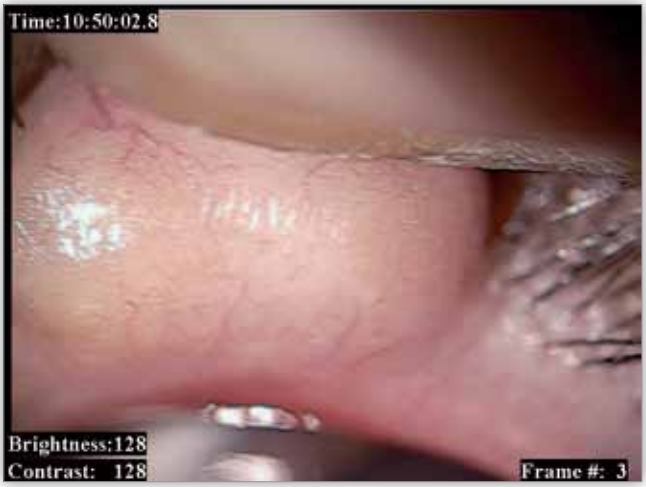

Fig. 1b. The tumor mass involving the margins of the eyelid.
Ryc. 1b. Guz obejmujący częściowo krawędź powieki.

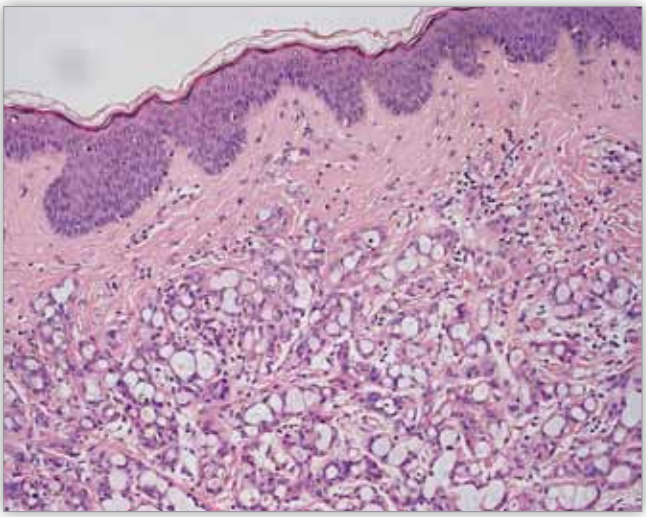

Fig. 2. Glandular structures with lumen formed by cells with intracytoplasmic vacuolization.
Ryc. 2. Poronne światła gruczołów utworzone przez komórki o zwakuolizowanej cytoplazmie.

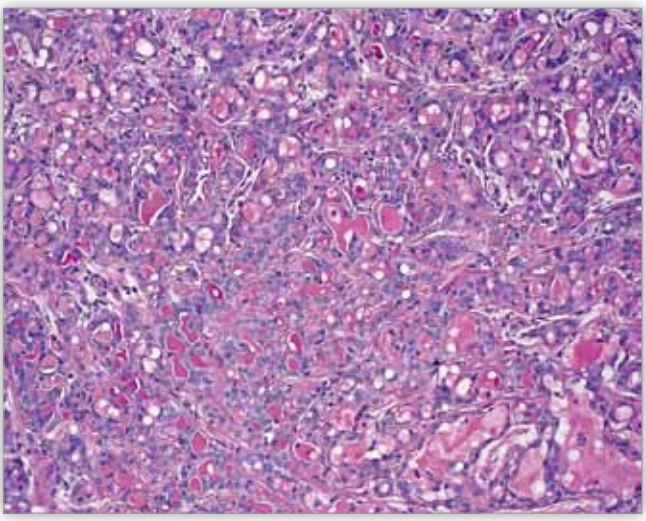

Fig. 3a. A PAS staining of the intracellular material.
Ryc. 3a. Wybarwiona metodą PAS treść wewnątrz komórek nowotworu.

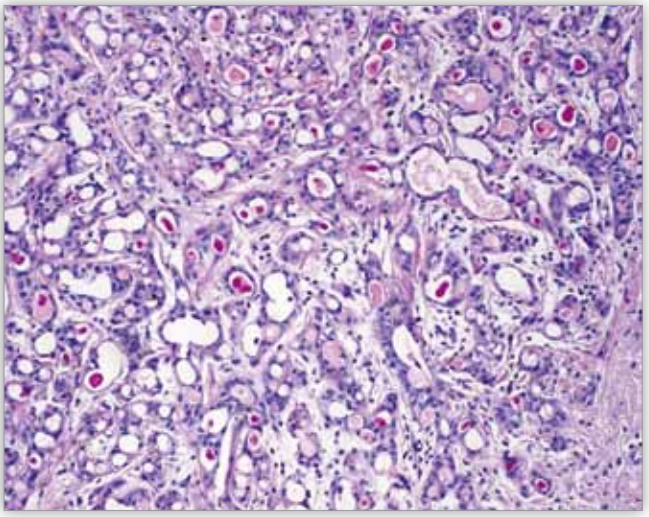

Fig. 3b. A PAS staining of the intracellular material with diastase predigestion.
Ryc. 3b. Barwiące się metodą PAS resztki materiału wewnątrzkomórkowego po wytrawieniu diastazą.

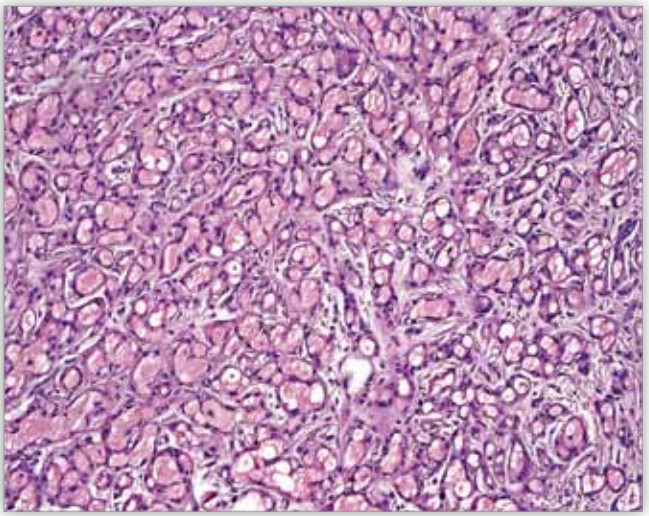

Fig. 4. Mucikarmin staining of intracellular material.
Ryc. 4. Barwienie mucykarminą treści wewnątrzkomórkowej.

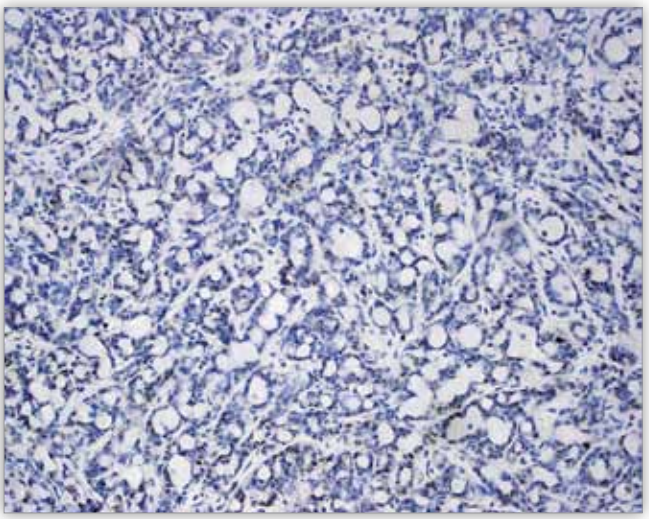

Fig. 5. Positive staining for S100 protein.
Ryc. 5. Dodatnie barwienie w kierunku obecności białka S100.

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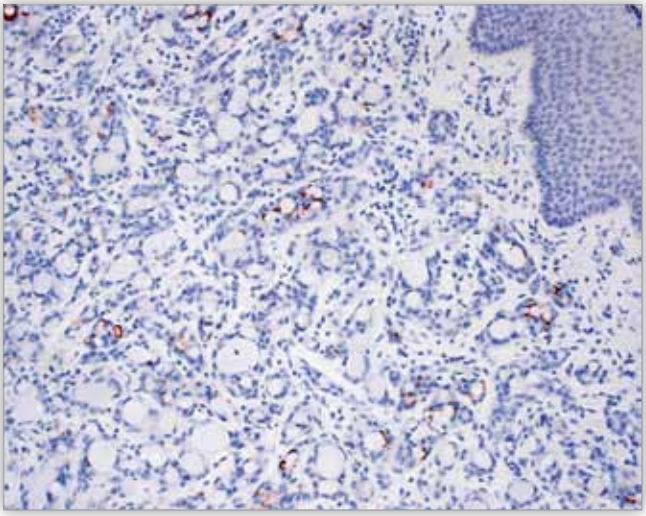

Fig. 6. Positive staining for epithelial membrane antigen (EMA).
Ryc. 6. Dodatnie barwienie w kierunku antygenu błony nabłonkowej (EMA).

Pathological findings

The tumor was composed of cells with intracytoplasmic vacuolization, which formed glandular structures with lumen. The neoplasm was arranged focally in a tubular-like pattern (Fig. 2). A PAS staining of the intracellular material with or without diastase predigestion was observed (Fig. 3a and Fig. 3b). Mitotic figures were absent. The mucicarmine preparations demonstrated the presence of mucin within the tumor cells (Fig. 4). There was also positive staining for cytokeratine, S100 protein (Fig. 5) and epithelial membrane antigen (EMA) (Fig. 6).

Discussion

In general, diagnosis of eccrine hidradenocarcinoma is challenging because of inconsistent nomenclature, rarity of these neoplasms and heterogeneous histological features with variable morphology of cells composing the tumor (2,3). Furthermore, they may be difficult to confirm histopathologically, because of similarities to either other primary malignancies or metastatic adenocarcinoma to the eyelid (4).

Among the eccrine malignancies the most common are: microcystic adnexal carcinoma, eccrine porocarcinoma and eccrine hidradenocarcinoma (3). They mostly occur on the face, scalp, lower extremities and trunk, but the eyelid location is typical for primary eccrine mucinous carcinoma (40%) (5,6).

Eccrine adenocarcinoma has been regarded as the classic form of eccrine carcinoma. Clinically it appears as a nodule and its histopathological features resemble a moderately to poor differentiated adenocarcinoma (7). There are three distinct variants of eccrine adenocarcinoma of the eyelid: mucinous, ductal and adenoid cystic. Well-differentiated ductal carcinoma has been classified as malignant syringoma, poorly differentiated one is classified as the histiocytoid variant (8).

Although the basic division of the sweat glands carcinoma into eccrine and apocrine is used universally, the absence of strict and homogeneous histological criteria limits the usefulness of this classification, all the more because its clinical importance is not proven yet (2). Ko et al., after careful histological and immunohistochemical evaluation of six hidradenocarcinomas, noted that all examined neoplasms displayed histological features considered eccrine (cuticular cells) as well as apocrine (decapitation secretion) and they proposed the term- "apoeccrine". This kind of differentiation would also explain the ability of these tumors to form all over the body, whereas the two former types of the sweat glands are normally found only in certain sites of the skin (2).

The pathological examination is usually insufficient to determine the eccrine or apocrine origin of the tumor, however, histological and in particular immunohistochemical parameters remain vital to assess the clinical course of the disease. Suggested pathological criteria for malignancy are: mitoses (also in clear cells, if present), atypical cells, perineural and vascular invasion, dispersed growth pattern and extension depth (2). Immunohistochemical parameters are thought to be even more important especially in the light of histological heterogeneity of these neoplasms and the most representative is positive staining for cytokeratin, CEA, EMA, S 100 protein, p53 and Ki67 (2).

All these criteria have great significance when preoperative estimation of malignancy is difficult, because tumors do not have any pathognomonic clinical appearance and resemble their benign counterparts.

They have to be differentiated from basal cell carcinoma, papilloma, keratoacanthoma, Kaposi's sarcoma, hemangioma, pyogenic granuloma (5) and Meibomian carcinoma (7). Additional immunohistochemical differential diagnosis of eccrine adenocarcinoma should consider metastatic adenocarcinoma, microcystic adnexal carcinoma and adenoid cystic carcinoma, all of which are positive for S100 protein (7).

In a study of 35 patients, Mehregan et al. concluded that all eccrine carcinomas have a tendency for local recurrence. Local lymphatic spread or distant metastasis occurs in less than 10% of cases (9). Nevertheless, eccrine malignancies differ from each other in relation to their biological behavior.

Microcystic carcinoma, mucinous carcinoma and adenoid cystic carcinoma have so called low malignancy potential, whereas eccrine porocarcinoma, malignant acrospiroma and malignant spiradenoma represent more malignant counterparts of these neoplasms (10).

Increased potential to expansive growth pattern and difficulties in treating recurrences are related mainly to histopathological differentiation (grading). El-Domeiri et al., divided 83 cases of sweat gland carcinomas into five groups according to their grading. Five years survival in the fourth group (very poor differentiated tumors), was 17% and in the fifth group (anaplastic tumors), was 0 (11). Lymph nodes metastasis occurred in 71% of cases with very poorly differentiated carcinoma, in 86% of cases with anaplastic tumors and only in 9% of cases with very well differentiated neoplasm (11).

Eccrine adenocarcinoma represents a well differentiated counterpart of a number of specific variants of eccrine carcinomas (7). Small number or absence of mitotic figures suggests its low malignancy potential and good prognosis. Despite the fact that eccrine adenocarcinoma is thought to recur one or more time in 50% of cases (12), this rate is mainly due to poorly differentiated variants.

Another significant factor, influencing recurrence, metastasis and survival rate is surgical method of excision. Complete

removal of these tumors is crucial, therefore the most accurate surgical method is Mohs micrographic surgery (3).

Extreme rarity of eccrine carcinomas, resulting in very small number of studies, mostly case reports, reveals a necessity of a systemic work-up including diagnostics, clear classification and histological appearance to ensure appropriate care.

Histological features of the tumor presented in our case correspond to the ductal variant of eccrine adenocarcinoma (malignant syringoma) (8) and it complies with the immunohistochemical criteria of malignancy described by Ko et al. (2). However, low malignancy potential of this neoplasm may be considered due to its well differentiation as a ductal counterpart of these adenocarcinomas and to the absence of pathological parameters of malignancy (8). These features in association with a complete surgical excision of the tumor may explain the benign clinical course of the disease with no evidence of local recurrence or lymph node or distant metastasis.

Conclusions

Sweat gland carcinomas are rare malignant tumors of the skin adnexa. Diagnosis of these carcinomas is difficult due to their infrequency.

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