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Congenital Infantile Fibrosarcoma: A Case Report and Literature Review

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ABSTRACT

Introduction: *Congenital infantile fibrosarcoma (CIF) is a rare soft tissue tumor, typically diagnosed at birth or during the first few months of life. In contrast to the adult form, it has a more favorable prognosis, despite sometimes misleading clinical and radiological features. Treatment usually involves complete surgical excision, but neoadjuvant or exclusive chemotherapy may be considered a therapeutic option, particularly when the tumor is located in areas where surgery would cause significant mutilation.*

Methods: *We report the case of a male newborn, born on March 9, 2022, presenting with a congenital and progressively enlarging swelling of the left forearm. Initial imaging suggested a hemangioma on Doppler ultrasound, while MRI findings were indicative of fibrosarcoma. The diagnosis of congenital infantile fibrosarcoma was confirmed by biopsy and histopathological examination. Although the multidisciplinary tumor board initially recommended amputation, a review of the literature guided the therapeutic decision toward exclusive chemotherapy using the VAC protocol (first cycle) and IVA protocol (second cycle), followed by a course of cyclophosphamide, vincristine, and doxorubicin.*

Results: *At 12 months, a marked reduction in tumor size was observed. At 24 months, a small, stable residual mass without any suspicious tumor activity remained. After three years of follow-up, the patient showed no recurrence, had satisfactory functional development of the upper limb, and exhibited no significant clinical sequelae.*

Conclusion: *This case demonstrates that an exclusively chemotherapeutic, limb-sparing approach can avoid major amputation while achieving complete control of congenital infantile fibrosarcoma. Continued clinical and radiological monitoring is essential to ensure the absence of recurrence. This therapeutic strategy may represent a valuable alternative for infants with fibrosarcomas located in anatomically critical sites.*

Keywords

Congenital infantile fibrosarcoma, Newborn, Exclusive chemotherapy, VAC, IVA, Amputation avoided, Long-term follow-up, Soft tissue tumor.

Introduction

Infantile fibrosarcoma (IFS) is a rare malignant soft tissue tumor, accounting for approximately 5 to 10% of sarcomas diagnosed in children under one year of age [1,2]. It typically appears at birth or during the first few months of life, frequently located in the extremities, and generally exhibits a less aggressive course than adult forms, despite the possibility of local recurrence or metastasis [3]. Diagnosis is made more challenging because the tumor can mimic benign lesions (such as angiomas or vascular malformations), thus requiring a multidisciplinary approach involving imaging, pathology, hematology, oncology, molecular biology, and surgery [4,5].

At the molecular level, the translocation t (12;15) (p 13; q25), involving chromosomes 12 and 15, results in the fusion of the ETV6 gene (located on 12p13) and the NTRK3 gene (located on 15q25). This is a major diagnostic marker of IFS and paves the way for specific targeted therapies [6]. Although the standard treatment traditionally relies on complete surgical excision, this can lead to significant functional sequelae, especially when located in the extremities [1,7]. In this context, chemotherapy plays a crucial role: used in a neoadjuvant setting, it often reduces tumor size, facilitates more conservative excision, or even allows avoiding surgery altogether. Furthermore, the advent of TRK inhibitors such as larotrectinib has demonstrated extremely high response rates (over 90%), opening the door to increasingly specific and non-mutilating approaches for IFS [8-10].

Objective

This work reports the rare case of an infant with congenital infantile fibrosarcoma of the left forearm, treated exclusively with chemotherapy, thus illustrating the major benefit of this therapeutic choice in preserving limb function while ensuring durable tumor control.

Materials and Methods

The patient is a male newborn, born on 03/09/2022, with no notable medical history, admitted for a congenital swelling of the left forearm, with sudden onset and progressively worsening evolution. Initial diagnostic investigations included a Doppler ultrasound, suggesting a pattern compatible with a hemangioma, as well as an MRI revealing a large mass suggestive of fibrosarcoma. A biopsy followed by histopathological examination confirmed the diagnosis of congenital infantile fibrosarcoma. During the multidisciplinary team meeting, amputation was initially proposed. However, based on literature data showing good outcomes with chemotherapy alone, a conservative treatment strategy was chosen.

Results

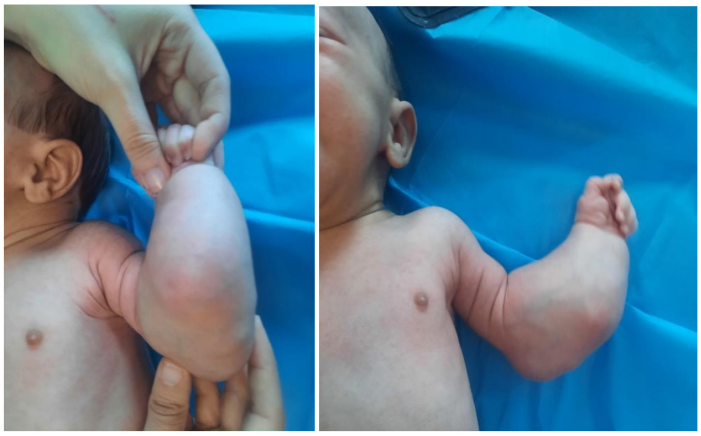

Figure 1: Newborn male (date of birth: March 9, 2022) presenting with a congenital mass of the left forearm, suggestive of infantile fibrosarcoma.

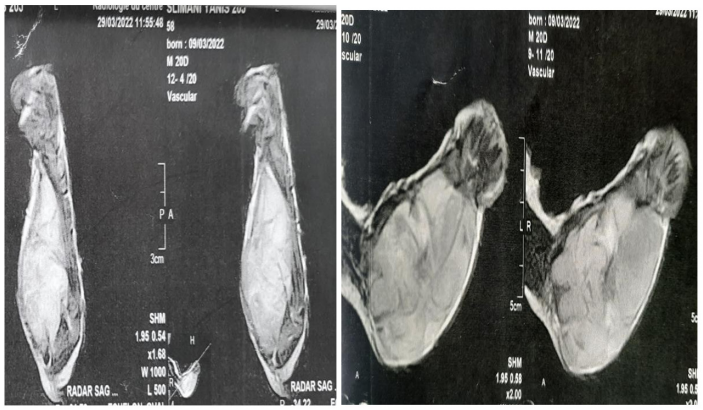

Figure 2: MRI of the infant's left forearm showing a large soft tissue mass suggestive of infantile fibrosarcoma.

Table 1: Treatment timeline and clinical evolution of congenital infantile fibrosarcoma.

Treatment Phase	Date	Action / Cycle	Evaluation
1st cycle (VAC)	05/18/2022	VA	
	05/25/2022	VAC1	
	06/08/2022	VAC2	
	07/06/2022	VAC3	CT scan: slight regression of the mass (45 × 42 mm)
2nd cycle (IVA)	07/28/2022	IVA1	
	08/23/2022	IVA2	
	09/14/2022	IVA3	
	10/12/2022	IVA4	MRI: significant tumor regression
	11/22/2022	IVA5	
Additional treatment	01/03/2023	Cyclophosphamide + Vincristine + Doxorubicin	
Clinical evolution	At 12 months		Significant reduction of the mass
	At 24 months		Persistence of a small stable residue (CT scan, PET scan), without suspicious tumor activity
	At 3 years follow-up		No recurrence, satisfactory functional development of the left forearm, without significant clinical sequelae

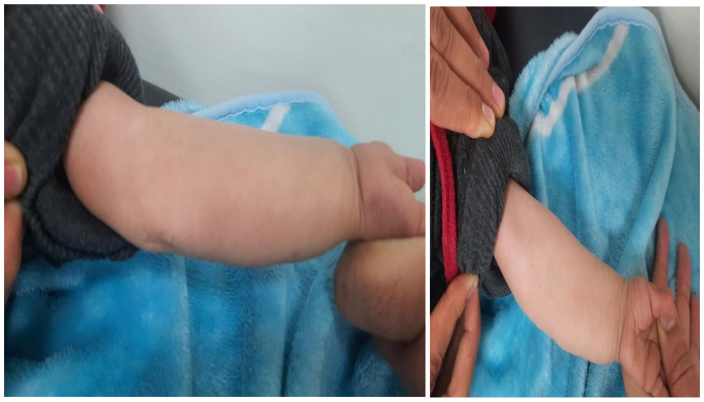

Figure 3: At 12 months of age, marked clinical regression of the left forearm mass.

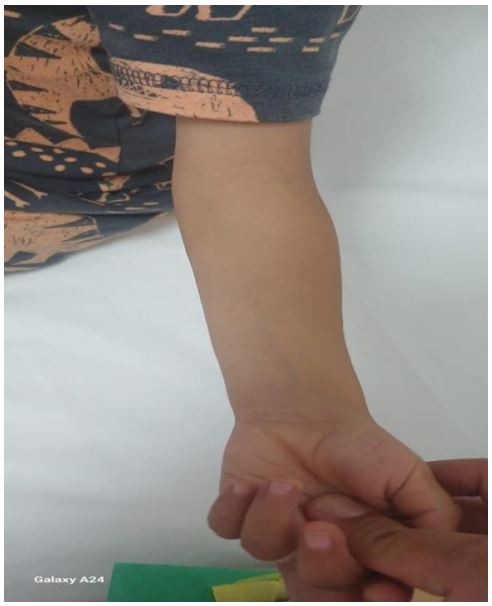

Figure 4: Recent image of the left forearm taken on June 25, 2025.

Discussion

The results of our study demonstrated a favorable response to exclusive chemotherapy in the treatment of congenital infantile fibrosarcoma of the left forearm. A clear tumor regression was observed from the early cycles, as confirmed by CT and MRI imaging, showing a progressive decrease in tumor size. At 24 months, a small stable residual mass with no metabolic activity on PET scan was documented, followed by the absence of recurrence at 3 years, indicating excellent local control without the need for surgery.

This positive response to chemotherapy, without requiring surgical intervention, is consistent with the findings of several recent studies. Demir et al. [11] showed that well-managed systemic chemotherapy can lead to significant and sustained tumor regression in patients with infantile fibrosarcoma, thereby reducing the need for mutilating surgery. Similarly, Sheng et al. [12] reported that VAC and IVA protocols are effective in inducing marked clinical and radiological responses, with good tolerance in children.

The presence of a stable residual mass without signs of metabolic activity on PET scan, as observed in our patient, is also well documented in the literature. Elmanzalawy A et al. [13] emphasize that this type of residual mass does not necessarily indicate active tumor recurrence and should prompt close monitoring rather than systematic surgical intervention, thereby avoiding overtreatment.

Furthermore, the absence of significant long-term functional sequelae in our case highlights that chemotherapy alone can preserve limb function — a crucial aspect in young, growing children. This conservative approach aligns with current recommendations aiming to minimize iatrogenic effects while maintaining optimal oncologic control [14].

However, the rarity of congenital infantile fibrosarcoma and the lack of large-scale randomized studies remain major limitations. Prospective multicenter research is needed to clearly define the indications and optimal duration of exclusive chemotherapy, as well as follow-up criteria for residual masses.

Conclusion

In conclusion, this case illustrates that exclusive chemotherapy can avoid major amputation while achieving optimal tumor control in congenital infantile fibrosarcoma, along with preservation of limb function. It is essential to ensure prolonged clinical and radiological follow-up to detect any potential recurrence at an early stage. This therapeutic approach may therefore represent a reference option for selected patients, provided that long-term, rigorous monitoring is guaranteed.

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