CASE REPORT

Unusual skin mass (primary cutaneous mucinous carcinoma)

Hussnain Javaid,^{1,2} Naeem Raza,³ Urwa Ejaz,⁴ Tariq Sarfraz⁵

¹Military Hospital Rawalpindi, Rawalpindi, Pakistan
²Queen Alexandra Hospital, Portsmouth, Hampshire, United Kingdom

³Department of Dermatology, Military Hospital Rawalpindi, Rawalpindi, Pakistan

⁴Army Medical College, Rawalpindi, Pakistan

⁵Department of Pathology, Army Medical College, Rawalpindi, Pakistan

Correspondence to
 Dr Hussnain Javaid,
 hussnainamc@gmail.com

Accepted 3 January 2018

SUMMARY

Primary mucinous carcinoma of the skin is a rare malignant tumour of sweat gland origin. Diagnostic concerns include its deceptively benign appearance in some cases and the difficulty in differentiating it from secondary mucinous carcinoma of skin metastasising from a primary source elsewhere. A case of a 75-year-old man is reported who presented with a slowly growing painless mass near the lateral canthus of the right eye for about 2 years. Clinically, a diagnosis of basal cell carcinoma was made whereas histopathology revealed mucinous carcinoma of the skin. The primary source of the tumour could not be found on detailed physical examination and laboratory investigations. Immunohistochemistry, performed later, was consistent with primary cutaneous mucinous carcinoma.

BACKGROUND

Primary mucinous cutaneous carcinoma is a rare but important cancer of the sweat glands with a low metastatic potential but high recurrence rate.

- It is easily mistaken to be a benign lesion, where no further investigation is done erroneously.
- The prognosis is excellent, when diagnosed accurately and treated timely, which is much better from the more common secondary metastatic deposits from a primary source (eg, in the gastrointestinal tract).
- No evidence-based guidelines are available for the treatment of the tumour.
- There is still some debate as to the exact origin of the tumour (eccrine or apocrine), but most authorities favour the origin to be eccrine sweat glands.

CASE PRESENTATION

A 75-year-old male patient presented with a swelling over his right temple, near the lateral canthus of the right eye, for a period of around 2 years. The swelling was increasing in size gradually ever since. Apart from the slight occasional discomfort, there was no history of pain, bleeding or discharge from the lesion. There was no history of diarrhoea, constipation, alteration of bowel habits, haematemesis or melena, long-term fever, cough, shortness of breath and weight loss or decreased appetite. Any history of previous cancer, radiation exposure or trauma to the area was absent. The family history of cancer or history of similar lesion was also denied.

On examination, the swelling was a 1.5×2 cm, spherical, pedunculated, non-tender mass, firm in consistency and having a translucent surface with visible telangiectasia ([figure 1](#)). No regional lymph nodes were palpable. Examination of salivary glands, thyroid gland, breasts and the abdomen was unremarkable. Examination of other organ systems was also unremarkable.

INVESTIGATIONS

Baseline blood tests, stool for occult blood, urine routine examination and blood prostate antigen levels were all unremarkable.

A clinical diagnosis of basal cell carcinoma was made, and the excised tumour with 3–4 mm tagged surgical margins was sent to the laboratory in normal saline for margin clearance on frozen section. After obtaining margin clearance, the surgical defect was repaired on the same day with primary closure, wound dressed and the patient asked to return after 5 days for inspection and removal of alternate stitches. The remaining sutures were removed on the seventh postoperative day.

Detailed histopathology report obtained after a week revealed a normal epidermis with the dermis being infiltrated by neoplastic cells arranged in nests and clusters, forming acini-like structures at some places. These structures were floating in mucin-filled cystic cavities, separated by fibrous strands. The individual neoplastic cells were cuboidal to columnar in shape, had hyperchromatic nuclei with raised N:C ratio. The margins, peripheral as well as deep, were clear of tumour.

Immunohistochemical studies revealed that the tumour was positive for CK7, whereas it stained negatively for Cytokeratin 20 (CK20), CDX2, BerEp4 and Thyroid Transcription Factor 1 (TTF-1), indicating primary nature of the tumour ([figure 2](#)).

Ultrasound abdomen, kidney, ureter, bladder and prostate as well as the contrast-enhanced CT of the chest and abdomen were negative for any other primary source of the tumour or any metastases from the skin to any other site.

DIFFERENTIAL DIAGNOSIS

- Basal cell carcinoma.
- Myxoid carcinoma.

TREATMENT

Surgical excision with margin clearance, as described above (please see under the 'Investigations' section).

To cite: Javaid H, Raza N, Ejaz U, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-222546

Figure 1 Lesion near the right lateral canthus, with visible telangiectasia.

OUTCOME AND FOLLOW-UP

Based on detailed physical examination, laboratory investigations, histopathological and immunohistochemical studies, the diagnosis was confirmed to be primary mucinous cutaneous carcinoma. The patient has been advised regular follow-up.

DISCUSSION

Primary mucinous carcinoma of the skin (PMCS) was first described by Lennox *et al* in 1952.¹ Approximately 215 cases of PMCS have been documented in literature since then.²

It usually presents in the skin of the head and neck area as a slow-growing, small (0.7–8 cm), painless nodule that is soft and may be indurated or ulcerated. It is usually translucent, reddish or grey blue in colour, being present for many years at presentation.^{2,3}

PMCS typically affects elderly men (mean age 63.5 years) more commonly.⁴ Most cases arise on the face, especially on the eyelid and scalp, but rare presentations in the gluteal region, axillae, vulva and extremities have also been reported.^{3,4} There is an increased predilection to white rather than African-American, East Asian or Indian individuals.^{2,4}

Microscopically, PMCS is characterised by neoplastic epithelial cell nests floating in mucinous lakes. As compared with the metastatic mucinous lesions, these nests are more organised with lesser hyperchromasia and fewer mitoses.

Immunohistochemical markers (like CK7, CK20, CDX2, TTF1, D2-40 and p63)^{5,6} and thorough clinical work-up are required to make a distinction between primary mucinous carcinoma and secondary mucinous carcinoma deposits from other tissues. Primary mucinous carcinoma tumour cells, like in our case, are CK7 positive and CK20 negative, similar to cancer of the breast, lung and gallbladder but different from gastrointestinal adenocarcinoma because these cells are CK7 negative and CK20 positive. Lung, breast and gallbladder cancers can be differentiated from a primary skin tumour based largely on clinical investigation with potential assistance from other immunohistochemical stains.⁷

Mardi and Diwana described a similar case of the carcinoma near the lateral canthus of the left eye of a 70-year-old patient. The lesion was a raised, freely mobile, firm skin mass of approximately 3.0 × 2.5 cm. The case was clinically diagnosed as fibroma, but detailed investigations proved it to be a PMCS.⁸

Treatment is wide excision of the lesion because of high recurrence risk.⁹ The options include standard local excision and Mohs micrographic surgery. Our case was treated with modified Mohs surgery, as margin clearance was achieved using frozen section technique. Mohs surgery is suggested as the preferred option as it has less risk of recurrence when compared with traditional surgical excision.^{7,10} The systemic review of Kamalpour *et al* about the postsurgical outcomes of the tumour by the type of surgery performed (ie, Mohs vs conventional excision) revealed that Mohs surgery was associated with lesser risk of recurrence and metastasis (among the 15 cases treated with Mohs surgery, only 2 cases (13%) recurred, and there were no cases of metastasis; among the 136 cases treated with excision, 46 cases (34%) recurred or metastasised).² Thus, it is recommended that a wider margin of at least up to 10 mm should be excised, especially if intraoperative frozen section facility is not available. However,

Figure 2 Histopathological and immunohistochemical findings. (A) Haematoxylin and eosin stain (×40): mucinous carcinoma showing nests of neoplastic cells. (B) CK7 immunostain (×40): tumour cells are strongly positive for CK7. (C) CK20 immunostain (×40): tumour cells are negative for CK20. (D) CDX2 immunostain (×40): tumour cells are negative for CDX2. (E) BerEp4 immunostain (×40): negative BerEp4 (a marker for basal cell carcinoma). (F) TTF-1 immunostain (×40): negative (a marker for lung cancer).

narrow margins may be considered if close histology control by an experienced pathologist (using Mohs, slow-Mohs, Tübingen or spaghetti techniques) is performed to reduce the risk of local recurrence and metastasis. Radiotherapy and chemotherapy were not employed as these tumours are usually resistant to these tools.¹ Due to the high potential of recurrence, patients are counselled for regular follow-up sessions, but again no set guidelines are available to determine the exact length of follow-ups.

Factors that could be associated with poorer outcome of treatment, that is, recurrence or metastases, include young age at presentation, site of tumour on the trunk, size larger than 1.5 cm and long periods without follow-up.⁴ Prognosis is variable in patients of Asian ethnicity.^{2,4}

Patient's perspective

Translated from Urdu: "I had this lesion for around 2 years and I thought it was just something minor. Finally, when I decided to get it reviewed it turned out to be a cancer. But, thank God it was in the earlier stages and has been treated adequately. I'll be happy to help anyone if they want to study my case".

Learning points

- Any suspicious-looking skin mass should be investigated.
- It is important to differentiate between primary mucinous carcinoma of the skin and metastasised mucinous carcinoma presenting on skin with the help of detailed physical examination, relevant laboratory investigations and immunohistochemistry of the excised tumour.
- Prognosis of primary mucinous carcinoma of the skin is excellent when treated timely.
- Follow-up is a must to detect any recurrence.

Contributors HJ planned to write the case report; major role in acquisition and analysis of all the necessary investigations and information related to the

patient; wrote the case report after thorough literature review; participated in the final approval of the version to be published. NR supervised and revised the case report; major role in acquisition and analysis of data; provided the final approval of the version to be published. UE played a role in writing the case report draft; literature review; participated in the final approval of the version to be published. TS supervised and edited the case report. All authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

© BMJ Publishing Group Ltd (unless otherwise stated in the text of the article) 2018. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

REFERENCES

- 1 Breiting L, Dahlström K, Christensen L, *et al.* Primary mucinous carcinoma of the skin. *Am J Dermatopathol* 2007;29:595–6.
- 2 Kamalpour L, Brindise RT, Nodzenski M, *et al.* Primary cutaneous mucinous carcinoma: a systematic review and meta-analysis of outcomes after surgery. *JAMA Dermatol* 2014;150:380–4.
- 3 Wright JD, Font RL. Mucinous sweat gland adenocarcinoma of eyelid: a clinicopathologic study of 21 cases with histochemical and electron microscopic observations. *Cancer* 1979;44:1757–68.
- 4 Laco J, Simáková E, Svobodová J, *et al.* Recurrent mucinous carcinoma of skin mimicking primary mucinous carcinoma of parotid gland: a diagnostic pitfall. *Cesk Patol* 2009;45:79–82.
- 5 Levy G, Finkelstein A, McNiff JM. Immunohistochemical techniques to compare primary vs. metastatic mucinous carcinoma of the skin. *J Cutan Pathol* 2010;37:411–5.
- 6 Ivan D, Nash JW, Prieto VG, *et al.* Use of p63 expression in distinguishing primary and metastatic cutaneous adnexal neoplasms from metastatic adenocarcinoma to skin. *J Cutan Pathol* 2007;34:474–80.
- 7 Kazakov DV, Suster S, LeBoit PE, *et al.* Mucinous carcinoma of the skin, primary, and secondary. *Am J Surg Pathol* 2005;29:764–82.
- 8 Mardi K, Diwana VK. Primary cutaneous mucinous carcinoma: a rare entity. *Indian Dermatol Online J* 2011;2:82–4.
- 9 Beteddini OS, Sheikh S, Shareefi F, *et al.* Primary mucinous adenocarcinoma of the scalp: a case report and literature review. *Int J Surg Case Rep* 2015;10:241–4.
- 10 Papalas JA, Proia AD. Primary mucinous carcinoma of the eyelid: a clinicopathologic and immunohistochemical study of 4 cases and an update on recurrence rates. *Arch Ophthalmol* 2010;128:1160–5.

Copyright 2017 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- Submit as many cases as you like
- Enjoy fast sympathetic peer review and rapid publication of accepted articles
- Access all the published articles
- Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow