

DERMATOLOGICAL SURGERY

DERMATOFIBROSARCOMA PROTUBERANS IN CHILDREN: THE SLOW MOHS MICROGRAPHIC SURGERY APPROACH

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Background: Dermatofibrosarcoma Protuberans (DFSP) is a rare slow-growing soft tissue tumour. The incidence in children is fewer than one per million before age 20 years. This locally aggressive tumour has a high recurrence rate due to its infiltrative growth pattern. Slow Mohs micrographic surgery (MMS) results in lower recurrence rates and smaller defect size for an equivalent cure rate compared to wide local excision. A Mohs map is constructed of the defect and sutures are used to mark areas. The Mohs layer is cut as per the map and each piece of tissue stained at the edges with a colour code to allow later orientation. The tissue is fixed in formalin and horizontal histological sections are prepared of the deep and peripheral margins. There are significant challenges in the histopathological interpretation of the sections. There is limited published data in the treatment of DFSP in children.

Observation: We present three children with DFSP, all treated with Slow MMS. CD34 immunohistochemistry and FISH COL1A1 gene translocation studies were also performed: Case 1: 4yo white male with DFSP on the occiput was completely excised after single stage Slow Mohs and reconstructed with Integra and a scalp split skin graft. Case 2: 8yo black female with DFSP on the calf, was completely excised with two stages of Slow MMS and reconstructed with Matriderm and a split skin graft. Case 3: 7yo Middle Eastern male with atrophic DFSP of the forearm, was completely excised with two stages of Slow MMS and reconstructed with Matriderm and a split skin graft.

Key message: DFSP can recur if inadequately treated and rarely can also metastasize. Slow MMS is our preferred surgical technique when treating DFSP in children. The main advantage is that the defect size is as small as possible with clear histological margins.