Congenital-Infantile Fibrosarcoma of the Foot â Avoidance of Amputation

Abstract:

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Abstract

Congenital-infantile fibrosarcoma is a rare entity with a five year survival rate of over 90%. Surgery is still the most common treatment modality with amputation often necessary. There have been reports supporting the use of neoadjuvant chemotherapy to debulk the tumour in an effort to facilitate limb sparing surgery. We report a case of a newborn who presented with a life threatening haemorrhage from a fibrosarcoma of the foot, successfully treated with Vincristine, Actinomycin and Cyclophosphamide (VAC) chemotherapy alone.

Introduction

Congenital infantile fibrosarcoma (CIF) is rare and represents <1% of all childhood cancers[1]. It is however the most common soft tissue tumour under the age of one year[2]. Historically wide surgical excision or even amputation has been required to treat CIF[3].

Case Report

A 12 day old boy presented with a large ulcerated soft tissue tumour on his left foot (Figure 1). A punch biopsy was performed. The deep dermis was essentially effaced by a spindle cell proliferation with a diffuse growth pattern. Prominent ectatic vascular spaces were identified and there was extensive interstitial haemorrhage. The initial histologic features were those of a primitive mesenchymal proliferation with the differential diagnosis, including Kaposiform haemangioendothelioma, congenital infantile fibrosarcoma (CIF) and, less likely, rhabdomyosarcoma. Plain x-rays of the foot demonstrated a large lobulated soft tissue mass with bones of the foot containing normal ossification centres for age. Magnetic Resonance Imaging (MRI) revealed a large 6.9 cm x 7.3cm x 8.2cm mass engulfing the entire left foot, splaying the metatarsals but without any bony infiltration (Figure 2).

The following day at change of dressing, he had a significant bleed from the biopsy site becoming rapidly haemodynamically unstable. He required emergency oversewing of the tumour to stop the bleeding. A single dose of intravenous Vincristine, Tranexamic acid and Prednisolone was administered. Further analysis of the histological specimens showed that immunohistochemical stains were negative for Desmin and Myogenin. D24-40 was limited to ectatic vascular channels and Glut 1 was negative. CD31 stains obvious vascular channels with unconvincing positivity in the intervening spindle cell population and CD34 was negative. A diagnosis of congenital infantile fibrosarcoma was confirmed histologically by the presence of the pathogonomic translocation t (12:15). He was commenced on a combination of Vincristine, Actinomycin and Cyclophosphamide. Total duration of chemotherapy was over a 28 week period. He is weight bearing in soft shoes and has full range of motion of his foot 24 months later.

Discussion

The differential diagnosis for a large, congenital, enlarging tumor of the foot includes a number of uncommon and rare entities. The most likely clinical scenario will involve differentiating a hemangioma from mimickers such as kaposiform hemangioendothelioma and tufted angioma. Other rare entities include lymphatic malformation, congenital hemangiopericytoma, embryonal rhabdomyosarcoma, infantile fibromatosis[4] or myofibromatosis, malignant fibrous histiocytoma, and malignant peripheral nerve sheath tumor[4]. Surgical excision currently remains the mainstay of treatment but the surgical approach has evolved from mutilating operations to more conservative organ-sparing procedures[5]. In some cases amputation is necessary if the extent of the tumour precludes surgical therapy. Preoperative chemotherapy may be useful for decreasing tumour bulk, enabling a more conservative surgical approach[6]. However there have been some reports of complete response following chemotherapy alone[7]. The complete remission rate is high after chemotherapy, with an overall survival rate of ~80%[8]. Kurkchubasche et al, reported that 43% of CIF treated with chemotherapy alone resulted in complete remission[9].

Our case demonstrates a conservative treatment approach resulting in a favourable response to chemotherapy avoiding amputation as was shown by Demir et al. in 2009[10]. It highlights the chemosensitive nature of these tumours. This case adds to a limited data set supporting chemotherapy alone followed by close observation as a treatment modality for a tumour with low metastatic potential in an effort to avoid limb amputation or at least facilitate limb sparing surgery. CIF is a rare tumour of childhood. The mainstay of treatment has been wide surgical excision, often requiring amputation. It is however a chemosensitive tumour with low metastatic potential. Our case illustrates the potential for avoidance of limb sacrificing surgery with the implementation of early aggressive chemotherapy.

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