Recurrent dermatofibrosarcoma protuberans of lower anterior chest wall in a young male with early fibrosarcomatous change

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing tumor of fibro-histiocytic nature considered of low- to intermediate-grade malignant mesenchymal neoplasm. It mainly affects young and middle-aged male adults with an indolent, very slow but infiltrative growth pattern. It originates from the dermis, from which it often invades the subcutaneous tissue and accounting for 6% of all soft tissue sarcomas and less than 0.1% of all cutaneous malignancies. We report a case of 7 year old male child presented with recurrent DFSP which exhibited early fibrosarcomatous changes. It is a rare finding and challenging entity in histopathological diagnosis. This subset of patients requires wide excision with a 3 cm safety margin and may require adjunct radiotherapy.

Keywords: *Swelling, Infiltrative growth, lesion, FNAC, rare*

INTRODUCTION

The peroneus (fibularis) tertius is a variably present muscle of the anterior compartment, contributing—albeit weakly—to ankle dorsiflexion and eversion at the talocrural and subtalar joints. It typically arises from the distal fibula, interosseous membrane, and anterior intermuscular septum and inserts on the dorsomedial base of the fifth metatarsal via a tendon; it may be absent or confluent with the extensor digitorum longus (EDL) (1). Olewnik's classification describes several insertion patterns, with type II most common (~84.8%), while the type IV pattern—a split insertion to the base and shaft of the fifth metatarsal—is uncommon (~1.5%) (2). Occasional tendinous connections with EDL have also

A 7-year-old male child presented to the burns and plastic surgery department with complaints of recurrent swelling over the left lower chest wall for three years (Figure A). He also gives a history of excision of lesion two years back from the same site with no documented histopathology report. Magnetic resonance imaging (MRI) showed a soft tissue lesion with no muscular plane infiltration. Fine needle aspiration cytology (FNAC) revealed a spindle cell

lesion. Excision of the lesion revealed a proliferation of bland spindle cells in a typical cartwheel or storiform pattern (Figure B). These neoplastic cells infiltrate into the subcutaneous fat in the lacelike pattern. Few mitotic figures were also noted (3-4/10 HPF), along with occasional pleomorphic cells; however no necrosis seen. The overlying epidermis was typically thinned out. Immunohistochemistry for CD34 was positive, however certain areas showed a focal loss. Smooth muscle actin (SMA) showed patchy immune-positivity which showed myoid differentiation. Based on overall features a final diagnosis of dermatofibrosarcoma protuberans (DFSP) with early fibrosarcomatous change was given.

FIGURES AND IMAGES

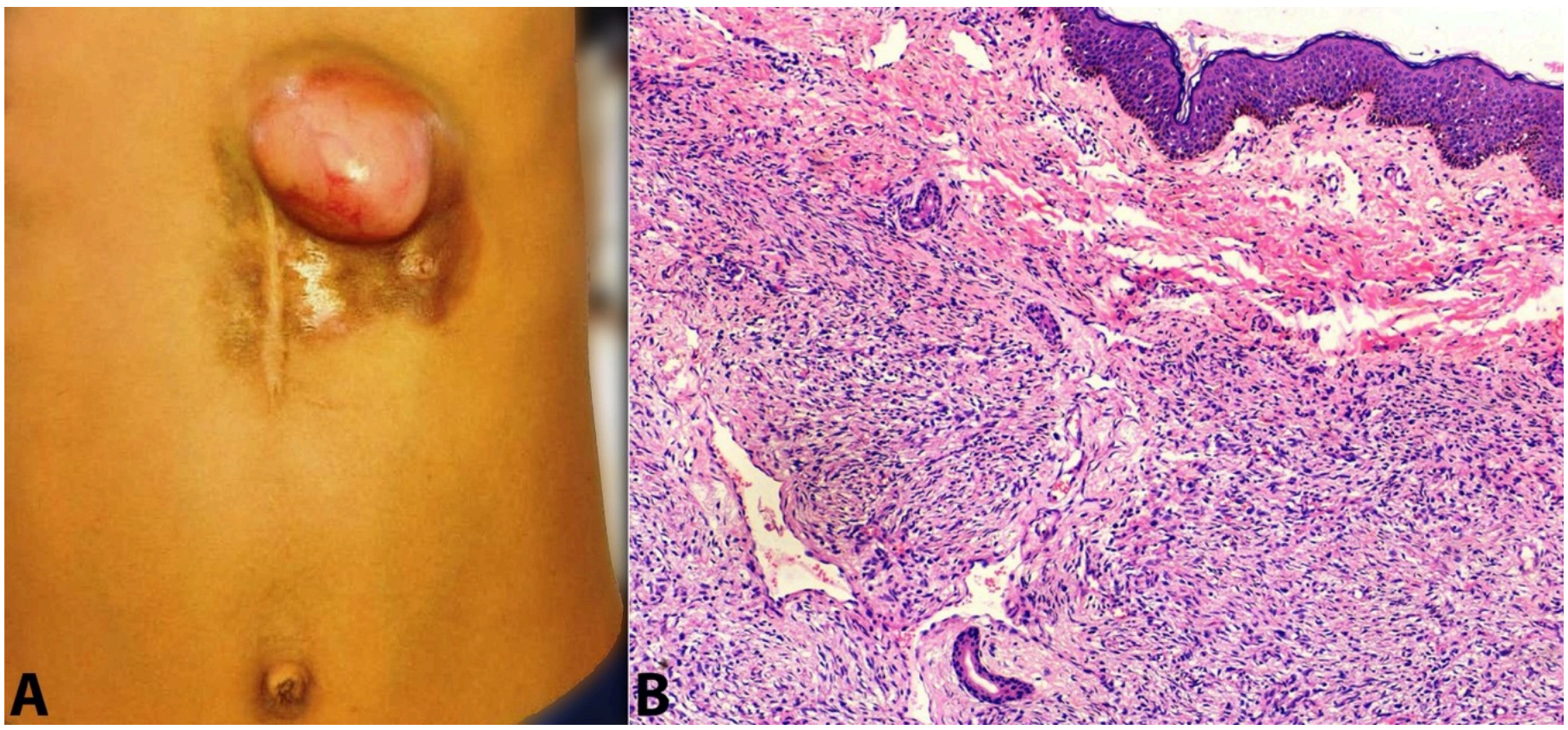

FIG A: Recurrent swelling over the left lower chest wall for three years FIG B: A proliferation of bland spindle cells in a typical cartwheel or storiform pattern

DISCUSSION

DFSP was initially reported in the literature as early as 1890, but in 1924 Darier and Ferrand first described DFSP as a distinct cutaneous disease and termed as "progressive and recurrent dermatofibroma". In 1925 Hoffman first termed it as "dermato-fibrosarcoma-protuberans".1-3

DFSP is a rare, slow-growing tumor of fibro-histiocytic nature considered of low- to intermediate-grade malignant mesenchymal neoplasm. Despite rarely metastasising it tends to recur. It originates from the dermis, from which it often invades the subcutaneous tissue and accounts for 6% of all soft tissue sarcomas and less than 0.1% of all cutaneous malignancies.2-3

It mainly affects young and middle-aged male adults with an indolent, very slow but infiltrative growth pattern. It most commonly occurs on the trunk (42-72%), followed by the proximal extremities (16-30%) and rarely occurs above the neck (10-16%).1-4

The origin of DFSP is still unclear or controversial but a genetic defect is present in 95% of cases due to either a reciprocal chromosomal translocation of t(17;22)(q22;q13) resulting in the formation of COL1A1-PDGFB fusion gene transcript, or the formation of a ring chromosome composed of hybrid material derived from t(17;22). This rearrangement leads to constitutive activation of the platelet-derived growth factor as a result of deregulated ligand expression.2-6

DFSPs usually arise as pink or violet-red plaques, while the surrounding skin may be telangiectatic. The main feature of DFSP is the horizontal spread of the tumour by creating neoplastic projections to all directions, like pseudopodia, resulting in the ejection of neoplastic cells up to 3 cm peripherally to the main tumour. This histological behaviour results in the high recurrence rate, as a result of satellite neoplastic cell populations that may be left in situ, during resection.3-5

Among the various treatment modalities, surgery is the gold standard treatment by wide excision with a safety margin of 3cm, including the underlying fascia since emphasis is on histological free margins for local control. Conventional chemotherapy is not commonly used to treat DFSP. Radiation therapy (RT) may be recommended if adequate wide excision may result in a major cosmetic defect or if the tumor margins are positive.1-6

CONCLUSION

DFSP is a rare tumor with intermediate-grade malignant potential. The fibrosarcomatous change requires careful histopathological evaluation with immunohistochemical support. Importance of diagnosis these changes is to consider them for further treatment like radiotherapy apart from standard surgical intervention.

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