Management of Recurrent Dermatofibro Sarcoma Protuberance of Scalp-a Reconstructive Challenge

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Introduction

Dermato fibrosarcoma protuberance (DFSP) of scalp is a relatively uncommon soft tissue neoplasm constituting less than one percent of all DFSP [4]. 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. All these features mandate that the management of DFSP scalp, especially extensive and recurrent lesions should be a team approach with close co-operation between the neuro-surgeon and the reconstructive surgeon.

Case Report

35 year old male, presented to us with a recurrent 8×8 cm ulcero-proliferative lesion over the vertex of the scalp. Patient was apparently asymptomatic 3 years back when he noticed a small painless nodular lesion over the vertex of the scalp. He underwent excision of the lesion twice in last 3 years followed by primary closure. He developed recurrence of lesion within 3 months of each excision.

Biopsy was not done during the first excision while during the second procedure it was consistent with DFSP scalp.

For last 1 year patient did not take any treatment and the lesion continued to increase in size. The swelling underwent extensive ulceration 3 months before reporting to us.

On examination the lesion was painless, hard, and nodular which bled on touch. There was presence of dirty granulation tissue over the ulcer with wound discharge. Margins were irregular and ulcer was found fixed to the underlying bone. In view of the aggressive ulcero proliferative picture a differential diagnosis of squamous cell carcinoma was kept (Fig. 1). A repeat incisional biopsy was done which again confirmed the diagnosis of DFSP scalp. No significant cervical lymphadenopathy was observed. Similar lesions were not seen in any other part of body. Systemic examination including CNS examination was normal with no evidence of raised intracranial tension or focal neurological deficit. Complete metastatic workup and routine investigations were performed. USG abdomen and Chest X-ray were done to rule out likely areas of secondaries and were found to be normal.

CT showed an extensive soft tissue lesion involving all the scalp layers over the vertex of the scalp with involvement of the underlying bone. Both outer and inner table were involved. However the area of bone involvement was much lesser than the soft tissue involvement. The intracranial involvement was extradural with no evidence of either dura breach or cerebral involvement. There was no evidence of any raised intra cranial tension (Fig. 2).

A wide excision of the lesion was planned keeping a margin of 5 cm (Fig. 3). All the old scar lines from previous surgery were also excised. The area of bone involvement was much lesser and full thickness piecemeal excision of the involved bone was done with 3 cm margin. Dura continuity was preserved and was found to be grossly free from the tumour (Fig. 4).

In view of extensive soft tissue lesion with intracranial involvement and multiple recurrences, definitive cranioplasty was deferred. The large skin defect was then covered with postero-laterally based scalp flap after ensuring complete

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Fig. 1 Extensive ulceroproliferative scalp lesion

haemostasis. Biopsy specimen showed free tumour 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 6 months (Fig. 5).

Discussion

D'Andree et al. summarized the historical backgrounds of this entity in their study and according to their report Taylor

Fig. 3 Bone defect with extensive removal of periosteum

first described a potentially relapsing neoplasia similar to a keloid. Clinical classification was done by Darier and Ferrand in 1924 and Hoffman named this tumour DFSP in 1924 [1, 2]. Bednar tumor is a rare pigmented variant of DFSP. Familial and congenital DFSP are also described in the literature. The most common location for this lesion is the trunk with vulva, penis, toe, breast, jejunum and scalp being the other rare reported sites. Less than 70 cases of scalp DFSP have been reported in the literature [4].

The tumour has low malignant potential and is considered to be locally invasive with low incidence of lymphatic or haematogenous spread. However there is pronounced and quick recurrence in cases of inadequate excision [4]. This tendency coupled with very good prognosis following adequate excision makes the management of this lesion

Fig. 2 CT showing full thickness calvarial spread with intact dura

Fig. 4 Excised specimen

debateable and interesting especially in scalp DFSP where there is paucity of good reconstructive tissue for scalp cover. Today various modalities of treatment other than wide surgical excision like immunotherapy, moh's micrography and radiotherapy are being advocated in various subset of patients [7].

However scalp lesions and their management provide some unique dilemmas.

Firstly, it has been our experience that majority of extra calvarial primary scalp lesions of any nature first present either to a Neurosurgeon, dermatologist or a general surgeon. These scalp lesions include, benign tumors like lipoma, peripheral nerve sheath tumours and malignant tumours like malignant fibrous histiocytoma, liposarcoma, leiomyosarcoma, and epithelioid sarcoma and metastasis. Of all these DFSP scalp is a rare tumour and frequently identified on histopathology only. In our extensive search of literature we have not found even a single case where a new scalp lesion was excised keeping in mind the diagnosis of DFSP. The relatively infrequent occurrence of DFSP on scalp lessens its clinical awareness and diagnosis is often made on histology.

Majority of the DFSP scalp on their initial presentation mimic a benign tumour. They are small, soft, non-tender and free from the overlying skin and underlying bone. Mostly MRI/CT or FNAC/Incisional biopsy is not performed for these lesions. Generally these are excised under local anaesthesia on a day care basis. Only on histopathology is the diagnosis confirmed. Many a times in primary or rural setup even biopsy is not done, as in our case. Quick and aggressive recurrence is the only thing which attracts the clinician's attention towards the problem.

Most of these patients are initially treated by a surgeon with limited reconstructive capabilities. There is tendency to excise the scalp lesions conservatively, keeping in mind primary closure of the scalp defect. In cases of DFSP scalp, a lateral margin of 3 cm results in a local recurrence rate of 10–20 % (McPeak et al., 1967; Roses et al., 1986; Gloster et al., 1996) and at 4 cm a recurrence rate of 6 % was found in 96 patients (Petoine et al., 1985) [7]. There was no

correlation between tumour size and lateral excision margin; hence all lesions of DFSP scalp irrespective of their size should undergo a wide excision [7]. The necessity for primary scalp closure on the scalp is what leads to inadequate excision and recurrence.

Secondly scalp as an anatomical region is unique, as it has limited tissue for reconstruction with a thin calvarial cover. Good reconstruction of scalp requires a thick, pliable hair bearing skin cover. Bone cover should be achieved using thin vascular, membranous bone. Thus the ideal scalp replacement should be durable, of the same thickness as the remaining scalp, hair-bearing, and reliable enough to allow for timely post-operative adjuvant therapy. Moreover calvarial involvement can lead to a quick intra cerebral spread of this lesion, making this easily manageable condition into an inoperable one.

Scalp reconstruction can be done using skin graft and flaps (Local Flaps or Free flaps). Free tissue transfer for reconstruction of large scalp defects has become popular. Taniguchi [5] presented a case of scalp DFSP with brain metastasis. 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 postoperative care and free tissue transfer generally results in a hairless reconstruction.

Skin grafts have major disadvantages such as missing hair and are prone to ulceration.

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.

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. This followed by cranioplasty and reconstruction with flap cover is the most suitable treatment approach [3].

Fig. 5 Well healed scalp flap

Hence we advocate that, in the management of DFSP of scalp, team approach is a must with effective co-ordination between the excising and the reconstructive surgeon from the outset.

Since 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 reports and see firstly whether 5 cm excision margin was taken or not and secondly if the underlying periosteum was excised. This should be done even in cases where tumour margins are said to be free on histopathology. The failure to excise the underlying periosteum is what we feel 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. This, we feel will help in reducing the incidence of recurrence to a large extent, as well as provide best possible aesthetic and functional result. All patients not opting for re excision should be advised a regular follow-up to detect recurrence if any as early as possible.

Only in such cases, where the adequacy of excision is doubtful or expertise for flap reconstruction is not available simple skin graft can be done. The advantage being that any recurrence can be detected early with skin grafts. However acceptance of skin grafts over bare calvarial bone can be uncertain, moreover skin grafts require very good care by the patient as they are insensate. Skin grafts are also prone to repeated ulceration even after minor trauma [6]. Moreover if definitive calvarial reconstruction is being contemplated in extensive lesions, then a flap either myocutaneous or fasciocutaneous is a must. In cases, where the tumour is extensive with inadequate local tissue for cover, a microvascular flap becomes the best option available. In cases of recurrent intra cerebral lesion, or lesions with extensive calvarial involvement, if a good skin flap is being given then calvarial reconstruction can be deferred for a later stage as in our case.

However at no stage adequate excision i.e. 5 cm margin and underlying periosteum should be compromised.

In India 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 local flap cover if possible will give the best results possible. Specialized treatment protocols like moms micrography and immunotherapy and radiotherapy have a role, especially in inoperable lesions, patient with multiple secondaries and distant metastasis or who are not fit for surgical excision.

Conclusion

It is a slow growing, locally aggressive fibrous tumour with a pronounced tendency for local recurrence, rarely metastasizing to regional lymph node or distant sites. Most lesions occur over the trunk or proximal extremities. In all confirmed cases of DFSP of scalp it is imperative to involve a reconstructive surgeon and use all methods of reconstruction after doing an aggressive excision of the tumour which involves a 5 cm margin and removal of the underlying periosteum. We feel that, with the involvement of a reconstructive surgeon at an early stage local scalp flaps can be used to provide a good hair bearing sensate flap cover with grossly reduced recurrence rates.

To conclude, we describe a relatively uncommon cutaneous tumour, in an uncommon site, the scalp. Awareness of this tumour at this site is important. Clinicians are urged to be more aware of this condition and obtain a histologic diagnosis by core or incision biopsy prior to excision. Given the propensity of recurrence and metastasis, a wide optimal surgical margin including the underlying periosteum should be ensured during excision. Moreover early re excision in case of inadequate excision and timely involvement of a reconstructive surgeon provides the best possible aesthetic outcome with low incidence of recurrences.

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