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CASE REPORT

Surgical decision criteria: Bednar tumour of the foot in a child

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Summary An 8-year-old boy was admitted for excision of a putative 'blue nevus' on the left foot. Histological examination and immunohistochemistry revealed a Bednar tumour, the pigmented variant of dermatofibrosarcoma protuberans. Surgical options considered by a multidisciplinary team included wide local excision, Mohs micrographic surgery or a staged excision with examination of several histological sections. The third alternative procedure was chosen after consideration of tumour and patient factors to achieve the best possible clinical, cosmetic and functional outcome. After the final surgical procedure with resection of the third metatarsal bone, all peripheral margins were free of tumour, and the interdigital space was reconstructed with a pedicled pulpa flap. Three years after surgery, there was no tumour recurrence, and further long-term follow-up for this patient will be provided.

Introduction

Dermatofibrosarcoma protuberans (DFSP) is an uncommon fibrous tumour that infiltrates deep into the dermis and adipose layers in a fascicular growth pattern. Histologically, the DFSP is a CD34+ spindle cell proliferation arranged in a storiform pattern with limited pleomorphism and a low mitotic index.[1] The pigmented variant, the Bednar tumour, is extremely rare and accounts for approximately 1–5% of all cases of DFSP.[1,2]

DFSP is of low malignancy with low potential for metastases, but it shows a high rate of recurrence.[3] Surgically, the standard treatment is wide local excision (WLE). Alternatively, many authors advocate Mohs micrographic surgical technique (MMS) as the treatment of choice for DFSP, even in the paediatric population.[1,4]

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We report our experience in a case of Bednar tumour occurring on the dorsal aspect of the foot of a boy, who, after careful consideration of tumour localisation and histology, underwent a staged surgical excision involving amputation of the third ray.

Case report and methods

An 8-year-old Asian boy presented with a slow growing 1 × 2-cm bluish plaque on the dorsal aspect of his left foot. A smaller, asymptomatic 'hyper pigmented' macula had been present since birth, which now appeared as a nevus-like lesion blue to grey in colour with a light scab and tender on palpation. Clinical diagnosis by a dermatologist was 'blue nevus', and the patient was referred for excisional biopsy.

Histopathology yielded a Bednar tumour reaching the resection margins. The tumour was composed of spindled cells of medium size arranged in a storiform pattern. It was highly cellular and abutted the overlying epidermis. Honeycomb infiltration of the adjacent subcutaneous fatty tissue was seen at the tumour margin. There was ample finely granular brown-black pigment in scattered multipolar stellate dendritic cells (Figure 1). Immunohistochemistry revealed expression of CD34 in the spindled tumour cells and HMB 45 in the scattered pigmented dendritic cells. Fluorescence *in situ* hybridisation (FISH) for an EWS 22q11 break-apart probe was positive and therefore consistent with a t (17; 22) translocation. Preoperative staging including radionuclide bone scan, computed tomography (CT) of the trunk and magnetic resonance imaging (MRI) of the foot, pelvis and skull revealed no metastasis.

Following careful consideration of different surgical options, including WLE and MMS, to preserve as much of the forefoot as possible, a staged surgical procedure was performed with all steps carried out under general anaesthetics.

The first operation after the biopsy was a wide excision along an accurately marked margin of approximately 1.0 cm (Figure 2). Tissue was excised perpendicularly (i.e., at a 90° angle), then mapped, dyed and sent to pathology for rush permanent paraffin-embedded sections. Orienting ink was applied and each block was step sectioned to ensure thorough sampling.[5] Subsequently, the edge was examined entirely (along with horizontal sections of the deep margin) for the presence of tumour by a pathologist with expertise and interest in DFSP. Immunohistochemical stains were not used at this stage. Meanwhile, the wound was covered with Epigard® (Medisave Medicalproducts, Wiesbaden, Germany). A second operation focused on re-excisions directly at the sites of the positive margins, which led to resection of the third metatarsal bone distal to the line of Lisfranc. In the same operation, primary wound closure was achieved by using the third toe as a vascularised flap to cover the interdigital space (Figure 3).[6] The envelope of the toes was used after removal of the bony and tendinous components for optimal adaption into the web space. Final histological examination revealed tumour-free margins.

Postoperatively, the patient developed a hypertrophic scar formation, which improved significantly under treatment with silicone gel sheeting and application of 585-nm pulsed dye laser. Four months after surgery, the patient was wearing normal shoes. At 3-year follow-up, neither clinical examination nor control magnetic resonance imaging (MRI) of the left foot revealed any evidence of local tumour recurrence. The patient is well with a normal gait and participates actively in sports, such as running and football.

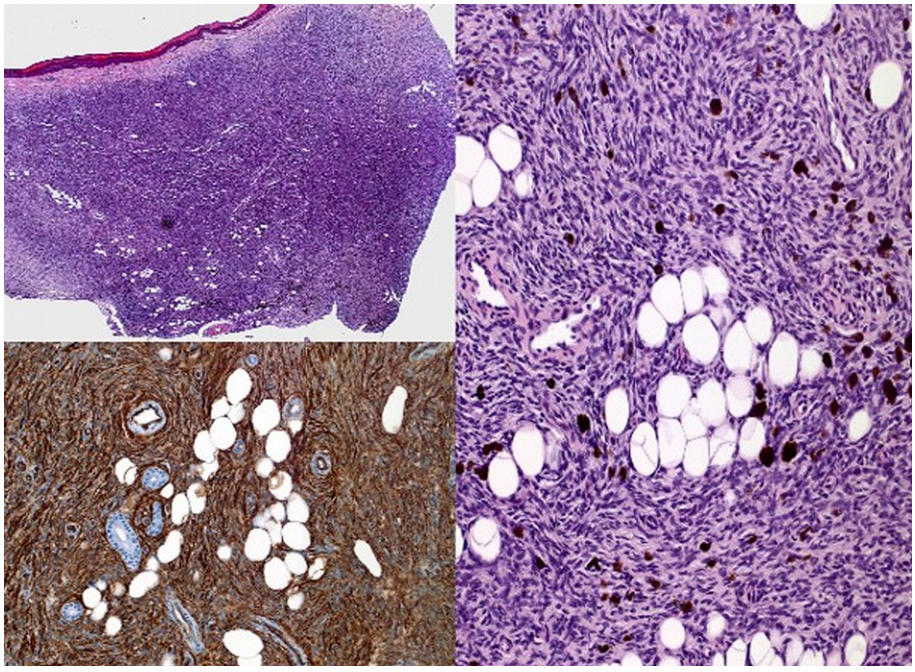

Figure 1 Tumour cells were spindled and arranged in a storiform pattern with scattered pigmented multipolar stellate cells. Immunohistochemistry demonstrated strong CD34 expression in the cytoplasm of all spindled tumour cells.