

Primary cutaneous mucinous carcinoma of the cheek. Case report

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SUMMARY: Primary cutaneous mucinous carcinoma of the cheek. Case report.

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Primary mucinous carcinoma of the skin (MCS) is a rare neoplasia described for the first time by Lennox et al. in 1952 and formally reviewed by Mendoza and Helwing in 1971. It is an uncommon subtype of sweat gland tumor. MCS affects men (58,8%) more than women (41,2%). It tends to occur in more elderly individuals (average 62,6 years, range 8-87 years), although the disease has been reported in a patient as young as 8 years old. In the english Literature are described 100 case of MCS. MCS has a varied clinical onset, typically presenting as an asymptomatic, slow-growing, painless, papular or nodular, subcutaneous or cutaneous, ranging from 5 mm to 120 mm. The lesion is frequently single and isolated red, pink, gray, blue or purple colored. Telangiectasia may or not be present. The surface may be smooth, ulcerated or crusted. Local recurrence occurs frequently (29,4%) after conventional excision, but the rate of distant metastasis is low (9,6%). In fact this tumour is typically avascular, a factor that help to explain its low rate of metastasis.

In September 2007, a 69 year old white man was referred to our observation for the evaluation of a painless, superficial nodular mass in left cheek that had slowly grown over 1 year to 2x1,4 cm current measures. The lesion was excised. Microscopic examination revealed a mucinous cutaneous carcinoma of the skin. Investigations for a primary visceral malignancy, including CT total body and colonoscopy, were negative.

RIASSUNTO: Carcinoma mucinoso cutaneo primitivo della guancia. Caso clinico.

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Il carcinoma mucinoso cutaneo (CMC) primitivo è una rara neoplasia cutanea descritta per la prima volta da Lennox et al. nel 1952 e formalmente classificata da Mendoza ed Helwing nel 1971. È un sottotipo non comune di tumore delle ghiandole sudoripare. Il CMC interessa più gli uomini (58,8%) che le donne (41,2%) e prevalentemente gli individui anziani (età media 62,6 anni, range di età 8-87). Nella letteratura in lingua inglese sono descritti 100 casi di CMC. Il CMC ha un esordio clinico variabile; tipicamente si presenta come un nodulo asintomatico, a lenta crescita, non dolente, papulare o nodulare, sottocutaneo o cutaneo, con dimensioni comprese tra i 5 e i 120 mm. Frequentemente si tratta di lesione singola, di colore rosso, rosa, grigio, blu, o porpora. Possono essere presenti teleangectasie. La superficie può essere liscia, ulcerata, o crostosa. La recidiva locale post-escissione si verifica frequentemente, ma la percentuale di metastasi a distanza è bassa (9,6%). Infatti, questo tumore è tipicamente avascolare, e ciò spiega la bassa incidenza di metastasi.

Nel settembre 2009, un uomo bianco di 69 anni giunse alla nostra osservazione per la valutazione di un nodulo superficiale, non dolente, alla guancia sinistra, che era lentamente cresciuto nel corso di un anno fino a misurare 2x1,4 cm. La lesione è stata asportata. L'esame microscopico rivelava trattarsi di un carcinoma mucinoso cutaneo. Le indagini volte ad individuare eventuali tumori primitivi viscerali, incluse TC total body e colonscopia, sono risultate negative.

KEY WORDS: Mucinous carcinoma - Skin.
Carcinoma mucinoso - Cute.

Introduction

Primary mucinous carcinoma of the skin (MCS) is a rare neoplasia described for the first time by Lennox et al. in 1952 (1) and formally reviewed by Mendoza and Helwing in 1971 (2). It is an uncommon subtype of sweat gland tumour. It is occasionally misdiagnosed as metastatic mucinous carcinoma (3,4). MCS is a low grade malignant, lymph node metastasis are uncommon though local recurrence is relatively frequent (1-4).

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We report a case of primary cutaneous mucinous carcinoma of the cheek.

Case report

In September 2007, a 69 year old white man was introduced to our observation for the evaluation of a painless, superficial nodular mass in left cheek that had slowly grown over 1 year to 2 x 1,4 cm current measures. The mass was hard and ulcerated. The regional lymph nodes were not palpable. The lesion was excised with 1 cm free margins under local anesthesia.

Microscopic examination revealed a mucinous cutaneous carcinoma of the skin. The neoplasia infiltrated the deep dermis, but not the subcutaneous. Surgical margins were negative for infiltration. Investigations for primary visceral malignancy, including total body CT and colonoscopy, were negative.

Discussion

Primary MCS is a rare low grade malignant subtype of sweat glands tumour. It is still controversial whether this neoplasia has eccrine or apocrine differentiation, but many authors favour the eccrine differentiation based on immunoistochemical studies and electron microscopic ultrastructural analysis. MCS affects men (58,8%) more than women (41,2%). It tends to occur in elderly individuals (average 62,6 years, range 8-87 years), although the disease has been reported in a patient as young as 8 years old. In the english Literature are described 100 case of MCS: the race was indicated in 64 cases: 39 White, 15 Black and 10 Asian (5). The incidence in men is twice that in women (6). This tumour commonly arises in the head or neck, and the eyelids the most common site (41%), followed by scalp (17%), face (14%), axilla (9%), vulva(4%), chest/abdominal wall (7%), neck (2%), extremity (2%), canthus (2%), groin (1%) and ear (1%) (5).

MCS has a varied clinical onset, typically presenting as an asymptomatic, slow-growing, painless, papular or nodular, subcutaneous or cutaneous, ranging from 5 mm to 120 mm. The tumour is a single isolated lesion red, pink, gray, blue or purple colored. Telangiectasia may or not be present. The surface may be smooth, ulcerated or crusted.

The differential diagnosis includes sebaceous cyst, hemangioma, adenocystic basal cell carcinoma, Kaposi's sarcoma, squamous cell carcinoma and metastatic adenocarcinoma; of these, the most important to consider is metastatic mucinous carcinoma of the skin. metastatic from another site. Infact, mucinous carcinoma may be very difficult to distinguish histologically from cutaneous metastases of breast, gastrointestinal tract, ovary, prostate and lung carcinomas (7). However the primary MCS produce a non sulfated mucin while metastatic mucinous

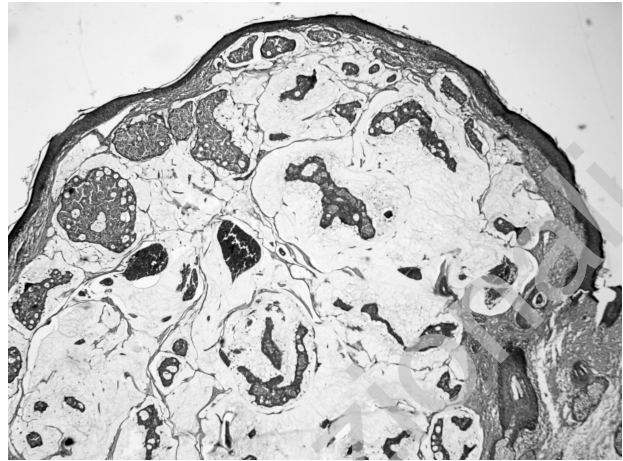


Fig. 1 - At low magnification (EE - 125x), the tumor has nodular appearance, raising the epidermis, with extracellular mucin.

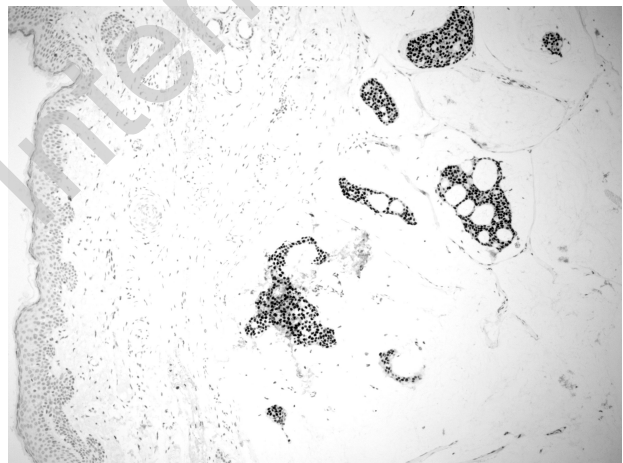


Fig. 2 - Immunohistochemistry (250x) shows the positivity for progesterone receptors.

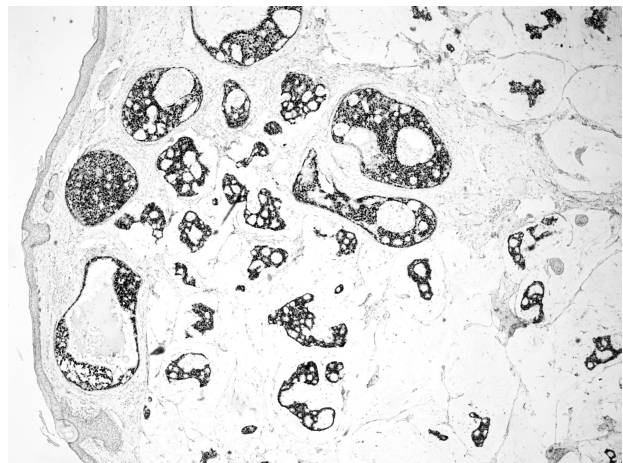


Fig. 3 - Immunohistochemistry (250x) shows diffuse nuclear positivity for estrogen receptors.

carcinomas, in particular these from gastrointestinal produce nonsulfated, neutral and sulfated mucins. A systemic evaluation of the patients, with breast examination, colonoscopy and total body CT scan is therefore mandatory for a definitive diagnosis of primary MCS.

Local recurrence occurs frequently (29,4%) after excision of MCS, but the rate of distant metastasis is low (9,6%) because the tumour is typically avascular (5).

Histological pattern of MCS is characteristic with cords and nests of basaloid cells embedded in pools of mucin and separated by thin fibrous septae. Mitoses are uncommon (1-3). Histochemically the mucin produced by the tumor is PAS- and colloidal iron-positive, and resistant to hyaluronidase. Alcian blu is positive at pH 2.5 but negative at pH 0.4 or in presence of sialidase. This histochemical pattern is consistent with the presence of a non sulfated mucoprotein, most likely sialomucin. Enzyme histochemistry is positive for succinic dehydrogenase, lactic dehydrogenase and isocitric dehydrogenase (supporting an eccrine origine) (8). Immunohistochemically tumour cells demonstrate consistently positive staining with low-molecular-weight cytokeratin and epithelial membrane antigen (EMA). CEA and S-100 protein expression is variable (3,9). Recently, primary MCS has been found to express estrogen and progesterone receptors in a manner indistinguishable from mucinous adenocar-

cinoma arising in the breast (10) with P53 and c-erb-2 negative (3).

The treatment for the primary MCS is wide local excision; it seems reasonable to recommend excision with at least 1cm margins. The metastasis to regional lymph nodes are rare; lymph node resection is indicated if nodes are clinically involved. MCS is unresponsive to radiation therapy or chemotherapeutic agents (11). Because of frequent local recurrences, close follow up is warranted.

In our case the patient showed painless, superficial cheek nodular mass strongly positive to cytokeratine 7, and estrogen and progesterone receptors, but negative for TTF1 and CK 20.

Conclusion

The primary mucinous carcinoma of the skin is an uncommon variant of sweat gland tumor with an indolent course and infrequent metastasis relative to true glands tumours. Clinical differentiation from metastatic mucinous carcinoma is not simple and a meticulous investigation for a primary tumour is necessary. Once diagnosed, the tumour requires wide local excision to prevent recurrence.

References

1. Lennox B, Pearse AG, Richards HG. Mucin-secreting tumours of the skin: with special reference to the so called mixed salivary tumor of the skin and its relation to hidradenoma. *J Pathol* 1952;64:865.
2. Mendoza S, Helwing EB. Mucinous (adenocystic) carcinoma of the skin. *Arch Dermatol* 1971;103:68-78.
3. Bellezza G, Sidoni A, & Bucciarelli E. Primary mucinous carcinoma of the skin. *Am. J. Dermatopathol.* 2000; 20,166-170.
4. Breier F, Clabian M, Pokieser W, et al. Primary mucinous carcinoma of the scalp. *Dermatology* 2000;200:250-253.
5. Steve R. Martinez, Shawn E. Young. Primary mucinous carcinoma of the skin: a review. *The internet Journal of Oncology*.2005 volume 2 number 2.
6. Snow SN, Reizner GT. Mucinous eccrine carcinoma of the eyelid. *Cancer* 1992;70:2099-104.
7. Karimipour DJ, Johnson TM, Kang S, Wang TS, Lowe L. Mucinous carcinoma of the skin. *J Am Acad Dermatol* 1997;36:323-6.
8. Headington JT. Primary mucinous carcinoma of the skin: histochemistry and electron microscopy. *Cancer* 1977;39:1055-63.
9. Carson HJ, Gattuso P, Raslan WF, Reddy W. Mucinous carcinoma of the eyelid: an immunohistochemical study. *Am J Dermatopathol* 1995;17:494-8.
10. Hanby AM, Mckee P, Jeffery M, et al. Primary mucinous carcinoma of the skin express TTF1, TTF3, estrogen receptor, and progesterone receptors. *Am J Surg Pathol* 1998;22:1125-31.
11. Yeung KY, Stinson JC. Mucinous (adenocystic) carcinoma of sweat glands with widespread metastasis: case report with ultrastructural study. *Cancer* 1977;39:2556-62.