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# Dermatofibrosarcoma Protuberans: The role of tissue expansion in reconstructive surgery of the scalp

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ABSTRACT

*INTRODUCTION:* Dermatofibrosarcoma Protuberans is an uncommon tumour, making up less than 0.1% of all malignancies. With regards to soft tissue tumours; this pathology is thought to make up less than 2% of the sum total. Traditionally treatment has been wide local excision, with or without adjuvant radiotherapy.

*PRESENTATION OF CASE:* We present a case of a 42 year old man referred by his GP with a lump on the right parietal region of the scalp. An USS done by his GP revealed a complex hypoechoic cystic mass, some 2 cm × 1 cm × 2 cm. Excision biopsy was performed and on review of the pathology it was noted that the lesion was a Dermatofibrosarcoma Protuberans. Due to the relatively low grade of this sarcoma, it was decided to treat with wide local excision with 2–4 cm margins. The expected residual scalp defect would be difficult to close with local flaps. To facilitate closure tissue expansion was undertaken for 6 weeks prior to definitive surgery.

*DISCUSSION:* With regards to tumours of the head and neck, use of a tissue expander has been recommended to improve cosmetic outcomes following respective surgery with wide margins. Ultimately the timing of tissue expansion i.e. before/after resection of the tumour, must weight the risk of delayed resective surgery on prognosis against the benefits of this reconstructive technique.

*CONCLUSION:* Head and neck tumours requiring careful reconstruction may benefit from tissue expansion to provide adequate volumes of matching soft tissue, as shown in this case.



## 1. Introduction

Dermatofibrosarcoma Protuberans is an uncommon tumour, making up less than 0.1% of all malignancies [1]. With regards to soft tissue tumours; this pathology is thought to make up less than 2% of the sum total [2]. There is a relatively low incidence of distant metastasis (<5%), even though the tumour can rapidly invade adjacent structures [1]. Recurrence is the major issue associated with this pathology, with a rate of up to 60% without wide margins [3]. Badeau et al. suggest surgical margins of between 2 and 3.5 cm will reduce the recurrence rates to around 8%.

## 2. Presentation of case

We present a case of a 42 year old man referred by his GP with a lump on the right parietal region of the scalp. The patient reports the swelling had been present for many years, but in the last few months it had gradually increased in size. He states that he had first noticed the lump after being bitten by a tick in this area. As it had grown in size it was becoming tender to touch; he himself denied any fevers or infective symptoms. An USS done by his GP revealed a complex hypoechoic cystic mass, some 2 cm × 1 cm × 2 cm. There was no significant past medical history or family history reported by the patient.

He was seen in the maxillofacial outpatients' clinic, where the decision was made to perform an excision biopsy of the lesion under local anaesthetic, on the presumptive diagnosis of an epidermoid cyst. Intraoperative findings were unremarkable and the sample was sent for histopathology. Upon review of the pathology it was noted that the lesion was a Dermatofibrosarcoma Protuberans (see Fig. 1). Clinical staging was completed with full body PET CT and MRI scalp to vertex (see Fig. 2) with final staging of T1N0M0, according to AJCC criteria. The case was discussed in the Head and Neck multidisciplinary meeting. Due to the relatively low grade of this sarcoma, it was decided to treat with wide local excision (WLE) with 2–4 cm margins. The expected residual scalp defect would be difficult to close with local flaps. To facilitate closure tissue expansion (see Fig. 3) was undertaken for 6 weeks prior to definitive surgery.

It was noted that the tissue expander was losing some volume between episodes of inflation and required a protracted expansion

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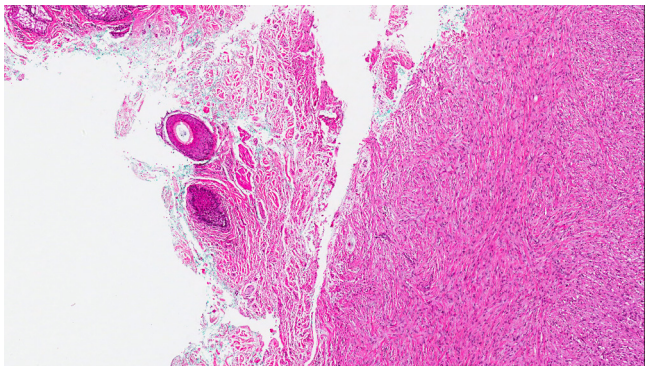

**Fig. 1.** Histological appearance of Dermatofibrosarcoma Protuberans. The tumour is located in the dermis and composed of fascicles of spindle tumour cells forming a protruding mass (hematoxylin and eosin × 2).

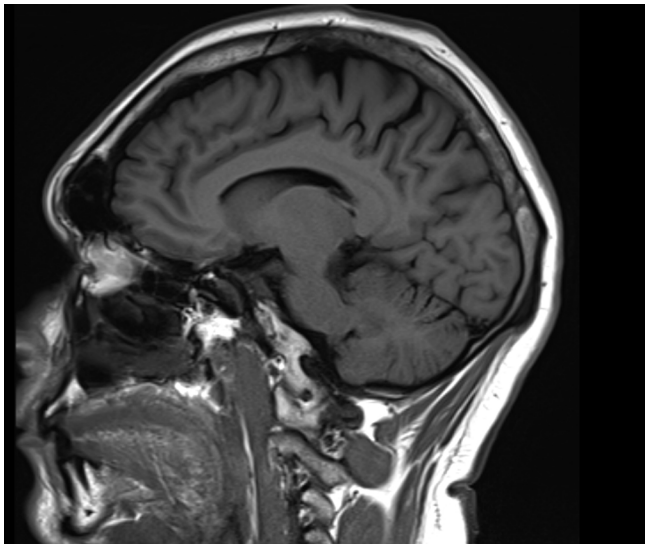

**Fig. 2.** MRI post initial procedure which shows small localised soft-tissue lesion in the right fronto-parietal region (measuring up to 1.5 cm) with no extension to the underlying bone.

period of nine weeks to achieve the desired tissue volume. At the time of definitive surgery the expander was removed and wide local excision of previous scar performed with 2 cm margins and also included pericranium (see Fig. 4). Histopathology from this specimen showed some residual Dermatofibrosarcoma Protuberans (33 mm × 15 mm) with clear margins; 2 mm deep and greater than 2 cm in all peripheral margins.

Our patient has been attending regular follow-up in the post-operative period, with no evidence of recurrence at this early stage.

## 3. Discussion

Traditionally treatment has been wide local excision, with or without adjuvant radiotherapy [1]. Studies have put forward that those patients that have positive or close margins may show lower recurrence rates, if treated with radiotherapy [1]. MOHS micrographic surgery has also been suggested to significantly decrease the rate of recurrence, when compared to surgery alone (6.3% vs 1.11%) [4]. Recurrence may also be higher due to the fact that lesion can be misdiagnosed as being benign [1]. Lesions in the head and neck have been show to be more likely to recur, with rates of up to 75%. It is possible that suboptimal surgery due to cosmetic concerns may be the cause of such a high recurrence rate [4].

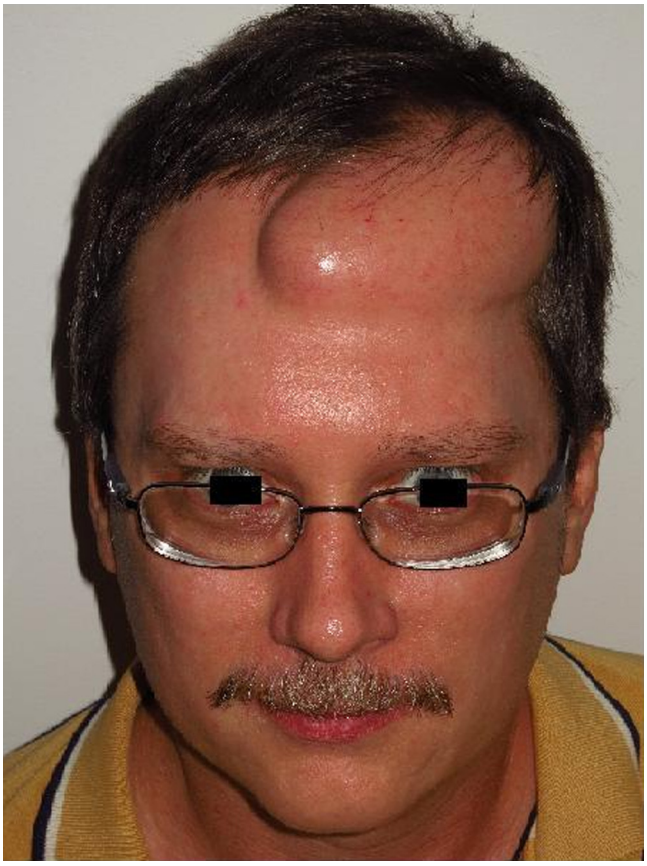

**Fig. 3.** Clinical picture with tissue expander in-place, in the front-parietal region.

**Fig. 4.** Clinical picture of the patient 1 week post-op after wide local excision of residual DFSP.

With regards to tumours of the head and neck, use of a tissue expander has been recommended to improve cosmetic outcomes following respective surgery with wide margins. Considerations when contemplating tissue expansion include the type and size of expected tissue defect, the availability of an expandable tissue plane such as the scalp, cheek or neck, the number and site of aesthetic units that will be affected. Ultimately the timing of tissue expansion i.e. before/after resection of the tumour, must weight the risk of delayed resective surgery on prognosis against the benefits of this reconstructive technique [3].

Treatment of Dermatofibrosarcoma Protuberans requires surgical excision with wide margins to prevent recurrence of the disease and regional or distant metastasis. While some suggest that MOHS micrographic surgery may provide lower rates of recurrence, there is no definitive evidence that this is the case [4]. Studies have also shown that the addition of radiotherapy may also have a positive impact on outcomes. Though such decisions should be made with a multi-disciplinary approach and take into consideration the grade of sarcoma as well as the presence of adverse prognostic factors especially margins.

## 4. Conclusion

Head and neck tumours requiring careful reconstruction may benefit from tissue expansion to provide adequate volumes of matching soft tissue, as shown in this case. Though this comes at the cost of delaying definitive cancer surgery, frequent patient visits for the addition of volume and an additional operative procedure to insert the device.

## Conflict of interest

None.

## Funding

None.

## Ethical approval

No ethics approval was required for this case report.

## Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

## Author contribution

Richard Maguire: Primary author and constructor of most of the body of report etc.

Dimitrios Nikolorakos—provided the case and pictures of the patient pre and post-op. Also assisted in correction of the draft to create final document. Alfred Lam—provided histopathology slide for the report.

## Guarantor

Dr Richard Maguire.

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