Repeated Local Recurrence of Dermatofibrosarcoma Protuberans in the Anterior Chest Wall of a Child

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a very rare, low-intermediate-grade sarcoma of the dermis layer of the skin; it has a marked propensity for local recurrence after excision. It is typically diagnosed during early adult life at a tumoral stage and it occurs rarely in children. This report is a case of childhood DFSP that had recurred five times within a period of eight years, each following surgical excision. This tumor also showed an uncommon myxoid change: Appearing as larger multi-nodular cutaneous plaque that arose at the site of excision of previous tumors some years earlier. The rarity of this tumor, and the fact that it is even rarer in children, prompted the report of this case.

Key words: Childhood dermatofibrosarcoma protuberans; myxoid change; repeated recurrence

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a low-intermediate-grade cutaneous sarcoma with marked propensity for local recurrence after excision.[1] It is very rare; the incidence is about one case per million per year. It behaves as a benign tumor but can metastasize in 2-5% of cases.[2] It is typically diagnosed during early adult life at a tumoral stage and occurs rarely in children.[3]

This report is a childhood DFSP with five local recurrences within eight years, each after surgical excision. The rarity of this tumor, and the fact that it is even rarer in children, prompted this report.

Case Report

Patient was a 14-year-old male admitted with 8-year-old history of recurrent mass in the anterior left shoulder. The mass, first noticed when he was 4 years old, and was initially slow-growing grew faster and bigger after each surgery. It was first excised about 2 years after it was first noticed, then 29 months after this first excision; the 3rd, 4th, and 5th excisions were done 19 months, 28 months, and 26 months, respectively, from each immediate previous excision. It was associated with itching, night sweats, fever, and remarkable weight loss. Following recurrence after the second surgical excision, ulceration and occasional bleeding started.

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Physical examination showed a cauliflower-shaped, ulcerated, 10 × 5 cm mass in the anterior left shoulder. The mass was firm, non-tender, warm to touch, and mobile over the surrounding structures. There was no surrounding edema, palpable lymph-nodes (in the axilla or cervical region) and no bruit. The rest of the physical examination was unremarkable.

Blood investigations showed a mild leucocytosis of 13,000/mm^3 and a negative reaction to human immunodeficiency virus (HIV) I and II antigens. Lateral chest radiograph showed a lobulated soft tissue mass anterior to the body of the sternum. It measured about 9 × 11 cm in the antero-posterior and cephalo-caudal diameters, respectively, [Figure 1] and had a spiculated margin in a black and white reconstructed image. [Figure 2]. There was no osseous involvement. Except for the overlying shadow of the mass in the left hemi-thorax, the chest radiograph was unremarkable [Figure 3].

The histology of the first excised specimen demonstrated the typical cart-wheel appearance of spindle cells that grow as sheets with few or no mitotic figures, in keeping with DFSP. A repeat histology of the 3rd excised specimen in the same center confirmed DFSP.

Post-operatively, his recovery was unremarkable and he was discharged for follow-up.

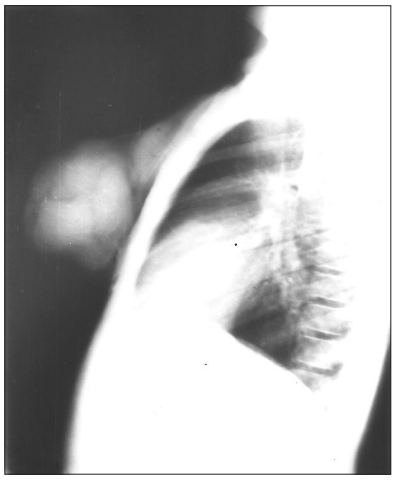

Figure 1: Lateral plain chest radiograph showing a lobulated soft tissue mass anterior to the sternum. Note that the lung fields are not involved

Figure 2: Black and white reconstructed image of figure 1 showing spiculation of margin of the soft tissue mass

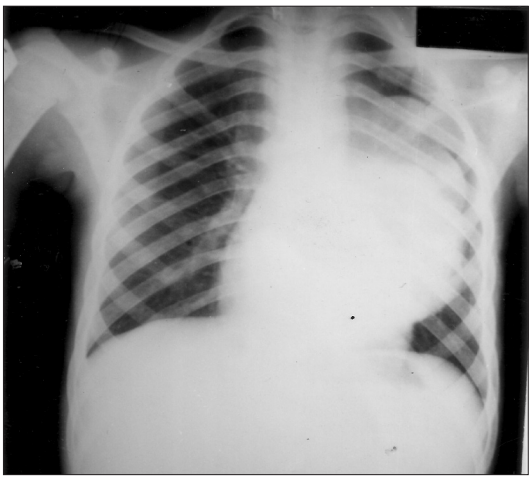

Figure 3: Postero-anterior chest radiograph of the patient in Figure 1 showing a large soft tissue mass overlying the left upper lobe and the left cardiac border

Discussion

Dermatofibrosarcoma protuberans (DFSP) is a fibrohistiocytic sarcoma and a typical superficial tumor involving the cutis and frequently the superficial subcutis.[4] When small, it has a characteristic histology of hour-glass shape, with a base in the dermis and two nodules, one superficial pressing the epidermis outwards (hence 'protuberans') and another larger one invading the deeper tissues. It usually occurs in the third and fourth decades of life but has also been described in children.[5] The occurrence of DFSP when our patient was about 4-years-old was rare.

Clinically, DFSP often begins as a non-protuberant plaque on the trunk, or on the extremities of middle-aged adults and occasionally persists as such.[6] Uncommonly, DFSP will show myxoid changes: Appearing as larger multi-nodular cutaneous plaques that arise at the sites of excision of previous tumors some years earlier.[7] DFSP in children does not show distinctive features when compared with adult forms except for a tendency for localization in the extremities like hands and feet.[3] The finding of a cauliflower-shaped, ulcerated mass in the anterior left shoulder in this patient would indicate that he had DFSP, which showed myxoid changes.

Diagnosis of DFSP is done by core needle or surgical incision biopsy. Microscopically, DFSP is composed of monomorphous spindle cells arranged in storiform pattern and embedded in a sparse to moderately dense fibrous stroma.[8] This finding was seen in the histology of excised biopsy specimens in this patient, thus confirming DFSP.

In this patient, the possible etiology of DFSP remains conjectural. There was no history of risk factors like trauma, irradiation, infarction, implanted foreign material, herbicides, chlorophenols, burn scars, injection sites, varicose ulcers, and at the site of prior immunization.

The cytogenetic features of DFSP include the translocation t (17; 22) (q22; q13) or more commonly, supernumerary ring chromosomes containing material from 17q22 and 22q13.[1] This rearrangement leads to constitutive activation of the platelet-derived growth factor receptor (PDGFR) as a result of deregulated ligand expression, thus providing a rationale for targeted inhibition of PDGFR as a treatment strategy for patients with unresectable, locally advanced, or metastatic DFSP.[1]

In the plain chest radiograph, DFSP appear as soft tissue mass or swelling without osseous involvement or calcification; this feature was seen in this patient. On computed tomography (CT) scan, it may appear as a moderately enhancing, well-defined, lobular, soft tissue lesion with tissue attenuation equal to or greater than that of skeletal muscle. Arteriogram will show mild to moderate hypervascularity. Bone scintigraphy may show increased accumulation of tracer.

The magnetic resonance (MR) appearance is non-specific and may range from prolonged T1 and T2 relaxation times to multiple nodular lesions.[9]

Surgery is the main therapy for DFSP but controversy exists regarding margin width and excision techniques with some advocating Mohs micrographic surgery with continuous histological control and others wide excision (3-cm margin of visibly uninvolved tissue, including the superficial fascia, for trunk and extremity lesions, and 2-cm margin for head and neck lesions).[10] Though chemotherapy and radiotherapy can sometimes be used and are effective, they are not curative.

DFSP has a particular propensity for local recurrence following seemingly adequate excision; metastases are rare, and usually follow repeated local recurrences. In this patient, surgery (wide excision) was the main treatment given, and there were five local recurrences within a period of 8 years. The repeated local recurrences have increased the metastatic risk, and thus necessitating a careful follow-up. It is very remarkable that this tumor had undergone an uncommon myxoid change: Appearing as larger multi-nodular cutaneous mass that arose at the site of excision of previous tumors some years earlier.

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