



# Recurrent Dermatofibrosarcoma Protuberans of the Scalp Reconstructed by Visor Flap

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Dermatofibrosarcoma protuberans (DFSP) is a locally aggressive skin tumor. It usually occurs on the trunk and extremities and, only about 10% of cases DFSP are found on head and neck region. Presence of the tumor on the head challenges surgical treatment because of need in wide resections of limited scalp tissue. Recurrent cases of DFSP often possess possibility of bone and intracranial involvement that dictates even greater resection with simultaneous closure to prevent meningitis and other fatal complications. Control of the wound margins either by means of frozen sections or by Moh's technique is a keystone of successful treatment.

Here we present a case of recurrent DFSP of the fronto-parietal area. The case is remarkable for intracranial extension of the tumor. Also we describe a rare use of bipedicle a. temporalis superficialis (visor) flap for reconstruction of composite defects of forehead resulted from wide resection of large recurrent DFSP lesion.

**Keywords:** dermatofibrosarcoma protuberans, surgical flaps, bipedicle a. temporalis superficialis flap, scalp

## Introduction

Dermatofibrosarcoma protuberans (DFSP) is a locally aggressive skin tumor of presumably mesenchymal origin. It is the most common type of skin sarcomas. The tumor usually affects people between 20-60 years; men are slightly more predisposed to it than women. Despite of locally aggressive behavior of classical dermatofibrosarcoma protuberans distant metastasis usually occurs only in 0.5% of cases and it most commonly affects the lungs. [1, 7]

Usual sites of occurrence include trunk and extremities and in only about 10% of cases DFSP occur in head and neck area. [1, 2]

Aberrations in 17<sup>th</sup> and 22<sup>nd</sup> chromosomes and previous local trauma are suggested to contribute to pathogenesis of DFSP. [3]

According to histological findings the tumor can be plaque, nodular, pigmented, fibrosarcomatous or juvenile type. Immunohistochemistry reveals CD34 antigen and

absence of S-100 marker, which is necessary for differentiation from other similar tumors. [1-6]

Due to highly invasive nature DFSP can affect different underlying structures (fat tissue, fascia, muscle, bone, etc.) and cause various correspondent symptoms. However, main invasion occurs horizontally, i.e. in the adjacent skin. Because of tentacle-like centrifugal growth, nests of the tumoral cells can microscopically be found in normally appearing skin as far as 9-15 cm away from the main lesion.

Typically the tumor has irregular borders and composed of firm subcutaneous (sometimes ulcerated) nodules of fleshy or reddish color. Less common presentation of DFSP is morphea-like, atrophic patch, which is prone to further ulceration.

Treatment modalities classically consist of wide resection with 2-4 cm margins of visibly unchanged skin. Chemotherapy with imatinib and radiotherapy can be used as adjuvant measures in selected cases. [1-7, 9,

11]

Although, DFSP rarely affects the scalp the localization itself challenges surgical treatment. This is particularly true in case of recurrent lesions, which invade bone and intracranial structures. Presence of intracranial extension requires more aggressive resection, including bone, dura mater and even brain structures apart from skin.

According to review of available literature several options of reconstruction can be exploited after classical wide excision or Moh's chemosurgery, including; a) wound grafting, b) reconstruction with local scalp flap(s), c) usage of tissue expanders prior to the main surgery, d) reconstruction with free flap, e) usage of tissue expanders, after the main surgery. [2, 4, 6, 11]

In case of intracranial involvement with invasion into dura and more deep structures resection must be followed by simultaneous reconstruction combined with one of the method of soft tissue coverage. This is necessary to prevent liquor leakage as it may have fatal consequences.

Here we present a case of recurrent DFSP of the frontal region with intracranial extension, which has been treated successfully by wide excision (including bone and intracranial structures), cranioplasty and transposition of bipedicle a. temporalis superficialis (visor) flap.

### Case report

A 36 years old female applied to neurosurgeon because of tumoral growth on the head, at locus of previous operation. Her disease began 7 years before a small nodule on frontal area of the scalp. It was excised at provincial healthcare unit under the local anesthesia without further pathologic examination. The tumor reoccurred next year. Totally, before current application she was operated 6 times, because of almost annual recurrence of the neoplasm. Pathological diagnosis of the tumor was established to be dermatofibrosarcoma protuberance.

In a last few months she had developed a headache, dizziness, general malaise and nausea. She was very anxious and depressed concerning course of her disease. Other findings of routine physical examination were in the normal range.

Local examination revealed scarred hairless oval-shaped area measured approximately 7x8 cm occupying center of fronto-

parietal area. Its borders were marked by postoperative scars, whereas the center contained crust covered nodularities and left lower quadrant was occupied by typical pinkish hard subcutaneous tumoral nodule. (Fig. 1)

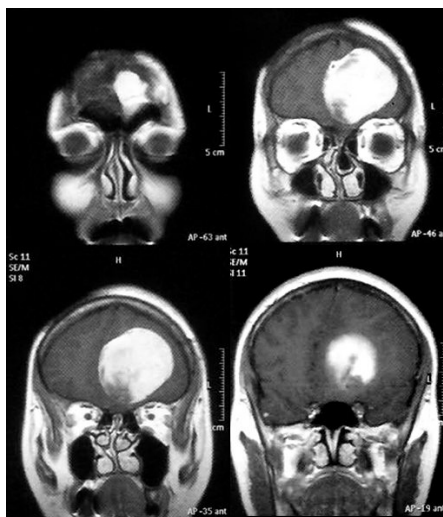
Magnetic resonance imaging of the brain revealed well-defined, 60x52 mm sized lesion, which continued from subcutaneous level to the extra-axial space through postoperative bone defect in the left half of frontal bone. The lesion homogeneously absorbed contrast agent being hypointense on T1-weighted and slightly hyperintense on T2-weighted images. Right-sided dislocation of the anterior median structures up to 8-10 mm, as well as diffuse edema of the left hemisphere was also noted. (Fig. 2)

After explanation of the details of treatment and taking written consent, the patient undergone operation. Because of nature of the post-resection defect, reconstruction initially included using of free radial forearm flap along with transposition of local scalp. However, the patient insisted on procedure with no risk of total flap loss. So, our final surgical plan included wide excision of the tumor within 3 cm of the healthy skin, frozen section biopsies from margins of the skin wound, en bloc resection of tumor bearing scalp along with underlying portions of frontal and parietal bones, resection of the intracranial part of the tumor with surrounding dura, closure of the dural defect by graft of fascia lata, closure of the bony defect by acrylic allotransplant and closure of the skin defect by bipedicle a. temporalis superficialis (visor) flap with subsequent skin grafting of the donor area (Fig. 3 a-e).

The operation proceeded according to plan described above; definitive closure began after 8 frozen biopsies taken from the edges of skin wound (2 pieces from each quadrant) had been reported to be tumor-negative.

The postoperative course was free of major complications. The patient was signed out at 10th postoperative day. As of the last control, which was 6 month after the last operation, she was satisfied with results of treatment and showed improved mood. No signs of recurrence were found. Deformities in form of "dog ears" at the edges of visor flap resolved simultaneously after 5th postoperative month.

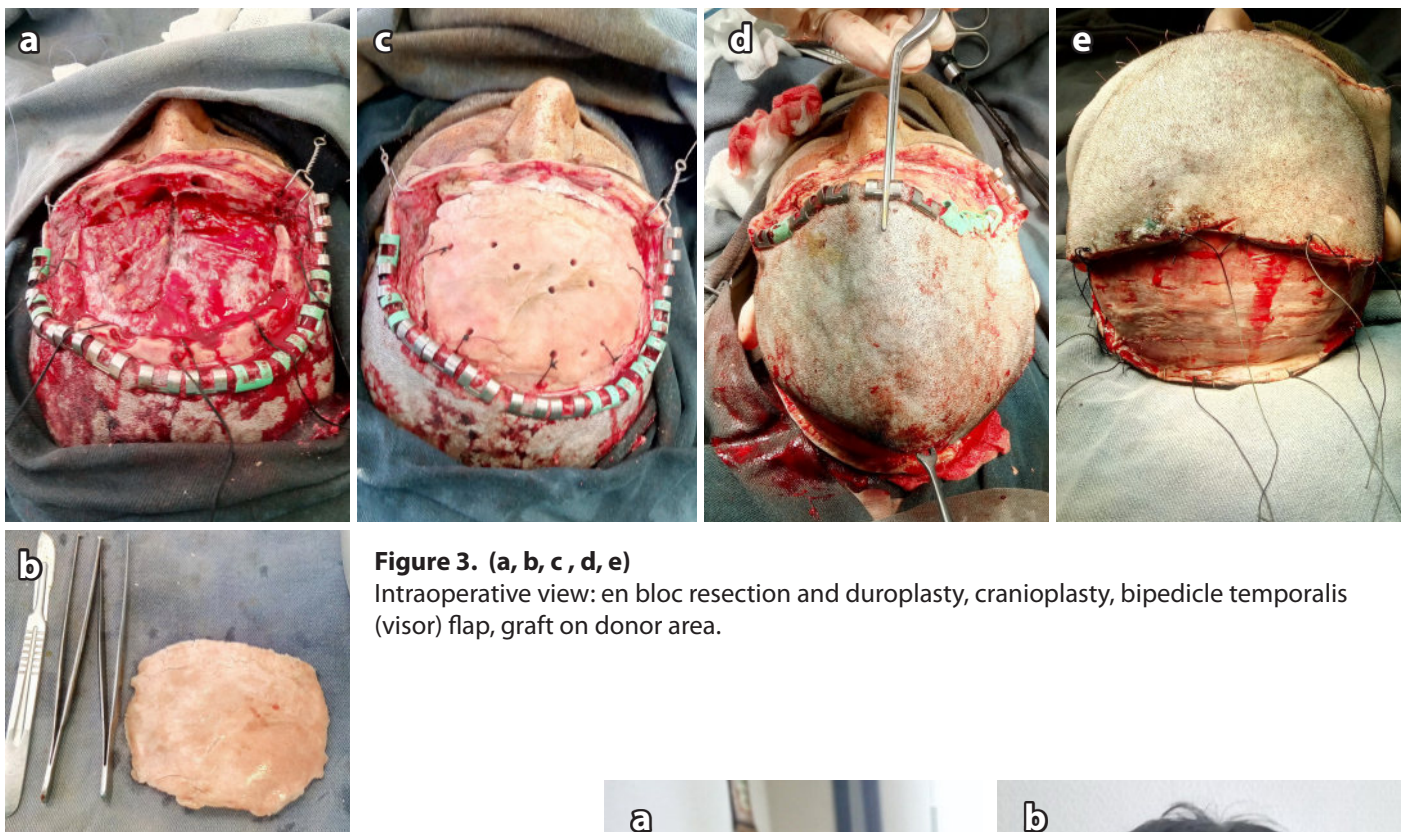
Surprisingly, the patient is not bothered by hair on the forehead region. She prefers carrying head covering veils and refuses further reconstruction to achieve hairless skin or to use any oth-



**Figure 1. (left)**  
Pre-operative appearance of the tumor.

**Figure 2. (right)**  
Pre-operative MR imaging (T2-weighted, coronal) shows intracranial extension of the tumor.





**Figure 3. (a, b, c, d, e)**

Intraoperative view: en bloc resection and duroplasty, cranioplasty, bipedicle temporalis (visor) flap, graft on donor area.



**Figure 4. (a, b)**

Post-operative view of the frontal area and donor site of the visor flap.

er method of hair removal. (Fig. 4 a-b)

### Discussion

Dermatofibrosarcoma protuberans (DFSP) is a rare skin tumor with locally aggressive behavior. Spread of tumoral cells far from the primary lesion is commonly known histological and clinical feature of the tumor. Therefore wide excision is keystone of the successful treatment. [1, 3-5]

Although DFSP of the head and neck constitutes less than 10% of cases, treatment of such entity is challenged by some anatomic and physiologic features. First, only limited amount of hair bearing scalp is available, which leads to an aesthetic problem in case of wide excision and skin grafting. However, this issue can be addressed by preoperative scalp expansion in case of small, non-

recurring and slowly growing lesions. Otherwise, postoperative scalp expansion may also be an option. Second, wide excision may exceed borders of scalp, requiring excision part of the ear, forehead or brow. In these cases additional aesthetic problems are to be considered. Third, cases where lesions invaded cranial bones and intracranial space require immediate reconstruction of multiple tissue layers to prevent meningitis and similar fatal complications. [8] The dura is classically repaired by fascil graft from fascia lata, temporal fascia etc. Alloplastic materials as well as biological glues may also be used for this purpose. [1] Bone grafts (from iliac crest or ribs), alloplastic materials (acrylic derivatives) or even autologous bone processed by autoclaving or irradiation can be employed to reconstruct defects of cranial

vault. Alternatively, the defect can be left open. Skin closure over cranial defect (either open or reconstructed) must be in the form of flap. [2, 4] Small to average wounds can be closed by transposition of the rest of the scalp with primary closure or grafting of the donor site. However, repair of large scalp defects, which exceeding 9 cm in diameter, often require free tissue transfer. Here muscle flap with overlying split thickness skin graft remains classical option, whereas fasciocutaneous flaps (like anterolateral thigh flap, transverse thoracodorsal artery flap etc.) can also be used in suitable cases.

In our case, we used classical dural repair by graft of fascia lata. Taking into a consideration large size of cranial defect (occupying large part of parietal and almost all frontal area) we decided on alloplastic bone replacement with methylmethacrylate material. Reconstruction of soft tissue coverage was done by large bipedicle a. temporalis flap (visor flap). The choice of this flap was directed by need in reconstruction that possesses low morbidity and no risk of total flap failure. Visor flap is widely known for its application in reconstruction of hear-bearing segments of upper and lower lips, and cheek in male patients, particularly after burn injuries. It provides ample amount of thick scalp skin and have robust circulation through both a. temporalis and depending on design can also include branches of retroauricular arteries. Here, we used the flap to cover almost entire parieto-frontal area over alloplastic material. This is, for our knowledge is a rare, "off label" use of visor scalp flap.

The drawbacks of our operation include translocation of hear-bearing skin on the forehead and creation of alopecia at the donor site of visor flap. Our patient refused further reconstruction. However, one can speculate on that further refinement can be done by laser hair removal, resurfacing of the forehead area by non-hear-bearing skin graft, reposition of the visor flap back to donor area and transfer of a free fasciocutaneous flap onto forehead area. Alopecia on the donor site can also be addressed by usage of tissue expanders.

By presenting this case we would like to raise awareness of DSFP in case of nodular lesions of scalp. Wide excision with subsequent control (either by frozen sections or by Moh's method) is sine qua non of successful treatment, especially in case of recurrent lesions. It has to be emphasized that recurrent DFSP of scalp has potential for intracranial extension, which necessitates even larger resection. According to our experience, visor scalp flap is a valuable option for reconstruction of composite defects resulted from such resections.

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