Position statement on management of Dermatofibrosarcoma Protuberans (DFSP)

BRITISH SOCIETY FOR DERMATOLOGICAL SURGERY

Dermatofibrosarcoma protuberans (DFSP) is a relatively rare malignancy, with an estimated incidence ranging from 0.8 to 5 per million people per year. It can occur at any age although the majority of cases present between 20 and 50 years of age. It occurs on the trunk in around half of cases but may also involve proximal extremities, head and neck.

DFSP presents usually as a cutaneous malignancy with slowly evolving indurated asymptomatic plaques, which may be atrophic or multinodular. The condition is asymptomatic and diagnosis is frequently delayed. Systemic metastasis is rare, with regional and distant metastases to the lung most common. The risk of metastasis may increase with incomplete initial treatment. The tumour has an infiltrative pattern, often with considerable subclinical extension and may involve surrounding muscle and fascia.

The primary treatment for most DFSP is surgical resection. Historically, wide excision margins, from 3 to 5 cm, were recommended, due to subclinical involvement. In recent years Mohs micrographic surgery has become the preferred choice of treatment, as this technique permits 100% tumour margin control.

The differences between wide local excision and Mohs surgery include the pathology processing and the detailed mapping with the latter. The Mohs technique has the advantage of sectioning en face along all margins. This increases the chance of detecting any residual tumour, which can then be precisely re-excised using mapping. This is particularly relevant for DFSP as it is a contiguous tumour, often with subclinical and asymmetrical extension. Accurate tumour removal ensures that no unnecessary additional normal tissue is removed. Although no randomised controlled studies exist, pooled data from the literature demonstrates a relative risk of recurrence of 15.9 (95% CI 7.2-35.5) comparing wide local excision to MMS. (Paradisi A et al. Cancer Treatment Reviews 2008; 34: 728-736).

Histological interpretation of Mohs surgery can be challenging and use of CD34 immunostaining may help in some cases.

Other treatment modalities for DFSP may include radiotherapy or imatinib.

Management of DFSP needs to be directed by a multidisciplinary team. Based on the available evidence in the literature, Mohs micrographic surgery is the initial treatment of choice for DFSP providing it is practical to perform. The specific circumstances of a case may determine an alternative treatment.