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Case Report

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Resection of a large scalp neurofibroma using multiple overlapping string sutures to control vascularity

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ABSTRACT

We report the excision of a large diffuse scalp neurofibroma in a 27-year-old patient with neurofibromatosis type 1. Two previous surgical attempts at removal had to be abandoned due to uncontrollable intra-operative haemorrhage. A haemostatic technique utilising multiple overlapping interrupted sutures prior to resection and skin graft enabling complete resection of the neurofibroma is described.

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Introduction

Neurofibromatosis type 1 (NF-1) is a neurocutaneous disorder that can be mapped to chromosome 17. The incidence is approximately 1 in 4000. Its classical features include dermal or plexiform neurofibromas, café-au-lait spots, freckles in the axilla and groin, optic nerve gliomas, Lisch nodules (iris hamartomas), and bony dysplasia.¹

Neurofibromas are benign tumours of the peripheral nerves of Schwann cell origin. Diffuse neurofibromas are an unusual form of this entity originating from tissue nerve fibres in the trunk, head and neck.² Neurofibromas can be unsightly and result in discomfort or pain. In the case of scalp neurofibromas, intracranial extension of an extracranial tumour has been described.³ Furthermore, rapid increase in the size of scalp neurofibromas may, as well as potentially being an indication of malignancy, result from traumatic or spontaneous intra-tumour haemorrhage.⁴

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We report a case of complete excision of a giant scalp neurofibroma using a novel technique of preskin incision sutures to control intra-operative haemorrhage.

Case report

A 27-year-old middle eastern male presented to the Neurosurgical department at Al-Basra Teaching Hospital with a large, pedunculated scalp mass involving the vertex and occipital area. The mass had been present since the age of one, initially as a small occipital nodule. It gradually increased in size during childhood and adolescence, but was not symptomatic and on inspection a primary diagnosis of a scalp lipoma was made. At the age of 16 the patient noticed a significant increase in size of the scalp mass to approximately 5×7 cm. At that time he underwent surgery with the aim of removing this lesion, but the procedure had to be aborted due to severe intra-operative haemorrhage. 5 years later, at the age of 21, he had a further attempt to have the lesion excised, but again, the procedure had to be terminated due to uncontrollable haemorrhage. Subsequently, the patient was averse to having further surgery. However, by the age of 27 the mass had further increased in size and was causing headache and neck pain radiating to both shoulders. The patient was also not able to sleep comfortably due to the mass itself. He dealt with the cosmetic problem of the mass by wearing a traditional hat.

On macroscopic examination (Figure 1, 2), the tumour measured 20×25 cm with a 14×17 cm scalp attachment. There was hyperpigmentation of the skin cover and evidence of previous surgical scars. The mass was soft, non-tender and freely mobile over the skull vault. On palpation, it felt multinodular and non-pulsatile; it was, however, compressible, which raised the suspicion of it having a rich vascular supply. There was no trans-illumination. On further external examination, the patient was found to have multiple café-au-lait spots, especially over the trunk and legs, two Lisch nodules and freckling in several skin folds. When taking his family history it transpired that his father had a nerve tumour removed surgically from his left arm several years previously. A diagnosis of Neurofibromatosis type 1 (NF-1) was made.

The patient underwent a skull X-ray and CT scanning of the head, both of which demonstrated intact, non-eroded cranial bone and no evidence of intracranial extension. The MRI of the head showed a large extracranial mass with heterogenic contrast enhancement. MR angiogram was performed demonstrating enlargement of the occipital arteries, which were the main blood supply of the mass.



Figure 1. Macroscopic view of the neurofibroma. The hyperpigmentation of skin is clearly visible. Note the scar from previous attempts at resection across the top of the mass.

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