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Reconstruction of a scalp and skull defect with free latissimus dorsi myocutaneous flap following dermatofibrosarcoma protuberans

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ABSTRACT

A 53-year-old male underwent more than 10 surgical treatments over 14 years, including a simple excision and local flap transfer, after recurrences of dermatofibrosarcoma protuberans of the head. Clinical results indicated that simple excision of dermatofibrosarcoma protuberans should not be performed as initial treatment. Instead, free flap transplantation, which permits a wide excision and complete reconstruction, should be the first treatment option.

Key words: *dermatofibrosarcoma protuberans; latissimus dorsi myocutaneous flap; skull defect*

INTRODUCTION

Dermatofibrosarcoma protuberans was first described as a progressive and recurring dermatofibroma by Darier and Ferrand in 1924[1,2] and was renamed dermatofibrosarcoma protuberans by Hoffman in 1925.[1,2] Dermatofibrosarcoma protuberans tends to occur on the trunk and proximal region of the extremities while it rarely develops on the head. It frequently recurs locally due to incomplete excision[1–3] but rarely metastases.[4]

We treated a patient who had undergone simple excision alone as the initial treatment of dermatofibrosarcoma protuberans on the scalp. More than 10 surgical treatments were followed, including simple excisions and local flap transfer due to recurrences of the tumours over a 14-year period, with additional metastases to the brain, scalp, and the skull. For the scalp and skull defects, we performed reconstruction by free latissimus dorsi myocutaneous flap transfer. Outcomes were satisfactory in terms of improved quality of life of the patient.

CASE REPORT

The patient noticed a mass 2–3 mm in diameter in the right occipital region at the age of 37 years. At the age

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of 39, an orthopaedic surgeon performed a simple excision of the mass as initial therapy. Histological examination revealed a uniform population of slender spindle cells arranged in a distinct storiform pattern, and the tumour was diagnosed as dermatofibrosarcoma protuberans. Recurrences occurred, however, and the patient underwent 11 simple excisions and a local flap transfer in several institutions including the departments of orthopaedics, thoracic surgery, and neurosurgery. He also underwent removal of lung tumours twice, at ages 50 and 52, respectively, due to dermatofibrosarcoma protuberans metastases.

At the age of 53 years, he underwent a twelfth simple excision, as well as artificial dura mater grafting and local flap transfer, which were the standard treatment for recurrent tumours. Postoperatively, however, cerebrospinal fluid leaked from the wound and meningitis ensued due to failure of union of the sutures. After treatment of the meningitis by antibiotics, a thirteenth procedure was attempted by a dermatologist to close the wound using a local rotation flap. However, the local flap did not survive and necrosis developed. The patient was therefore introduced to our department.

Physical examination revealed a skin defect of about 8 cm in diameter due to skin necrosis in the right lateral occipital region. The artificial dura mater and outflow of cerebrospinal fluid was observed (Fig. 1). Magnetic resonance imaging revealed a partial defect of the skull and multiple metastases to the brain. Wide excision of the tumours was found to be impossible.

Two weeks later, a free latissimus dorsi myocutaneous flap transfer was performed to cover the scalp and skull defects. The operation was started with the patient in the left side decubitus position. The scalp defect was 13 cm in diameter after excision of the skin necrosis. The superior thyroid artery and external jugular vein were ablated from the right neck for preparation of the recipient vessels.

A latissimus dorsi myocutaneous flap measuring 40 cm × 10 cm was harvested from the right back. The donor site was closed primarily. In the right neck, the superior thyroid artery and external jugular vein were anastomosed end-to-end to the subscapular artery and vein, respectively. The exposed portion of the latissimus dorsi myocutaneous flap was covered with a split-thickness skin graft. Urokinase was administered at 240 000 units per day for one week to prevent thrombosis. The latissimus dorsi myocutaneous flap survived completely (Fig. 2).

There was no recurrence of cerebrospinal fluid leakage or meningitis postoperatively. The quality of life improved for the patient as he was allowed to take a bath. Two months after the latissimus dorsi myocutaneous flap transplantation, further radiation therapy was performed for dermatofibrosarcoma protuberans of the head. A total of 120 gray of gamma irradiation was administered, and did not affect blood supply to the myocutaneous flap. However, the patient died one year after transplantation surgery due to metastasis to the abdominal cavity.

Figure 1 Artificial dura mater (arrow) was exposed due to the skin defect following surgery.

DISCUSSION

Scalp defects develop due to trauma,[5] burns,[6] malignant tumours,[7–9] and radiation therapy[10] for malignant tumours. For patients with scalp abrasion after trauma, replantation of the avulsed scalp has been performed with excellent results,[5] whereas reconstruction of the scalp defect following resection of a malignant tumour is difficult. Dermatofibrosarcoma protuberans is classified as a fibrohistocytic tumour of intermediate-grade malignancy.[4] Dermatofibrosarcoma protuberans is reported to exhibit frequent local recurrence[1–3] but to rarely exhibit metastasis.[4] Simple excision of the tumour was initially performed on this patient, but frequent local recurrence

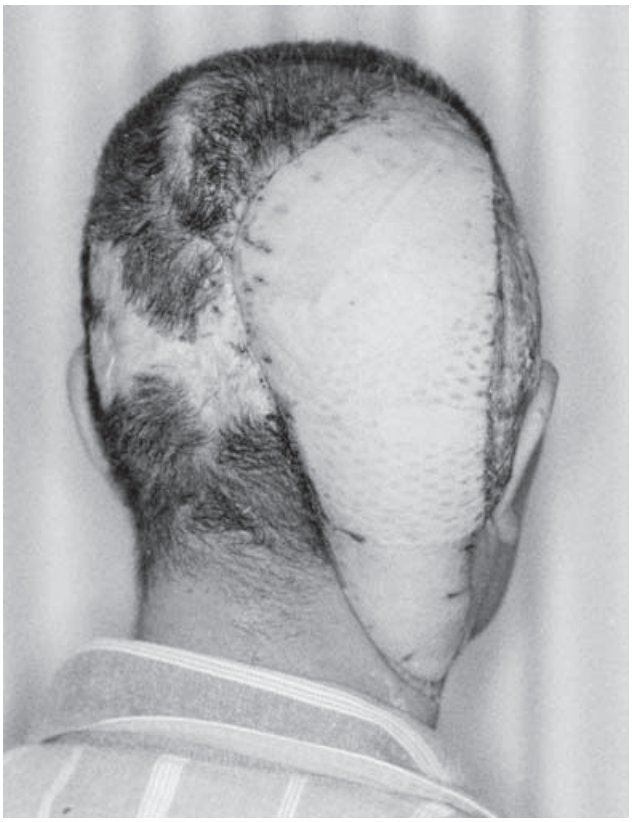

Figure 2 Coverage of the scalp and skull defects was achieved using a free latissimus dorsi myocutaneous flap.

of the tumour subsequently occurred because wide excision had not been performed, thus leading to metastases to the brain and the lung. The patient died 14 years after the initial surgery from metastasis to the abdominal cavity. The case illustrates that simple excision of dermatofibrosarcoma protuberans should not be performed as the initial treatment. Instead, free flap transplantation, which enables wide excision and complete reconstruction, should be selected.

Arnaud et al.[1] noted that it is important to excise a tumour with a 5 cm margin during the initial procedure to improve treatment results of dermatofibrosarcoma protuberans. For patients who develop dermatofibrosarcoma protuberans on the trunk, it is possible to cover the skin defect after wide excision using a free skin graft and a local rotation flap.[1,2] The scalp region features peculiarities such as a limited amount of skin due to inclusion of the face and coverage of the cranium. Because of these peculiarities, the chosen means of reconstruction of the scalp affects treatment results. As free flaps for the treatment of scalp defects associated with malignant tumours, reconstructions by radial forearm flap[11,12] or groin flap[4,13] have been reported. The vascular pedicle for these flaps is not long, however, and the size of an elevated flap is limited, making them inappropriate for reconstruction after a wide excision.

Dermatofibrosarcoma protuberans must be excised along with the skin with a 5 cm margin,[1] thus resulting in an extensive scalp defect. After wide excision of dermatofibrosarcoma protuberans, reconstruction with a latissimus dorsi myocutaneous flap should be the first choice because it permits the harvesting of a wide myocutaneous flap that includes a long and thick vascular pedicle.[7–9,14] At the time of his initial visit to our department, the patient had already developed a metastasis of dermatofibrosarcoma protuberans to the brain, making radical treatment impossible. Free latissimus dorsi myocutaneous flap transplantation was performed as salvation treatment for his skin necrosis, which was resulted from previous surgical failures. As a result, leakage of cerebrospinal fluid and meningitis was prevented, leading to a satisfactory improvement in the quality of life of the patient.

In conclusion, simple excision must not be performed as the initial treatment for dermatofibrosarcoma protuberans of the head. Wide excision of dermatofibrosarcoma protuberans and free flap transplantation, which enables reliable reconstruction of the scalp defect after excision, should be the first choice for treatment.

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