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Case Report

Malignant cylindroma of the scalp treated with staged perimeter excision: A case report and literature review[☆]

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ABSTRACT

Malignant cylindroma is a rare and poorly understood cutaneous malignancy. There is a paucity of literature on these lesions, with only a select number of case reports and limited guidelines on management. We present a case of a 60-year old patient with a malignant cylindroma of the scalp treated surgically with staged perimeter excision and summarize our review of the literature with a focus on management of this potentially aggressive disease.

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Introduction

Cylindromas are rare tumours of the apocrine or eccrine glands classified within the family of skin adnexal tumors. The term cylindroma was first used by Billroth in 1859 to describe an orbital tumour

[☆] The current work has not been presented in any part in another meeting.

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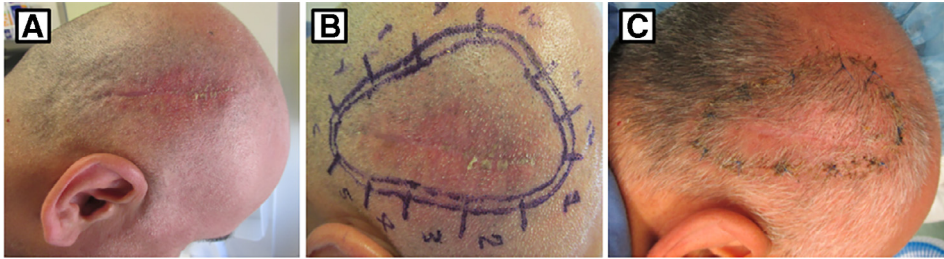


Figure 1. Perimeter excision resection method for malignant cylindroma. (A) Initial patient presentation with scar resulting from two previous excisional biopsies. (B) Pre-operative markings for perimeter biopsies with pathological specimens labelled 1–12 for separate analysis. 2 cm margins marked with wider margins favoured superiorly due to positive margins on previous resection. (C) Post-operative photograph with prolene stitch marking each separate biopsy specimen and remainder of incision closed with gut suture (Patient consent was obtained for inclusion in this case report and for photo-documentation).

with cylindrical shapes seen on histology¹. These cutaneous lesions are typically categorized as other skin cancers according to their malignant potential.

Benign cylindromas (BC) occur as solitary or multiple lesions. The lesions are usually slow-growing, firm, pink to red in colour, and commonly within the head and neck region². These lesions are typically painless, and surgical intervention is often not required. The presence of multiple cylindromas is associated with hereditary conditions including familial cylindromatosis (FC) and Brooke-Spiegler syndrome (BSS). FC is characterized by multiple cylindromas, whereas patients with BSS are also affected by trichoepitheliomas and spiradenomas³. These lesions can cause disfigurement and alopecia, sometimes requiring surgical excision. These familial syndromes have been shown to have an increased incidence for malignant transformation of benign lesions⁴.

In rare instances, malignant cylindromas (MC) occur in patients without any known hereditary risks. These cutaneous lesions arise from benign lesions or *de novo*, and tend to behave aggressively with a high risk of recurrence and metastasis⁵. First described in 1929 by Wiedman, less than 50 cases of MC have been reported⁶. As such, there lacks information regarding the etiology, histogenesis, treatment options, and recurrence rates of MC². Wide local excision with or without adjuvant radiotherapy is the most common treatment method^{5,7}. Other treatment modalities include Mohs micrographic surgery (MMS)⁶. We report a rare case of a solitary MC in a 60-year-old patient with no family history of associated syndromes, managed with staged perimeter excision (SPE, also known as spaghetti technique) and close follow-up.

Case presentation

A 60-year-old man initially presented to his family physician with a three-year history of a persistent firm, raised nodule in the right parietal region. He reported a rapid growth of the lesion over two months, prompting referral to a community surgeon. The patient was otherwise well with no constitutional symptoms and no significant past medical history.

The surgeon performed an excision and primary closure, and the pathology reported the lesion to be consistent with MC with positive superio-medial and deep margins. The surgeon attempted a re-excision with 5mm margins. Unfortunately, margins remained positive after the re-excision and the patient was referred to a tertiary centre. Staging CT showed no evidence of regional or distant metastasis. The case was presented at Cutaneous Multidisciplinary Rounds, where wide excision with perimeter margin control was recommended. As MMS is not available at our centre, SPE⁸ was planned for margin clearance prior to definitive resection.

2cm margins were marked outside of the pre-existing scar, except the superio-medial margin where a 3cm margin was marked due to previous positive margins. Under local anesthesia, 2mm wide perimeter biopsies were taken circumferentially (Figure 1) and closed primarily. Fortunately, all initial 12 pathology margins were clear, and the patient was scheduled for a definitive resection.

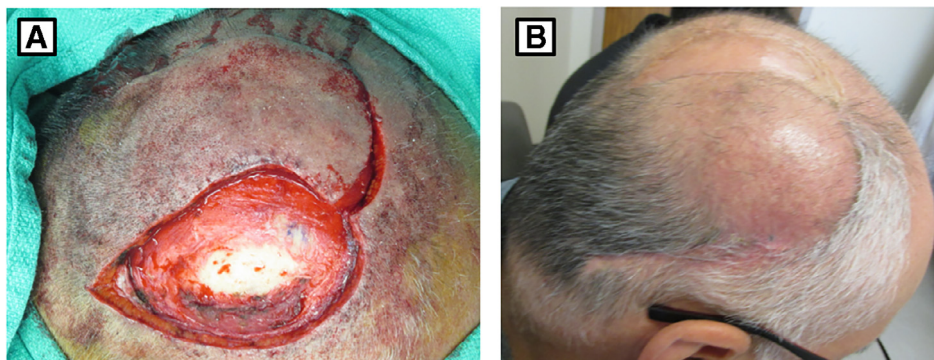


Figure 2. Definitive resection of malignant cylindroma. (A) Intra-operative photograph with definitive resection of malignant cylindroma at level of galea with frozen section control. Bone burred centrally to ensure local control. (B) 3 months post-operative. (Patient consent was obtained for inclusion in this case report and for photo-documentation)

Under a general anesthetic, the lesion was resected en-bloc using the predetermined margins. Dissection was carried deep, and the lesion was removed in the galeal plane. The lesion was not found to be adherent to underlying bone and the intraoperative frozen sections of the deep margin were negative. However, due to its aggressive behavior, the decision was made to burr the outer table of the calvarium to ensure local control (Figure 2). A large local transposition flap was designed over the occipital vessels and transposed into the defect. The donor site was covered with split thickness skin graft from the thigh. Final pathology demonstrated clear margins and the team decided not to proceed with any adjuvant therapy. The patient is being closely followed clinically and has no signs of recurrence at six months.

Pathological features

Multiple sections of the initial biopsy and re-excised tissue demonstrated infiltrative growth of the dermis and subcutaneous fat tissue (Figure 3 A–C). The tumor exhibited individual nodules composed of undifferentiated basaloid cells with small darkly-stained nuclei and scant cytoplasm. Focal atypical features were noted, including irregular asymmetrical proliferations of multiple nodules and strands of hyperchromatic cells with reduced basement membrane material, and loss of jigsaw pattern. The cells showed prominent cytological atypia with nuclear pleomorphism and increased mitotic activity; the nests stained positively for cellular proliferation marker Ki-67⁹. These features are consistent with a well-differentiated MC.

Literature review

An electronic literature review was conducted on PubMed and Western University library database to identify case reports describing MC using the search terms “malignant cylindroma,” “cylindrocarcinoma” and “transformation cylindroma”. A summary of these cases is shown in Supplementary Table 1.

Discussion

MC is a rare and aggressive cutaneous malignancy. The literature is sparse on its natural history and treatment options. Our current literature search revealed a total of 36 English published cases (Supplementary Table 1). Of the reviewed cases, one third had a family history of cylindromas or a diagnosis of BSS. Surgeons should be aware of this genetic connection and elicit appropriate family history during initial consultation. The natural history of these lesions is thought to occur most often through malignant transformation of BC. This can present as slow and indolent change, leading to late

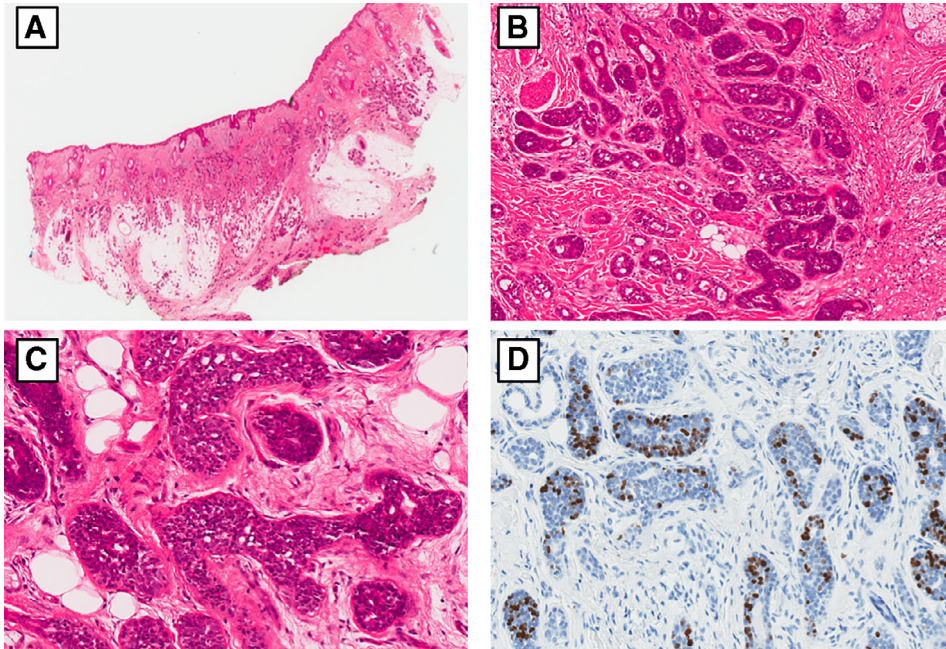


Figure 3. Histological features of malignant cylindroma. (A) Hematoxylin and eosin (H&E) staining of paraffin fixed tissue biopsy specimen viewed at 2× magnification. The tissue section demonstrates cylindroma-like nests infiltrating into the dermis and subcutaneous tissues. (B) Higher 20× magnification of the tissue specimen showing irregular aggregates of malignant cylindroma cells with focal absence of basement membranes and loss of ‘jigsaw’ pattern. (C) 20× magnification highlight cytological atypia. (D) Immunohistochemical staining for Ki-67, Positive stained cells appear brown. (Patient consent was obtained for inclusion in this case report and for photo-documentation)

diagnosis and distant metastasis⁷. In other patients, the malignant transformation is noticed by sudden rapid growth, pain, and bleeding. This lesion may also arise de novo without a pre-existing lesion and behave aggressively early in the course of disease¹⁰. Prompt biopsy and referral for a definitive removal is crucial for a patient who reports any change in tumour appearance.

Histologically, BC show irregular islands of basal cells surrounded by a hyaline sheath². MC are characterized by loss of hyaline sheath, anaplasia, pleomorphic nuclei, abundant mitosis, stromal invasion, loss of palisading at the periphery, and loss of “jigsaw puzzle” arrangement⁶.

From the literature review, MC is generally managed with wide excision or MMS⁵. A few patients with advanced disease were managed with adjuvant radiotherapy and chemotherapy⁷. The low number of reported cases preclude accurate comparison of recurrence rate with various treatment modalities. Among cases that describe long-term outcome, 33% of patients either presented with recurrence, or developed a recurrence or distant metastasis during follow-up. Shortest time to the detection of metastasis was eight months, however local recurrence was detected up to seven years after the initial excision.

We performed a SPE to ensure clear margins before definitive resection. SPE has been described for other skin lesions with a propensity for positive margins in cosmetically sensitive areas⁸. This decreases the necessity of multiple procedures and limits the size of resection while maintaining oncological safety. This option is especially useful in aesthetically sensitive areas and when MMS is not available.

Conclusion

The etiology and pathophysiology of MC remains unclear. MC should be regarded as highly aggressive lesions, with the potential for local invasion and metastasis up to seven years after initial

treatment. Wide excision should be the treatment of choice. Currently, there are no established clinical guidelines. Close long-term follow-up is necessary for possible recurrence and metastasis. Discussion in a multidisciplinary team may be considered as well, along with other treatment modalities such as lymph node dissection, chemotherapy, and radiotherapy.

Conflicts of interest

The authors declare no potential conflicts of interest.

Supplementary material

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.jpra.2019.04.003](https://doi.org/10.1016/j.jpra.2019.04.003).

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