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Case Report

Recurrence case of rare scalp dermatofibrosarcoma protuberans: Two case reports of a wide radical excision, craniectomy bone involvement followed by cranioplasty and reconstruction

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Abstract

Background: Dermatofibrosarcoma protuberans (DFSP) is a rare low-grade sarcoma of the fibroblast originating from the dermal layer of the skin, characterized by a locally aggressive growth and high rate of local recurrence.

Case Description: Two patients underwent a wide radical excision of recurrent scalp DFSP which was reconstructed with translational skin flap and split-thickness skin graft. We described above cases several years ago with a local excision of the tumor; recently, they developed local recurrence of DFSP with calvarial involvement. We then performed a wide radical excision, with craniectomy of the cranial defect followed by cranioplasty using titanium mesh, continuing with reconstruction.

Conclusion: A successful treatment and management depends on achieving local control and preventing cosmetic and functional deficit; all efforts should be made for complete excision. Postoperative follow-up recommended for highly suspicious cases and annual checkups should be performed up to 5 years after definitive therapy.

Key Words: A wide radical excision, craniectomy and cranioplasty, recurrence of dermatofibrosarcoma protuberans, translational flap and split-thickness skin graft

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a neoplasm originating from the fibroblast with characteristic aggressive mesenchymal tumor; its local recurrence rate is up to 60%.[6] DFSP infiltrates their surrounding as pseudopods, resulting in incomplete total removal with a potential to recur in the future. A single-center series of 90 patients in South Korea suggested that a wide local excision with adequate lateral and deep margin can effectively control local recurrent rate for DFSP; recurrence-free survival was 87% at 6 years and 77% at 7 years.[4] Scalp DFSP is a relatively rare case constituting less than 1% of all DFSP.[3]

This article reported our recurrent case of scalp DFSP, reviews this locally aggressive growth tumor with high rates of local recurrence, surgical technique, and its management, as well as with follow-up recommendations for this unusual neoplasm.

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CASE REPORTS

Case 1

We present a case of a 29-year-old adult male whose main complaint was a recurrent lump, a painless lesion, on his mid frontal region after tumor excision who was diagnosed with DFSP 4 years ago [Figure 1a and b].[2] History revealed that the DFSP recurred in the second year after the first tumor excision [Figure 1c]. On physical examination, there was a irregular, skin-colored, multiple lobulated mass, measuring 12 × 10 × 5 cm in diameter, with a fixated base; head computed tomography (CT) shown no intracranial mass [Figure 1d].

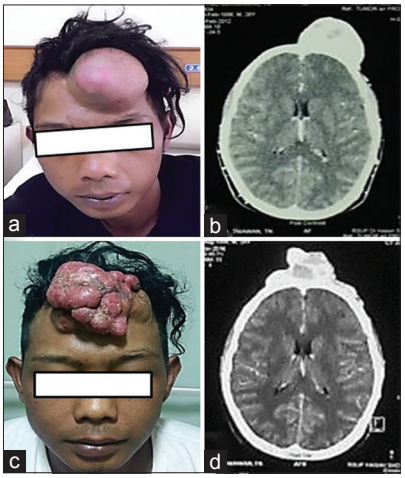

Figure 1: (a) Lump in mid frontal region in male patient (before excision in 2012), mass size 12 × 10 × 6 cm. (b) Head CT scan showing soft tissue attenuation irregular mass single lesion lobulated in mid frontal region. (c) Lump in mid frontal region in same patient after operated 4 years ago with mass size 12 × 10 × 5 cm. (d) Head CT scan showing soft tissue attenuation irregular mass multiple lesion lobulated in mid frontal

Under general anesthesia, the patient was positioned supine with his back elevated approximately 20 degrees. A wide radical excision with 4 cm distance from neoplasm margin and translational skin flap line was drawn [Figure 2a-c]. After a wide radical excision was performed, calvarial defect was observed without involvement of duramater; frontal bone craniectomy was performed [Figure 2d]. A wide bone defect 10 × 8cm was made, and cranioplasty using titanium mesh was placed [Figure 2e]. The skin defect was then reconstructed by translational skin flap and split-thickness skin graft [Figure 2f and g]. An irregular, skin colored, multiple lobulated mass, measuring 12 × 10 × 5 cm in diameter, was excised along the surrounding skin in radius 4 cm from neoplasm edge [Figure 2h].[4]

As in histopathological findings, using hematoxylin and eosin (HE) stained sections showed a densely cellular and poorly circumscribed tumor in the dermis layer, comprising interwoven bundles and fascicles of uniform spindle shaped cells arranged in a "storiform" or "cartwheel" pattern [Figure 4a]. The tumor cells had monotonous appearance with oval nuclei, vesicular chromatin, inconspicuous nucleoli, and scanty-to-moderate cytoplasm. A panel of immunohistochemistry (IHC) comprising CD-34 and vimentin was applied. Tumor cells were strongly positive for both CD-34 [Figure 4b] and vimentin expression [Figure 4c].

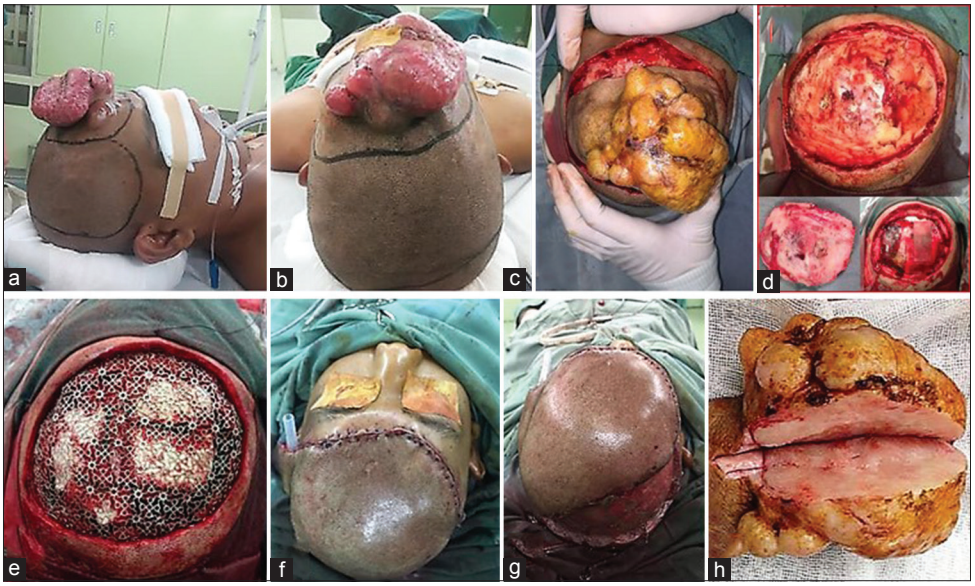

Figure 2: (a-c) A wide radical excision with 4 cm distance from neoplasm margin and translational skin flap line was drawn. (d) After wide excision was performed, calvarial defect was observed without involvement of duramater below; as frontal bone craniectomy was performed. (e) A bone defect 10 × 8cm was made and cranioplasty using titanium mesh was placed. (f) The skin defect was then reconstructed by translational skin flap and (g) split-thickness skin graft. (h) Mass with an irregular, skin colored, multiple lobulated mass, 12 × 10 × 5 cm in diameter was excised

Case 2

A 49-year-old male presented with the main complaint of a recurrent lump, a painless lesion, on his mid parietooccipital region after tumor excision; the patient was diagnosed with DFSP 3 years ago. History revealed that the recurrence of DFSP occurred in the second year after the first tumor excision. On physical examination, there was an irregular, skin-colored, multiple lobulated mass, measuring 8 × 6 × 4 cm in diameter, with a fixated base; head CT showed no intracranial mass [Figure 3a].

Under general anesthesia, the patient was positioned. A wide radical excision with 4 cm distance from neoplasm margin and translational skin flap line was drawn [Figure 3b]. After a wide excision was performed, calvarial defect was observed without involvement of duramater; parietooccipital bone craniectomy was performed [Figure 3c]. The skin defect was then reconstructed using translational skin flap and split-thickness skin graft [Figure 3d and e]. An irregular, skin-colored, multiple lobulated mass, measuring 8 × 6 × 4 cm in diameter, was excised along surrounding skin in radius 4 cm from neoplasm edge.[4] HEstained sections showed a densely cellular and poorly circumscribed tumor in the dermis layer comprising interwoven bundles and fascicles of uniform spindle shaped cells arranged in a "cartwheel" pattern. It had monotonous appearance with oval nuclei, vesicular chromatin, inconspicuous nucleoli, and scanty-to-moderate cytoplasm.

DISCUSSION

The management of scalp DFSP, immunohistochemical demonstration of CD34, and vimentin are important features for diagnosing DFSP; in which both proteins showed strong positivity were noted in our cases. Recurrent lesions, masses measuring more than 5 cm, and those with calvarial involvement need team effort from both neurosurgeon and plastic surgeon for reconstruction. One study observed recurrence rates after wide local excision performed during 2000–2012.[1] The pooled recurrence rate was 8.5% in 1432 patients; wider excision resulted in a lower recurrence rate. Another study showed that wide local excision <3 cm resection margin resulted in an increased recurrence rate.[7] After a wide local excision of the lateral margins were decided based on location of DFSP (head and neck area or not), the size of the tumor (≤ or ≥5 cm), recurrence status,[4] and bone craniectomy was done, a composite scalp reconstruction become necessity; since neurosurgeon have limited reconstruction capabilities, team effort and coordination with the reconstructive surgeon become crucial. In our cases, because there were calvarial involvement, a wide excision of cranium with a least 3 cm margins were performed; both outer and inner table of calvarium were involved with an intact duramater. After we removed the cranial bone, we found that duramater continuity was preserved and was found to be grossly free from the DFSP. We then used titanium mesh or implants to close the bone defect as artificial bones.

Scalp reconstruction was then performed using translational local skin flap and split-thickness skin grafts. Local skin flaps in our cases are the most suited for reconstruction because they have similar characteristic feature with the defect area. Other options are free flap reconstruction that require technical expertise, increased operative time, and intensive postoperative care; most of free tissue transfers commonly resulted in hairless reconstructive area. Wide excision followed by cranioplasty and flap reconstruction provide the best possible esthetic and functional results.

Clinically, DFSP initially presents typically superficial in location as a cutaneous pink to red-bluish painless

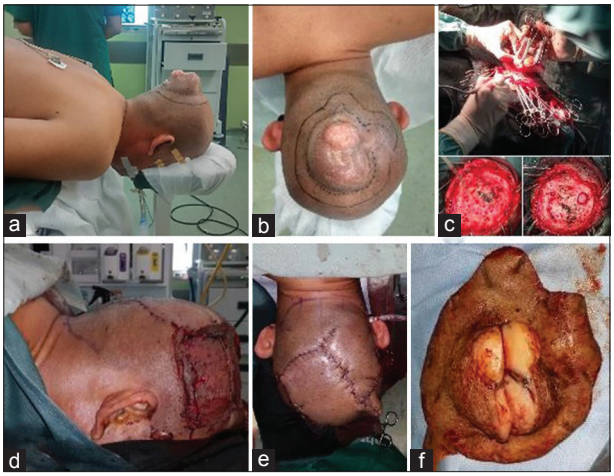

Figure 3: (a, b) A wide radical excision with 4 cm distance from neoplasm margin and translational skin flap line was drawn. (c) After wide excision were performed, calvarial defect was observed without involvement of duramater below; as frontal bone craniectomy was performed. (d, e) The skin defect was then reconstructed by translational skin flap and (f). Mass with an irregular, skin colored, multiple lobulated mass, 8 × 6 × 4 cm in diameter was excised

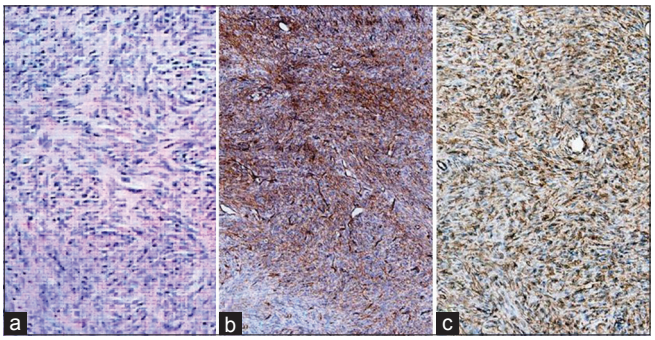

Figure 4: (a) Section showing spindle cells arranged in short fascicles, storiform pattern, oval nuclei, vesicular chromatin, inconspicuous nucleoli and scanty to moderate cytoplasm (HE, 100×). (b) Section showing tumor cell diffuse strong positive staining for CD-34 (100×). (c) Section showing tumor cell diffuse strong positive staining for vimentin (100×)

plaque which grows more nodular-exophytic growth pattern with time. Histopathologically, DFSP consisted densely packed, monomorphous, clump of spindle-shape cells arrange "storiform" or "cartwheel" of uniform appearing fibroblast; whereas at the periphery there was diffuse infiltrative into subcutis. IHC staining demonstrate spindle cells with strong positivity for CD-34 and vimentin expression as an important feature for diagnosing DFSP. CD-34 positivity is particularly useful for the differential diagnosis of DFSP from the fibrohistiocytics lesions that shown negative CD-34 expression. The fibrosarcomatous variant of DFSP (FS-DFSP), loss of CD-34 gradually less than 50%, represents a form of tumor progression and associated with a significantly more aggressive clinical course than ordinary DFSP.[5] A proper decision regarding the surgical margin for aiming a complete resection of DFSP and its reconstruction are challenging, not to mention that the fact we can face a major complication after surgery such as a flap failure. Other modalities such as postoperative chemotherapy and/or radiotherapy should be also considered after we can prove the evidence of malignant changing of DFSP into more aggressive variant. Although scalp DFSP have been rarely reported infiltrate into intracranial.

CONCLUSION

A successful treatment and management depends on the achievement of local control and the prevention of cosmetic and functional deficit; all efforts should be made for complete excision with negative margin. Postoperative follow-up using magnetic resonance imaging or positron emission tomography is recommended for highly suspicious cases, and annual checkups should performed up to 5 years after definitive therapy.

CONSENT

Informed consent was obtained from both patients for publication of this two case reports and any accompanying images. Their family was present at the time.

COMPETING INTERESTS

The authors declare that they have no competing interests.

AUTHORS' CONTRIBUTIONS

All authors had examined, treated, observed and followed up the patient and participated in writing the manuscript. All authors has read and approved of the final manuscript.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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