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Van Nes rotationplasty as a treatment method for Ewing's sarcoma in a 14-month-old[☆]



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ABSTRACT

INTRODUCTION: In recent years, the rotationplasty procedure has become popular amongst tumour surgeons as an alternative to endoprosthetic replacement or amputation. There are very few documented cases of this technique in young patients with malignancy.

PRESENTATION OF CASE: We describe an extremely rare case of Ewing's sarcoma in a 14-month-old boy that involved the entire length of the left femur. At initial presentation, pulmonary metastatic spread had occurred and there was no neurovascular involvement. Complete response to neo-adjuvant chemotherapy was achieved prior to performing the definitive surgical procedure.

DISCUSSION: This case highlights the many reconstructive options and difficulties encountered in managing such extremely young patients with aggressive malignant disease. In this case, a complete femoral excision was necessary and various treatment options were explored. These included irradiation and re-implantation, endoprosthetic replacement and manufacturing a custom growing prosthesis. Taking future functional, psychological and social implications into consideration, we performed a total femoral excision and Van Nes rotationplasty of the tibia at our institute. Histological analysis of the tumour resection showed clear tumour margins and at 1 year clinical review, the patient demonstrates good functional outcome with no evidence of disease recurrence.

CONCLUSION: Van Nes rotationplasty is a viable reconstructive option in young patients with sarcoma of the femur. We believe this to be the youngest reported case of rotationplasty in current literature.

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1. Introduction

Ewing's sarcoma was first described in the 1921.¹ It constitutes 3% of all paediatric bone tumours and is the second most common malignancy after osteosarcoma. Ewing's sarcoma has an approximated incidence of 2 million per year in the under fifteen year age group. It is more common in the adolescent years, and carrying a male preponderance with a male:female ratio of 1.3–1.5:1. Clinicopathological studies have identified a translocation t(11;22)(q24;q12) that results in the formation of *EWS-ETS* fusion gene responsible for the pathogenesis of the tumour.²

The mainstay of treatment includes chemotherapy, radiotherapy and surgery. Many surgical procedures (limb sparing, sacrificing and allografting) have been described in the literature. The rotationplasty technique was originally performed by

Borggreve in the 1930s³ and was later popularised by Van Nes in the 1950s in children with proximal femoral focal deficiency (PFFD).⁴ The Van Nes rotationplasty technique has since grown favour amongst tumour surgeons worldwide as an alternative to endoprosthetic replacement or amputation, either as a primary or alternative procedure.⁵ The use of Van Nes rotationplasty in tumour surgery involving either the femur or tibia has been widely documented in terms of surgical technique, rehabilitation and functional and psychosocial outcomes.⁶

We describe the case of a 14-month-old boy who was referred to our specialist Bone Tumour Unit after initial diagnosis was made at his local hospital. We believe this to be the youngest reported case of its kind, treated with left femoral excision and Van Nes rotationplasty.

2. Case presentation

A 14-month-old boy presented locally with a painful left leg. He had been unable to weight bear in recent weeks as this had increased the severity of pain. Initial clinical assessment was performed at the referring hospital consisting of blood analysis and radiological investigations.

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Fig. 1. Pre-operative MRI with STIR sequence (left femur): demonstrating expansion of the left femur from the neck of the femur into the shaft with bowing and marrow signal abnormality of lower third of the shaft of the femur.

Plain radiographs of the left femur demonstrated Ewing's sarcoma that involved the entire length of the femur. Subsequent MRI revealed expansion of the femur from the neck into the shaft with bowing and persisting marrow signal abnormality in the lower third of the femoral shaft (Fig. 1). A CT guided needle biopsy and bone marrow trephine was performed, demonstrating a high grade small round cell tumour. This expressed CD99 with a strong perimembranous stain molecular genetic studies confirmed a positive FISH for EWS gene rearrangement with RT-PCR analysis detecting a EWS/FL11 type 2 transcript. A histopathological diagnosis of Ewing's sarcoma/PNET was made.

He was consequently referred to our unit and discussed at the National Sarcoma MDT meeting. A staging chest CT and ^{99m}Tc bone scan was performed which showed evidence of pulmonary metastatic spread (Fig. 2).

An informed multi-disciplinary team (MDT) decision to proceed with total femoral excision and a Van Nes rotationplasty was made. Pre-operative management involved oncological therapy for the metastatic disease and stem cell transplantation. The patient received 6 cycles of VIDE (Vincristine, Ifosfamide, Doxorubicin and Etoposide) chemotherapy (Euro Ewing 99 protocol) and GM-CSF therapy. A good response was seen treatment and repeat CT chest showed complete resolution of the pulmonary metastases after the 5th chemotherapy cycle.

The surgical procedure was performed at our institute. Post procedure a total femoral resection specimen (175 mm long) was sent for histological analysis (Fig. 3). Soft tissue dissection showed clear tumour margins. Microscopic examination demonstrated only focal areas of intramedullary fibrosis with greater than 80% tumour necrosis following chemotherapy, confirming a complete response to neo-adjuvant treatment (Fig. 4).

Post-operative management involved immobilisation in a hip spica for 6 weeks with a further 12 weeks administration of chemotherapy. Clinical review at 6 weeks was reassuring as he was able to crawl without any restrictions. The neo hip joint was stable on examination demonstrating 20° flexion and extension, and 10°

Table 1

Specific principles for the Van Nes rotationplasty procedure.

The foot and ankle MUST be disease free
The nerve supply to the foot and ankle MUST be preservable
The vascular supply to the foot and ankle MUST be either preservable or restorable after resection of diseased vessel segment
Muscle power MUST be restorable in the ankle following surgery
Large tissue segment resections are achievable

Table 2

Indications for the Van Nes rotationplasty procedure.

Lesions of the distal or proximal thirds of the femur and proximal tibia
Expected remaining growth in contralateral leg > 10 cm in young children
Extent of tumour size resection that may leave poor bone or soft tissue stock for reconstruction
Distal vascular supply compromised secondary to tumour (reconstructable by segmental resection and anastomosis)
Physical function and activity of major importance to child/adolescent and outweighs importance of cosmetic appearance
Previous failed reconstructive attempts
Late complications (leg length discrepancy/inadequate bone stock/failure of endoprosthesis)

of internal and external rotation. There were no vascular or neurological complications following the procedure and at 12 weeks, the patient was referred to our limb fitting service.

At 1 year follow up, the patient is able to stand and walk independently with the use of a prosthetic leg (Fig. 5). He displayed 50°, 10° and 60° of flexion, hyperextension and abduction respectively. Repeat MRI demonstrated no evidence of recurrence. Chest radiograph and abdominal ultra-sound were also clear of disease.

3. Discussion

The Van Nes rotationplasty is an extensively documented procedure with favourable published results.^{7,8} We believe this to be the youngest documented case, following a review of current literature.

The principles (Table 1) and indications (Table 2) for the Van Nes rotationplasty must be addressed before undertaking the procedure.^{9–11} The procedure involves rotating the foot 180° about a vertical axis so that ankle dorsiflexion simulates knee flexion. Proposed advantages of this procedure is that it allows function more akin to transtibial rather than transfemoral amputation due to retention of voluntary control of motion at the 'knee' level and the ease of application of a below knee prostheses. Pre-operative planning consisted of taking into account both the differential growth in the operative leg (proximal femoral and distal tibial epiphysis and by growth of the calcaneotalar unit) and growth in the contralateral leg (proximal and distal femoral epiphyses).

This case demonstrates the difficulties posed by an extremely young patient presenting with an aggressive malignancy that involved the entire length of the femur. We explored various treatment and reconstructive options in his management. We highlight some learning points in this case report, particularly in the pre-operative planning stages.

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