CASE REPORT

Late Recurrence of Dermatofibrosarcoma Protuberans in a Previously Skin Grafted Forearm

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Introduction

Dermatofibrosarcoma Protuberans (DFSP) is an extremely rare low-grade sarcoma of fibroblast origin, with an incidence in the United States estimated at less than five cases per million persons per year [1, 2]. Though metastasis of DFSP is rare, local recurrence rates as high as 60 % have been reported [1, 2]. This is most likely a result of the tumor's highly irregular three-dimensional shape with “finger-like” extensions, which allow for extensive involvement of subcutaneous tissue, muscle, fascia, and bone [1, 2]. Majority of local recurrences occur within the first 3 years of resection, with recurrences at greater than 10 years from index resection very rarely reported [3].

Case Report

We present a highly unusual case of a middle-aged male confirmed to have DFSP of the volar forearm, at a site that was previously resected and skin-grafted as a child nearly 40 years

prior. The patient was a 42-year-old Caucasian right-hand-dominant man who was referred to our specialty hand clinic for further management of biopsy-confirmed DFSP of his left volar forearm. The patient had a prior history of surgery at that same location, where a mass was excised and the defect was covered with a skin graft at the age of three. The patient and his family were unaware of any tissue diagnosis and long-term follow-up had not been recommended. He remained asymptomatic and noted no changes in the appearance of the grafted area for nearly four decades. Approximately six months prior to presentation, the patient noticed three “bumps” within the previously grafted area, but remained asymptomatic. One month prior to referral to our clinic, an excisional biopsy was performed, confirming DFSP.

Physical examination findings at the time of our initial evaluation were mostly unremarkable; range of motion, strength and nerve testing were all grossly normal. Examination of the skin revealed a well-healed skin graft over the left volar forearm with some darkening along the edges of the graft. (Figure 1) Plain radiographs were normal, while an MRI demonstrated abnormalities relative to the skin without deep invasion.

Fig. 1 Pre-operative appearance of patient's volar forearm with planned surgical margins marked

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Initial management consisted of radical resection of the tumor with margins of 3 cm circumferentially around the lesion, extending from just proximal to the wrist crease to the mid forearm (Fig. 2). The tumor was harvested, along with the adhered volar forearm fascia, with care taken to protect the underlying neurovascular structures and flexor tendons. No gross extensions of tumor were identified, and the specimen was sent to pathology. After tourniquet release and achievement of hemostasis, a series of Integra Template® skin substitute sheets (Integra LifeSciences, Plainsboro, NJ) were sewn in place and stented. A short arm splint was applied.

Pathologic assessment of the tissue revealed clear margins at the distal, radial and ulnar borders of the specimen, with positive margins at its proximal and deep aspects. Microscopically, low-grade spindle cell proliferation of the dermis was seen extending to edges of the specimen, with low mitotic activity, minimal nuclear polymorphism, positive CD34 stain and a negative S-100 stain. An oncologic surgeon was consulted and adjuvant therapy was not adopted, as it was felt an adequate final resection could be performed at the time of definitive reconstruction. Four weeks after the initial surgery, re-excision of the proximal and deep margins, with definitive reconstruction using a STSG of 10 by 12.5 cm harvested from the right thigh, was performed (Fig. 3). Pathology at that time showed all margins free of tumor. Thirteen months after excision of the tumor and skin grafting, the patient continued to do well, with the skin graft appearing appropriately healed and well incorporated. Clinically, there were no signs of recurrence on examination.

Discussion

This case represents a very unique scenario involving an already-recognized rare tumor. Though the patient's original diagnosis as a child is not confirmed, a recurrence of DFSP is the most-likely etiology. It is estimated that only six percent of DFSP is present in children less than 16 years of age [4].

Fig. 2 Intraoperative view of resected tissue from initial surgery and the resulting forearm defect

Fig. 3 Intraoperative view of forearm after placement of split-thickness skin-graft (STSG) during definitive reconstructive surgery

Though the characteristics of DFSP are similar in children and adults, the sites of occurrence often differ; DFSP has a predilection for the legs and acral regions in children versus mainly affecting the trunk and proximal extremities in adults [2, 4]. Though this epidemiologic data suggests that DFSP in the forearm of a 3-year-old child would be extremely rare, the high rate of local recurrence that seen in inadequately-excised DFSP does support a diagnosis of DFSP as a child in the presented case. However, a delay of 40 years between primary DFSP resection and recurrence would be longest ever reported [5].

Far less likely is the possibility of the presented case of DFSP occurring as a neo-tumor within the previously skin grafted area. Though there are reports of DFSP arising from previous trauma, burns or immunization sites, the occurrence of DFSP from autografted skin has never been reported [6].

Achievement of negative margins remains the top priority of surgical management for DFSP. Tumor depth and margin status were the strongest predictors of future recurrence in a large study performed at a major cancer center [5]. Post-operatively, follow-up and surveillance should include evaluation every six to 12 months, in conjunction with patient education on self-examination. In the case of recurrence, surgical re-resection is preferred, though the use of targeted radiotherapy as an alternative or and adjuvant may be considered. If the disease is locally advanced or non-resectable, or in the rare case of metastasis, a multi-modal approach including radiotherapy, targeted therapy and/or chemotherapy in consultation with a medical oncologist is warranted.

Compliance with Ethical Standards All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Conflict of Interest Author M. Gaspar, Author G. Landes, and Author A.L. Osterman declare that they have no conflict of interest.

Informed Consent Informed consent was obtained from the individual participant included in the study.

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