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Case Report

Malignant pilomatrixoma case report: what kind of treatment is the most suitable?

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ABSTRACT

Malignant pilomatrixoma is a rare malignancy with just 136 cases reported in the English literature. The tumour is described as locally aggressive, with possibility of reoccurrence and distal metastasis. Histology remains the gold standard in diagnostic. Wide surgical excision is preferred to simple excision. In the present study a case of first malignant pilomatrixoma in our hospital is described. We made an excision of the tumour and the tissue was sent on histopathology. The skin defect was covered with a local ulnar flap. Through our case report and literature review we focus on the most suitable treatment option. The tumour was not excised with appropriate safety surgical margin based on the histopathology report. Patient refused another excision and any additional treatments. The most optimal treatment has not yet been established. Most of the article's state that primary wide surgical excision should be done, with surgical margins still being debateable. The best results were given when using Mohs micrographic surgery.

Keywords: Tumour, Skin, Rarity, Histology, Surgery

INTRODUCTION

Malignant pilomatrixoma, in other words calcifying epitheliocarcinoma of Malherbe was first described by Lopansri and Mihm in 1980.[1-5] It is an extremely rare malignant hair follicle carcinoma.[2,4,6-9] Until now there were just 136 cases reported in the English literature.[9] The tumour is locally aggressive, has a strong tendency to reoccur and can metastasize to lungs, spine, abdominal viscera, regional lymph nodes and brains.[1,2,4,6-9] The most common sites of tumour presentation being head and neck nevertheless it can also present on the back, upper and lower extremity, chest inguinal and axillary regions and on the buttocks.[1,5,6,8,9]

This is the first known case of malignant pilomatrixoma in our department. There is not a lot information available on what type of treatment of for this malignancy is the best course of action.[2,5] Most of the authors recommend wide surgical excision, with surgical margins from 0.5 centimetres (cm) to 2 cm.[1,2,3,5] Through our case report and review of the literature, we would like to discuss what kind of treatment is the most suitable.

CASE REPORT

64-year-old male was admitted to the Department of Plastic Surgery and Burns, University Medical Centre (UMC) Ljubljana in April 2019, with a tissue mass on the right forearm first noticed two years ago (Figure 1). Upon

examination, it was a 6x5 cm, firm, non-tender mass, superficial and easily mobile. It gave a view of an ectopic ossification or calcification. No associated lymphadenopathy was seen. At first cytological punction was advised, which showed cystis epidermoidei, partially calcified. We advised an extirpation of the cyst.

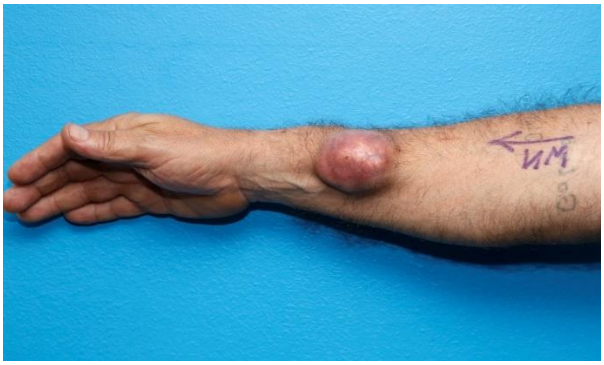

Figure 1: Pre-operative clinical presentation of malignant pilomatrixoma.

The mass was excised, and the tissue was sent on histopathology. The skin defect was covered with a local ulnar flap. The gross specimen was an elliptical, 75x49 mm large sample of skin with 20 mm of soft tissue. In the soft tissue, a 55x44x35 mm large, firm tumour mass was observed. Cut section showed unilocular cystic formation, with calcified content.

Microscopy revealed a tumour composed of proliferating large anaplastic hyperchromatic basophilic cells (Figure 2). Numerous atypical mitotic figures were seen. Areas of necrosis without the tumour's infiltration into adjacent tissues were observed. The tumour involved the whole layer of the skin, mostly the subcutaneous tissue. There was no evidence of any lymphovascular or perineural invasion. However, even though the inked surgical margin was free, the tumour was very close to it (0.5 millimetre (mm) away). The diagnosis of malignant pilomatricoma was made. Because the tumour was not excised with the appropriate safety surgical margin based on the histopathology report, another excision was advised, but the patient did not agree with it.

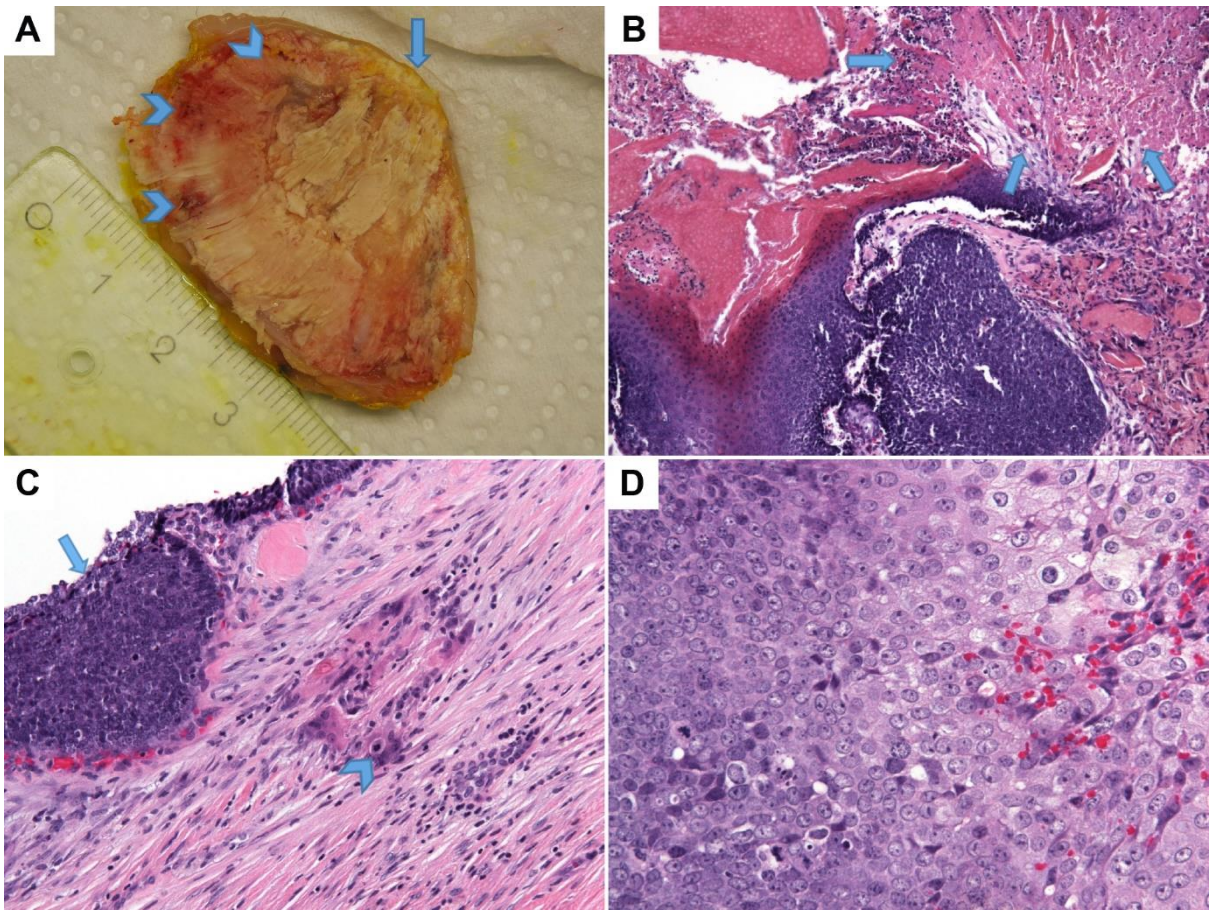

Figure 2. (A) Malignant pilomatrixoma. Cross section of the calcified tumour showing skin infiltration (arrow) and focal haemorrhagic necroses (arrowheads); (B) the latter is showed also histopathologically (arrows). (C) In addition to areas of proliferating atypical basaloid cells with scanty cytoplasm (arrow) with numerous mitoses, small infiltrating islands of pleomorphic squamous tumour component with atypical mitoses (arrowhead) are present as well. (D) Higher magnification of pleomorphic epithelial cells with prominent nucleoli and mitoses. Haematoxylin-eosin, original magnifications 100x (B), 200x (C), and 400x (D).

The patient was sent to the Institute of Oncology Ljubljana for further investigation and treatment. The ultrasound of axillary nodes showed chronical changes. Additional radiotherapy was advised, but the patient refused the treatment. Up till now the patient is doing well but has been advised to do an ultrasound of the abdomen and a chest X-ray and follow ups every 4 months.

At the last follow up in October 2020 at our institution the local ulnar flap was in place, the postoperative scar was without any inflammation. The patient had no complaints.

DISCUSSION

Malignant pilomatrixoma is a rare cutaneous adnexal tumour, with a male predominance, mostly occurring in middle-aged adults.[1-3,5,6,9] Clinical presentation of a tumour is most commonly asymptomatic as a single, hard, firm, slow growing, painless, dermal, or subcutaneous mass, with bluish or flesh-coloured skin discoloration or ulceration.[1,2,5] It can also present as a rapid growth of a longstanding mass or rapidly enlarging new mass.[2] Tumour can be ranged from 0.5 cm to 20 cm in size, mostly presented on the face and in around 10% in the upper extremity.[5,8] The clinical differential diagnosis can vary from an epidermal cyst, benign or proliferating pilomatrixoma, to vascular lesions.[1,9] The only possible way to differentiate between malignant pilomatrixoma and other lesions is by histologic evaluation.[5] Atypical and frequent mitoses, central necrosis, basaloid cells with nuclear pleomorphism, ulceration, perineural and vascular invasion are the most reliable indicators of malignancy.[2,4,5,7]

The main treatment option, by most of the authors, is surgical excision.[4] Some authors believe that local reoccurrence and metastasis are particularly common when simple excision, rather than wide excision is performed.[3,5] Although Melancon et al established that the differences in metastasis rate did not reach significance.[5] Wide excisions should comprise surgical margins from 0.5 cm to 2 cm even though clear recommendations are not yet established.[1-3,5,7] Herman et al compared 6 patients with wide excisions and 6 with simple excisions and in all patients the tumour reoccurred and metastasized.[8]

We performed surgical excision with surgical margin of 0.5 mm and by complying with the reconstructive ladder, as it was given 3; we closed the exposed tissue with a local ulnar flap. After reading the histopathologic report we realized we did not use the appropriate surgical margin and that a re-excision was advisable. A series of radiation therapies were considered. Radiation therapy can be used alone or in the metastatic disease and reoccurrence 8, however its potential in stopping the disease progression is rather debatable.[2] Although Papadakis et al in their report suggests that radiotherapy is a good alternative in those cases where adequate surgical excision is not possible.[1]

Liu et al describes positive effects on the local disease control in combining adjuvant radiotherapy with a wide surgical excision 2, while Aherne et al state that the role of adjuvant radiotherapy is unclear.[7] In contrast, systemic chemotherapy has not proven any positive effects.[5]

As an alternative to the previously described treatments Mohs micrographic surgery has been proposed, which offers complete tumour margin control. Till this date there were at least 2 cases treated successfully in this way.[5,8] Despite its effectiveness numbers of treating patients are too small for reaching any definite conclusions.

CONCLUSION

Malignant pilomatrixoma is a rare hair follicle tumour, locally aggressive, has a propensity for local reoccurrence and distant metastasis. The most optimal treatment has not yet been established. Most of the article's state that primary wide surgical excision should be done, with surgical margins still being debateable. Radiation therapy gave mixed results and its role as an adjuvant therapy or as a therapy in halting the disease progression is still unclear; chemotherapy is ineffective. The best results were given by Mohs micrographic surgery. The problem is that the number of patients receiving it, was too little to make any final decisions in its effectiveness.

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