Definitive cranioplasty and scalp reconstruction in recurrent dermato fibrosarcoma protuberance of scalp

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) of the scalp is a relatively uncommon soft-tissue neoplasm. Calvarial involvement can lead to a quick intracranial spread making the tumor incurable. The management of DFSP scalp, especially recurrent lesions with calvarial involvement should be a team approach with close cooperation between the neurosurgeon and the reconstructive surgeon. The failure to excise the underlying periosteum is one of the major reasons for early calvarial spread. At no stage adequate excision, i.e., 5 cm margin and removal of underlying periosteum should be compromised. In developing world where follow-up of patients is difficult and specialized health services are at a premium we believe that a wide surgical excision, including the underlying periosteum and cranioplasty followed by local flap cover if possible will provide the best results with the shortest downtime.

Key words: Cranioplasty, dermatofibrosarcoma, reconstruction, scalp

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) of the scalp is a relatively uncommon soft-tissue neoplasm constituting less than 1% of all DFSP.[1] Locally aggressive nature of lesion, potential intracranial spread and lack of adequate scalp tissue for reconstruction, makes this lesion unique and its proper management challenging. In cases having calvarial involvement reconstruction becomes difficult due to the composite tissue loss. All these features mandate that the management of DFSP scalp, especially recurrent lesions with calvarial involvement should be a team approach with close cooperation between the neurosurgeon and the reconstructive surgeon.[1]

CASE REPORT

Two patients both 35-year-old males, presented with recurrent 4 cm × 4 cm (case 1) and 8 cm × 8 cm (case 2) ulcero-proliferative lesions over scalp vertex. They had undergone excision of the lesion twice in last 3 years followed by primary closure in the case 2 and excision followed by split skin graft (SSG) in the case 1. Both developed recurrence of lesion within 3 months of each surgery. Biopsy performed during the surgery was consistent with DFSP scalp. The tumors underwent extensive ulceration 3 months before reporting to us.

On examination, the lesions were painless, hard and nodular, which bled on touch [Figure 1]. Ulcers were found fixed to the underlying bone with presence of granulation tissue and wound discharge. No significant cervical lymphadenopathy was observed. Central nervous system examination was normal. Ultrasonography abdomen and chest X-ray were done to rule out likely areas of secondaries.

Computed tomography (CT) showed an extensive soft-tissue lesion involving all the scalp layers over the scalp vertex with involvement of the underlying bone. Lesion also involved the previous SSG in case 1. Both outer and inner table of calvarium were involved with intact dura [Figure 2].

Wide excision of the lesion, including the previous SSG was planned keeping a margin of 5 cm. Full thickness excision of the involved bone was done with 3 cm margin.

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Dura continuity was preserved and was found to be grossly free from the tumor [Figure 3].

The bone defect was covered with porous polyethylene implant and secured with 1.5 mm plates in the second case [Figure 4]. The large skin defect was then covered with posterolaterally based scalp flap after ensuring complete hemostasis in both patients. Biopsy specimen showed free tumor margins, consistent with the diagnosis of DFSP scalp. Patient was put on follow-up and observation. There has been no recurrence of lesion for last 1 year [Figure 5].

DISCUSSION

Hoffman named this tumor DFSP in 1924 with trunk being the most common site.[1-3] Less than 70 cases of scalp DFSP have been reported in the literature.[1,4,5] The most widely used is World Health Organization classification that classifies DFSP as intermediate tumor of fibrohistiocytic origin.[3] It has variable malignant potential and is considered to be locally invasive with low incidence of lymphatic or hematogenous spread.[6,7] However, there is pronounced and quick recurrence in cases of inadequate excision.[1,5] This tendency coupled with very good prognosis following adequate excision makes the management of this lesion debatable, especially in scalp DFSP where there is a paucity of good reconstructive tissue for scalp cover.[1] The relatively infrequent occurrence of DFSP on scalp lessens its clinical awareness and diagnosis is often made on histology.[1]

Most of these patients are initially treated by a neuro-surgeon with limited reconstructive capabilities. DFSP is a curable locally aggressive lesion. However, in scalp, calvarial involvement can lead to a quick intracerebral spread, making this easily manageable condition into an inoperable one. The necessity for primary scalp closure in such cases is what leads to inadequate excision and recurrence. No correlation between tumor size and lateral excision margin has been established; hence all lesions of DFSP scalp, irrespective of their size should undergo a wide excision (at least 5 cm)

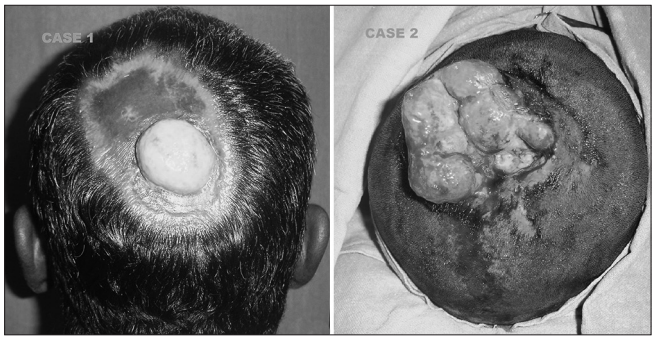

Figure 1: Recurrent ulceroproliferative lesions over scalp-case 1 and 2

Figure 2: Computed tomography scan showing full thickness bone involvement-case 1

Figure 3: Wide excision of lesion including soft-tissue, periosteum and bone-case 1

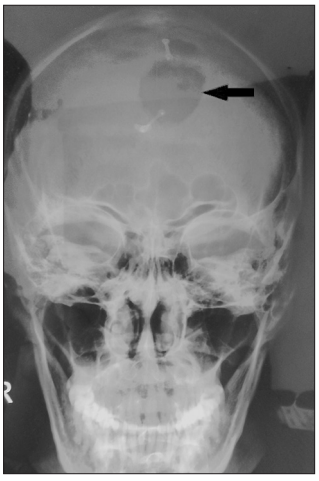

Figure 4: Post-operative X-ray showing radiolucent implant secured with plates-case 2 (Arrow)

Figure 5: Late post-operative showing well-settled scalp flap after excision and definitive cranioplasty-case 2

along with excision of the underlying periosteum to prevent calvarial involvement. If the bone is involved at least 3 cm margin is essential.[1]

When wide excision of the scalp and bone is done, a composite scalp reconstruction becomes a necessity. Scalp has limited tissue for reconstruction with a thin calvarial bone. Good reconstruction of scalp requires a thick, pliable hair bearing skin cover with bone cover.[1] In the management of DFSP of scalp, team approach is a must with effective coordination between the excising and the reconstructive surgeon from the outset.

In most of the primary cases the diagnosis is made on histology, every effort should be made by the treating surgeon to go back to the patient's records and see firstly whether 5 cm excision margin was taken or not and secondly if the underlying periosteum was excised.[1] This should be done even in cases where tumor margins are said to be free on histopathology. The failure to excise the underlying periosteum is one of the major reasons for early calvarial spread. In all patients with sub optimal resection, an option of re-excision with 5 cm margin, including the underlying periosteum with scalp flap reconstruction should be given to each patient.[1]

If the cranium is involved and/or intracranial extension is detected, a wide excision of the affected cranium with at least 3 cm margins should be done. In cases of wide bony defects, it becomes impossible to primarily close the bone defect as occurred in our case. In these situations, an auto-graft or implant is required for reconstruction.

Of all the techniques for calvarial reconstruction, the use of autologous split calvarial bone graft is considered as the best procedure as it entails the use of native bone and eliminates the use of foreign body; thus, reducing the rate of infection and implant extrusion. In patients with neoplastic diseases it is difficult to harvest split calvarial graft and will require neurosurgical expertise, which is not readily available. Use of a rib or iliac crest bone grafts leads to additional procedures, increasing patient morbidity and carries the risk of unpredictable results due to long-term bone re-absorption.

In the current era, commonly used alloplasts include silicone prostheses, titanium implants and a variety of artificial bones.[8] Porous polyethylene implant is one of the most frequently used. It is a highly stable and slightly flexible porous alloplast that has been shown to exhibit rapid tissue and bone in-growth. The implant is easy to shape, yet strong enough for use in the craniofacial skeleton. As a result, it has been used to repair cranial defects, facial deformities and in auricle reconstruction.[8] Disadvantage of the polyethylene implant is its radiolucency on conventional CT and magnetic resonance. It provides a stable, esthetic, permanent calvarial replacement without any additional procedures is easy to perform and does not require extensive instrumentation.

Keeping these factors in mind a detailed discussion was done with patients and all possible outcomes explained. While the first patient refused any bone reconstruction, the second patient decided on implant reconstruction.

Scalp reconstruction can be performed using the skin graft and flaps (local flaps or free flaps). Local scalp flaps when possible are most suited for the reconstruction since they have similar characteristic features with the defect area. However, the relative inelasticity of scalp skin limits reconstruction without skin grafting of donor site.

Taniguchi presented a case of scalp DFSP with brain metastasis.[2,6] They performed a wide excision and reconstruction with a latissimus dorsi myocutaneous free flap. They concluded that a simple excision should not be performed as the initial treatment for DFSP scalp to which we agree. They however promoted a free flap reconstruction in scalp DFSP in all cases. This requires technical expertise, increased operative time, intensive post-operative care and free tissue transfer generally results in a hairless reconstruction.

Wide excision with underlying periosteum followed by cranioplasty and flap reconstruction is the most suitable treatment approach.[3] This, we feel will help in reducing the incidence of recurrence to a large extent as well as provide best possible esthetic and functional result.

In extensive tumors with in adequate local tissue for

cover, microvascular flap becomes the best option.[2] In lesions with extensive calvarial involvement, cranioplasty can be deferred for a later stage if a good skin flap is being given as in our first case. However, at no stage adequate excision, i.e., 5 cm margin and underlying periosteum should be compromised.

It is a slow growing, locally aggressive fibrous tumor with a pronounced tendency for local recurrence, rarely metastasizing to regional lymph node or distant sites. Clinicians are urged to be more aware of this condition and obtain a histologic diagnosis by core or incision biopsy prior to excision. In all confirmed cases of DFSP of scalp, it is imperative to involve a reconstructive surgeon. In the developing world where follow-up of patients is difficult and specialized health services are at a premium we believe that a wide surgical excision, including the underlying periosteum and cranioplasty followed by local flap cover if possible will give the best results possible with the shortest downtime. Specialized treatment protocols such as mohs micrographic surgery, immunotherapy and radiotherapy have a role, especially in inoperable lesions, patient with multiple secondaries and distant metastasis or who are unfit for surgical excision.[9]

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