Journal of Plastic, Reconstructive & Aesthetic Surgery (2014) 67, 1222—1229

JPRAS
An International Journal of Surgical Reconstruction
www.JPRASurg.com

Dermatofibrosarcoma protuberans in children and adolescents: Clinical presentation, histology, treatment, and review of the literature

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Received 10 September 2013; accepted 14 May 2014

KEYWORDS
Dermatofibrosarcoma protuberans;
Children;
Treatment;
Adolescent

Summary *Purpose:* Dermatofibrosarcoma protuberans (DFSP) is a rare, low-grade malignant tumor. It is characterized by aggressive local infiltration, leading to a propensity for recurrence. In children, DFSP is even less common and likely misdiagnosed or underdiagnosed. This study is a review of DFSP in the pediatric population and aims to identify factors for successful treatment.
Materials and methods: From July of 1986 to 2011, a total of 159 patients were diagnosed with dermatofibrosarcoma protuberans at Kaohsiung Chang Gung Memorial Hospital, Taiwan. Subject to the age classification of our institution, patients under the age of 18 are defined in the pediatric category, of which159 cases were identified as our research subjects. Detailed data, including demographic data, imaging studies, pathology, treatment methods, and outcomes, of these identified patients were collected, reviewed, and analyzed.
Results: A total of 13 patients, consisting of six male and seven female patients, were identified based on our criteria. Two had the lesions noticed at birth. Most patients experienced a variable period of quiescence, followed by a rapid growth phase. All 13 patients underwent wide excisions. Post-excision reconstruction included direct closure in three cases, skin grafting in three cases, and local or free flap reconstruction in seven cases. Of 13 patients, four received postoperative radiotherapy. All patients survived without recurrence up to July 2011, with follow-up periods ranging from 20 months to 19 years.

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http://dx.doi.org/10.1016/j.bjps.2014.05.031

Conclusion: Clinicians should be aware that DFSP is known to occur among children. Owing to its relatively low incidence, its presence may be confused with commoner lesions such as hemangioma, fibroma, or atrophic plaques without nodule. The confusing situation, as a result, frequently leads to delayed diagnosis. Vigilance in its diagnosis allows for treatment at manageable sizes as well as ensures complete excision. Reconstructive options, such as skin grafting, and modalities, such as adjuvant postoperative radiotherapy, are suggested to best complement each other. The former minimizes disfigurement while the latter minimizes recurrences.

Introduction

Dermatofibrosarcoma protuberans (DFSP) is an uncommon soft-tissue tumor with low to intermediate grade malignancy, characterized by high rates of local recurrence yet low risk of metastasis.[1,2] DFSP was first described by Darier and Ferrand in 1924. Its occurrence in childhood was nevertheless not mentioned in the literature prior to 1957.[2] It is mostly located on the trunk and the proximal portion of the limbs. In 10—15% of cases, DFSP affects the head and neck regions, chiefly involving the scalp, cheek, and supraclavicular areas.[3] The initial presentation of DFSP is of an asymptomatic, indurated plaque with a hard consistency, which is fixed to the skin with not deep layers.[4]

DFSP typically ranges from 1 to 5 cm in size and tends to increase in dimension over years. The size would keep enlarging if left untreated.[5] The age of presentation varies widely, from being present congenitally to being diagnosed in patients over the age of 90. However, it mostly occurs to those of age between 20 and 50.[6,7] In case of pediatric DFSP, the proportion has been reported to be between 6% and 20% in the literature.[8,9]

The main histologic characteristic of DFSP is its capacity to invade surrounding tissue to a considerable distance from the central focus of the tumor. The histologic subtypes include pigmented DFSP (Bednar tumor), giant cell fibroblastoma (GCF), atrophic DFSP, sclerosing DFSP, granular cell variant of DFSP, and fibrosarcomatous DFSP (DFSP-FS).[9—15,17] The tumor with fibrosarcoma component (DFSP-FS) is a special variation with a higher rate of local recurrence after surgery and has an increased risk of distant metastasis.[13—15]

In many cases, DFSP diagnosed in adults has been present since childhood, mainly due to its slow growth and lack of symptoms. The treatment of choice for nonmetastatic DFSP is complete surgical resection with wide margins. Recurrence rate ranging from 26% to 60% has been reported in cases of conservative resection with undefined surgical margins.[18] In contrast, wide local excision (WLE) with a margin of at least 2—3 cm of normal tissue from the tumor boundary results in much lower recurrence rates ranging from 0% to 30%.[18—21] The objective of this study was to review our experience with the diagnosis and treatment of DFSP among children and the important factors contributing to successful treatment.

Patients and methods

In a 25-year period from 1986 to 2011, a total of 159 patients were diagnosed with DFSP at Kaohsiung Chang Gung Memorial Hospital, Taiwan. In our institution, children are categorized as individuals under the age of 18. This category of pediatric patients was identified from the 159 cases and enrolled in our study. Detailed data were collected and analyzed, including demographic data, disease factors such as age of onset and operation, history of trauma, and related symptoms. Details of intervention were also studied in detail, including types of surgical treatment, resection margins, reconstructive procedures, the use of postoperative radiotherapy, and postoperative complications. Outcome information such as recurrence, duration of follow-up and overall recovery were also collected and analyzed.

Results

Thirteen patients were identified, representing 8.3% of the entire cohort of DFSP patients. Girls outnumber boys at 7 to 6, for a male-to-female ratio of 0.86. Congenital presence of these lesions was noted among two cases (Patients 11 and 12), or 15.4% of all childhood DFSP. A history of trauma was described in two cases (Patients 2 and 10). All patients experienced a variable quiescent stage, followed by a rapid progression stage. The majority of DFSP cases were distributed over the trunk region (seven patients), followed by extremities (three patients), back (two patients), and head (one patient) (Figure 1A and B).

Most cases were treated by plastic surgeons with WLE, and where necessary, with immediate reconstructions. Three cases were closed primarily, while another three necessitated skin grafts, with seven other patients undergoing local or free flap reconstructions. The surgical margin was not recorded in one of these patients (Patient 6) who was operated by a non-plastic surgeon, which was yet considered a conservative resection with clear margin. Three patients underwent adjuvant postoperative