Multifocal, Recurrent Microcystic Adnexal Carcinoma (MAC) of the face: A Diagnostic, Therapeutic and Reconstructive Challenge

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Abstract

Educational Objective: At the conclusion of this presentation, the participants should be able to recognize the diagnostic and reconstructive challenges of treating microcystic adnexal carcinoma (MAC) of the face with Mohs micrographic surgery (MMS).

Study Design: Case report.

Results: This case highlights the diagnostic challenges presented by MMS for large facial defects; extensive resection required the patient to have serial sections over several days, with a widening facial defect and concern for wound exposure. Traditional frozen section analysis is not sensitive enough to detect small foci of infiltrative tumor. Modified, or slow Mohs, presents an advantage in which a broad loafing technique is used. Rush processed, paraffin embedded specimens that represent en face analysis of the margin (modified Mohs) was necessary to correct both false negative and false positive margins obtained with traditional Mohs.

Conclusions: MAC is a rare skin tumor with propensity for the central face. Typical defect size is 6-14 times the size of the original lesion. MMS has been shown to improve outcomes and spare wide margins around critical structures of the face; however modified Mohs may be necessary to clear margins prior to reconstruction of large defects.

Case Description

• 38-year-old male presented to the Dermatology clinic with a right cheek lesion, and biopsy-proven recurrent microcystic adnexal carcinoma in 2014
• Past medical history significant only for previous microcystic adnexal carcinoma (MAC) of the contralateral (left) cheek treated in 2011
• At that time the patient was treated with Mohs micrographic surgery (MMS) and local flap reconstruction, followed by radiation therapy.
• Pre-operative consultation, he was also noted to have a 3.5 x 4.5 cm area of thickening of palpation of the skin, without overlying ulceration or color change, at the angle of right mandible. This was concerning for an additional focus of tumor (compared to right cheek), which was confirmed via frozen section biopsy at the time of MMS.
• No communication between the two foci was discovered during MMS, and both were cleared following 13 MMS stages with 26 sections. The lesion appeared to be confined to the dermis.
• The final post-MMS defect size measured 9.8 x 14.4 cm.
• Right Selective Neck Dissection (levels I-III) as a result of CT scan with enlarge ipsilateral jugular nodes and right superficial parotidectomy.
• Reconstructed defect one week later with anterior lateral thigh free tissue flap and large right cervico-facial advancement flap.

Discussion

• Diagnostic dilemma
  • Newly detectable disease, multi focal primary of a rare entity
  • previous negative margins, presentation of disease far from the contralateral side of the face, failure of pathologic analysis to demonstrate a physical connection between the three separate foci of MAC detected
  • Versus recurrence of contralateral side
  • the distance between known positive margins was less than 2 cm, potential for false-negative margins because of the difficulty in tracking the tumor through tissue with traditional Mohs technique and permanent analysis which uses “bread-loafing”, the time-course of this slow-growing tumor could support recurrence, the known behavior of the tumor supports insidious perineural spread with very rare multifocality reported in the literature.

• Risk Factors
  • Radiation Exposure
    • Radiation Exposure has been mentioned as a risk factor for MAC however it is unlikely that this played a part in the course of our patient as MAC is a slow growing neoplasm with late stage presentation (3 years from first excision to second procedure).
  • Case Report in which patient presented with de-differentiated more extensive and histologically aggressive form of MAC 6 months after complete resolution with Radiation therapy treatment.

  • Ultraviolet Radiation
  • Predominance for the left face in a series of patients
  • Possibility of positive margins remains following traditional MMS, and some authors advocate sending an additional layer for “modified Mohs” after tumor clearance via frozen section
  • Particularly true for peri-ocular tumors or tumors with peri- or intraneural invasion
  • In modified Mohs, the specimen is fixed in formalin and embedded in paraffin for permanent sections, but is processed in face, similarly to the traditional frozen section Mohs technique
  • While this allows the pathologist to examine the entire involved margin, rather than representative sections of the margin (as in standard “bread loaf” processing), it also allows for a range of immunohistochemical stains, as discussed earlier.
  • This may be of particular importance with small foci of peri- or intraneural invasion, but should also be considered in cases such as ours, with an apparent multifocal growth pattern. The obvious drawback is the delay in processing required for permanent sections.


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References