



Surgical Management of Primary Cutaneous Mucinous Carcinoma

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Abstract

Primary cutaneous mucinous carcinoma (PCMC) is a rare non-melanoma skin cancer that most commonly presents in the face. This malignancy is difficult to differentiate from metastatic mucinous carcinoma with a visceral source. Thus, PCMC is a diagnosis of exclusion, after internal malignancy has been ruled out. Given its rarity, there is currently a paucity of sound evidence behind its diagnosis and management. We present the case of a 74-year-old male with recurrent mucinous carcinoma of the right lateral orbital rim and our approach to management.

Introduction

Primary cutaneous mucinous carcinoma (PCMC) is a rare non-melanoma skin cancer which represents 0.005% of all malignant epithelial neoplasms [1]. These adnexal tumours have been thought to be of eccrine origin, though this is currently under debate [2]. Clinically, PCMC presents as an indolent, papillomatous or pedunculated, solitary, flesh-coloured to translucent nodule which may or may not be ulcerated [3]. These lesions are capable of rapid enlargement [4]. Histologically, PCMC is a lobular dermal tumour with extension into the subcutaneous tissue and its composition consists of mucinous collections loculated by fibrous trabeculae [5]. Herein, we present the case of a 74-year-old male with recurrent mucinous carcinoma of the right lateral orbital rim and our approach to management.

Case Presentation

The patient is a 74-year-old man without any prior history of malignancy who initially presented to us three years ago with a soft tissue mass overlying the right lateral orbital rim. It had been stable for 15 years and only started to grow over the course of the preceding year. Clinically it presented as a benign subcutaneous cyst and was excised under local anesthesia. At surgery it looked more like a ganglion rather than a cyst. The histopathological examination of the specimen yielded findings consistent with PCMC. Because of this unexpected pathology report, the lesion was re-excised shortly after with a 1cm margin around the initial excision, and serial deep margins were examined intra-operatively under frozen section until the underlying zygomatic bone was reached. Permanent pathology showed complete lesion excision with clear margins.

The patient was subsequently referred to the regional oncology service to rule out a possible gastrointestinal (GI) source of mucinous carcinoma. He denied any complaints of GI symptoms or any family history of GI cancer, polyps or inflammatory bowel disease. The patient was cleared by the oncology service after clinical examination and colonoscopy biopsies which were unremarkable for GI malignancy.

The patient presented one year post the wide local excision with a similar looking lesion in the same location. An excisional biopsy revealed an epidermal inclusion cyst. Twenty months after the wide local excision, this patient presented to us for the third time for a possible PCMC recurrence with a 6-month history of growth. A re-excision was carried down to the periosteum overlying the lateral orbital rim, with permanent pathology documenting recurrent PCMC with clear resection margins. Post-operative head and neck CT scan did not show any residual disease or metastasis.

Currently, the patient is once again under the care of the oncology service. A multidisciplinary discussion with radiation oncology and head and neck oncology deemed radiation therapy to be reasonable for this recurrent condition. The patient was treated with 50 Gy in 20 fractions. He has been followed up closely without any signs of recurrence for three months since his last operation.

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Received Date: 16 Oct 2016

Accepted Date: 19 Dec 2016

Published Date: 27 Dec 2016

Citation:

Hanna SA, Hu J, Allard-Coutu A, Thoma A. Surgical Management of Primary Cutaneous Mucinous Carcinoma. *Clin Surg*. 2016; 1: 1262.

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Discussion

PCMC tends to occur in the periocular region and has a 20% incidence rate in the face [6]. The regional and distant metastatic rates for this tumour have been estimated to be 10% and 3% respectively, with few reported cases [7-11]. With respect to demographics, these tumours tend to occur most frequently in Caucasian and African-American males over the age of 60 [3]. PCMC generally has a favourable prognosis, however, local recurrence following excision is quite common and has been reported to occur in up to 36% of cases [1,2].

When making a diagnosis of PCMC, special attention must be paid to ruling out the presence of a primary tumour elsewhere. Systemic imaging and tissue-specific staining may aid in this endeavour, however there are currently no established guidelines to this end. Sites to consider include the breasts, lungs, ovaries, prostate, gastrointestinal tract, pancreas and kidneys [1,11]. PCMC is difficult to differentiate from metastatic visceral mucinous carcinoma as they may appear remarkably similar histologically [5,12]. Positivity for tumour protein p63 is associated with adnexal origin and may be used to support a diagnosis of PCMC [13]. It is worth noting that metastatic mucinous carcinoma very rarely presents in the face and rather presents in the skin overlying, or in close proximity to, the affected viscera [14,15]. The authors suggest a low threshold to biopsy lesions suspicious for PCMC. Histopathological studies demonstrating large loculated pools of mucin and positivity for tumour protein p63 support a diagnosis of PCMC after a visceral source has been ruled out.

There is currently no high level evidence to guide management of this malignancy, as the majority of publications are case reports. With this in mind, local excision is preferred over radiation and chemotherapy given the high morbidity of these options as well as the low mortality and low metastatic potential of the neoplasm [2,5,7-10]. Furthermore, as per Snow and Reizner [11] the available literature seems to indicate that PCMC may be resistant to chemotherapy and radiotherapy. Due to the overall rarity of this tumour, there is no standard of practice for its surgical management. However, typical resection margins for these tumours are between 1.5 to 2 cm [3]. In light of this and the fact that these tumours most commonly occur in locations that present a reconstructive challenge, it would behoove the practicing surgeon to be aware of this diagnosis and its management.

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