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An Infantile fibrosarcoma with sural nerve grafting of median nerve: A case report

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Abstract

Infantile fibrosarcoma is a disease that is characterized by rapidly progressing soft tissue tumors, mostly over the distal extremities of children during their infancy period. The management of this cancer includes chemotherapy and complete surgical resection, which has previously shown a favorable prognosis and a greater overall survival rate. This case study is an examination of a child who presented with left wrist swelling at the age of six months. After his diagnosis, the child received nine cycles of chemotherapy before surgery and operated at 18 months of age. His post-operative history was unremarkable, and he did not show any signs of relapse or metastasis. Our case is suggestive of the hypothesis that chemotherapy before surgical excision results in a good prognosis and a greater survival rate. In the future, the patient's condition should be followed-up regularly for early recognition of recurrence.

Keywords: Infantile fibrosarcoma; Case report; Sural nerve; Orthopedics; Saudi Arabia.

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1. Introduction

Congenital/Infantile fibrosarcoma is classified as a low-grade " Non-Rhabdomysarcoma Soft Tissue Sarcoma (NRSTS)" [1, 2]. The tumor is of a mesenchymal cell origin with malignant fibroblasts within a collagen background [3]. It constitutes <1% of all childhood carcinomas [1]. It is most commonly seen in the first year of life, with an incidence of 24.5% [4], but it can also be developed congenitally or during the first five years of the child's life [5, 6]. Infantile fibrosarcoma (IFS) most often appears on the patient's extremities, particularly their distal segments (70%), followed by the head and the neck [7]. It typically presents as a painless, poorly defined mass, which can come in different sizes and consistencies [8]. IFS tend to proliferate; therefore, early clinical diagnosis is often challenging and erroneous [9-11]. It has a lower incidence of metastasis (10%), and a >90% probability of long-term survival at 5 years, as well as a generally satisfactory response to chemotherapy [1, 3]. Nevertheless, the standard treatment has primarily been surgical excision, with preoperative chemotherapy being successfully undergone since the 1980s [12]. This study is a case report of infantile sarcoma with a brief and relevant literature review.

2. Case description*:*

2.1 History*:*

An eighteen-month-old baby boy came to the orthopedics clinic with the complaint of left wrist swelling. The swelling appeared shortly after birth. At the age of six months, the child underwent a biopsy and was diagnosed with infantile fibrosarcoma in the left wrist. His biopsy showed tumor cells of various morphology: spindle-shaped, cigar-shaped, ovoid, vesicular nuclei with small nucleoli and eosinophilic cytoplasm, with mixed foci of more, primitive small round cells. Variable-sized, thin-walled, and irregularly branching (staghorn configuration) vascular spaces seen. Mitotic figures were frequently visible with atypical forms. Areas of hemorrhage and necrosis were also identified. Immunohistochemical stains showed positive staining for CD99, bcl2, CD34, highlighted the vascular spaces and showed focal positive staining in the tumor cells. There was scattered positive staining with smooth muscles actin within the tumor cells. Ki67 showed a high proliferative index. However, the tumor cells were negative for desmin, AE1/3 end S100. All the changes were suggestive of infantile fibrosarcoma with near certainty, representing different stages of maturation of the same (single) entity. The patient received nine cycles of chemotherapy after diagnosis. He

had no history of pain or trauma in addition to unremarkable past medical and family histories.

2.2 General and physical examination:

The physical examination showed no swelling in any other body parts. The child's chest was clear, and the cardiovascular, abdominal, and lymph node examinations were unremarkable. The rest of the physical examination was also unremarkable. However, the musculoskeletal examination showed swelling and a biopsy scar over the volar side.

2.3 Laboratory investigations:

All laboratory investigations parameters were within the normal range.

2.4 Radiological Investigations:

X-ray left forearm:

The radiograph revealed a faint periosteal reaction in the diaphysis of the radius, which might represent osteomyelitis, a fracture or sarcoma.

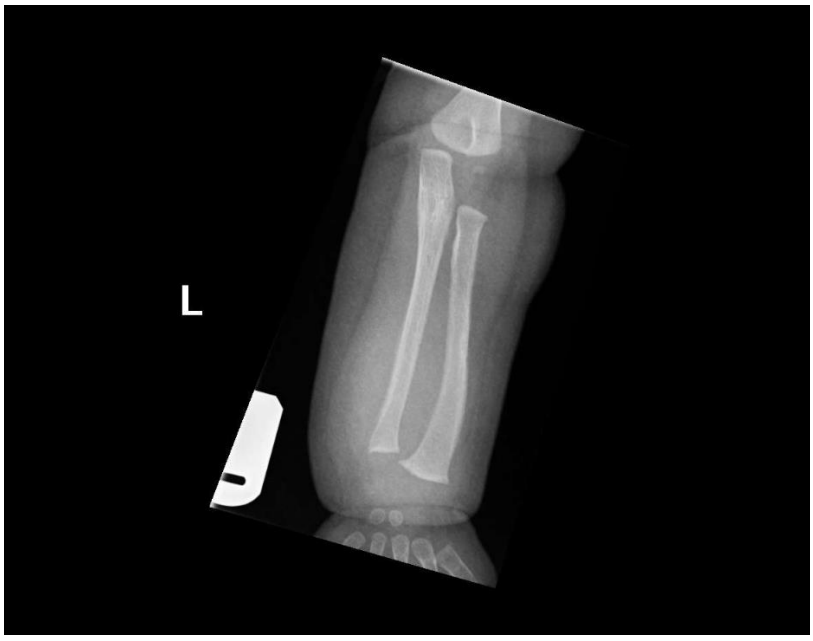


Figure 1, X-ray left forearm and wrist.

MRI forearm and wrist:

The MRI of the wrist showed an infiltrated deep mass lesion at the palmar aspect (flexor region) of the distal forearm, extending just at the level of the wrist between the flexor digitorum superficialis. It was profound at the distal forearm, which had a measurement of 5 cm in the craniocaudal dimension x 2.6 cm in the transverse dimension x1.7 cm in the anterior posterior dimension.

It extended predominantly between the flexor digitorum superficialis and the flexor digitorum profundus. It also extended around the flexor digitorum superficialis and extended superficially to the superficialis at the distal forearm. It insinuated between the bellies of the flexor digitorum profundus. The mass was closely attached to the superficial part of the flexor digitorum profundus and could not be separated from it. It partially surrounded the radial aspect of the flexor digitorum superficial. The mass was also touching the superficial part of the flexor pollicis muscles and myotendinous junction and could not be separated from them. The flexor carpi radialis tendon appeared at the radial aspect and closely adhered to the mass lesion. It completely encased the median nerve, which could not be identified at the middle of the mass lesion. The ulnar artery and nerve were passing along the other aspect of the mass, which appeared separated at the most part, except for one image where the ulnar nerve was located at the edge of the mass with no flat plane. The radial artery was at the radial aspect and appeared separated. The interosseous neurovascular bundle appeared to be separated by the deep flexor muscles. The mass mainly showed a heterogenous high T2 signal intensity with areas of low T2 and low to intermediate T1. The most distal extension was noted at the level of beginning of flexor retinaculum.

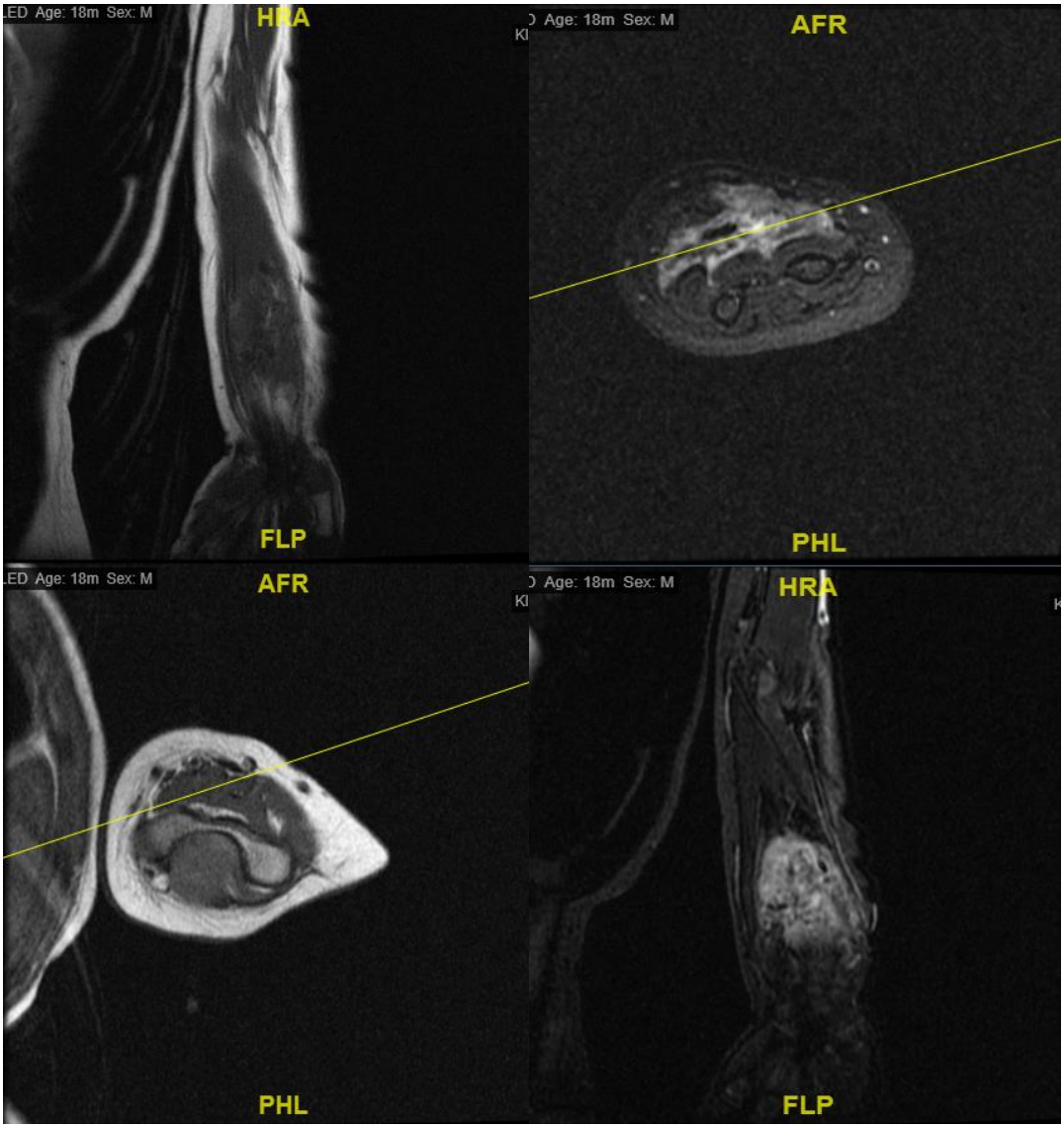


Figure 2 MRI forearm and wrist.

Bone scan whole body:

The whole-body multiple static planar, anterior, and posterior images were acquired. A baseline study showed uniform skeletal uptake. No focal abnormality was seen in the left radius or ulna bones.

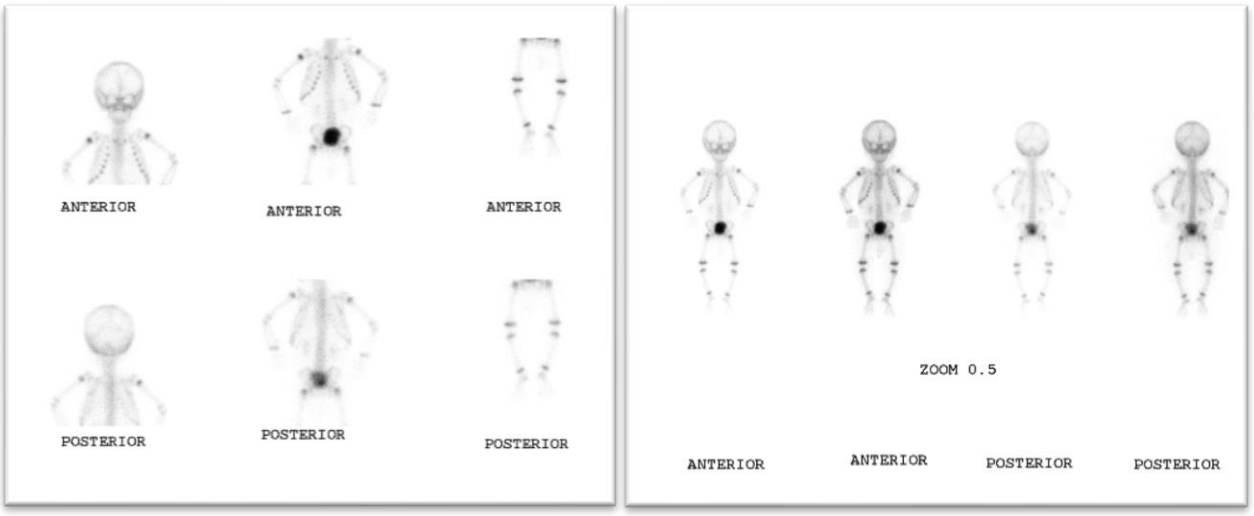


Figure 3 Bone scan.

2.5 Treatment plan:

The child's chemotherapy was completed one month before admission. Consequently, the multidisciplinary team planned a sarcoma resection of the left wrist.

Surgical treatment:

A sarcoma resection of the left wrist was performed under general anesthesia by the orthopedic team. The technique used was a volar approach with an elliptical incision. Afterwards, there was a wide margin excision which included the median nerve, Flexor Carpi Radialis (FCR), Flexor Digitorum Superficialis (FDS) and Flexor Digitorum Profundus (FDP). The radial artery/nerve and ulnar artery/nerve were then preserved. The plastic team identified the motor and accessory branch of the median nerve, and then used a sural nerve graft with fasciculations. The full-thickness flap was used to cover the wound. After the surgery was completed, the post-operative phase was uneventful. Since then, the child is having a regular follow-up and no signs of recurrence have been identified.

Excision biopsy:

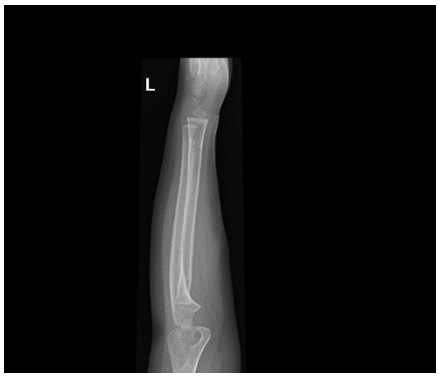
Excision biopsy showed a minimal residual tumor tissue, which was consistent with the previous diagnosis of infantile

sarcoma. Most of the tumor cell was replaced by the therapy-related changes, including hemorrhage, chronic inflammation and fibrosis. All the margins were adequately clear.

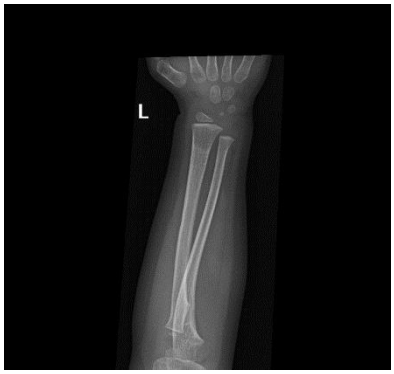
Post-operative radiological investigations:

The following investigations were performed when the child was at the age of 23 months, which was five months after the surgical removal of the sarcoma.

X-ray wrist: unremarkable.



a



b

Figure 4 (a and b) X-ray of the left Forearm.

CT chest with contrast: no metastasis

MRI wrist: Enhancement in the middle forearm reaching to the skin, most likely representing post-surgical changes.

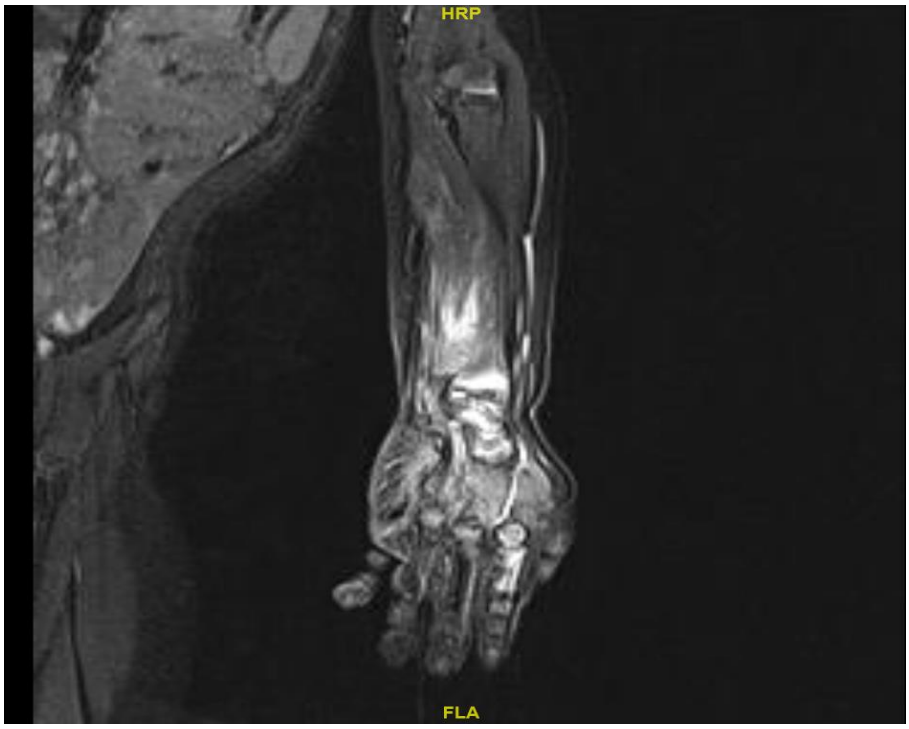


Figure 5 MRI forearm.

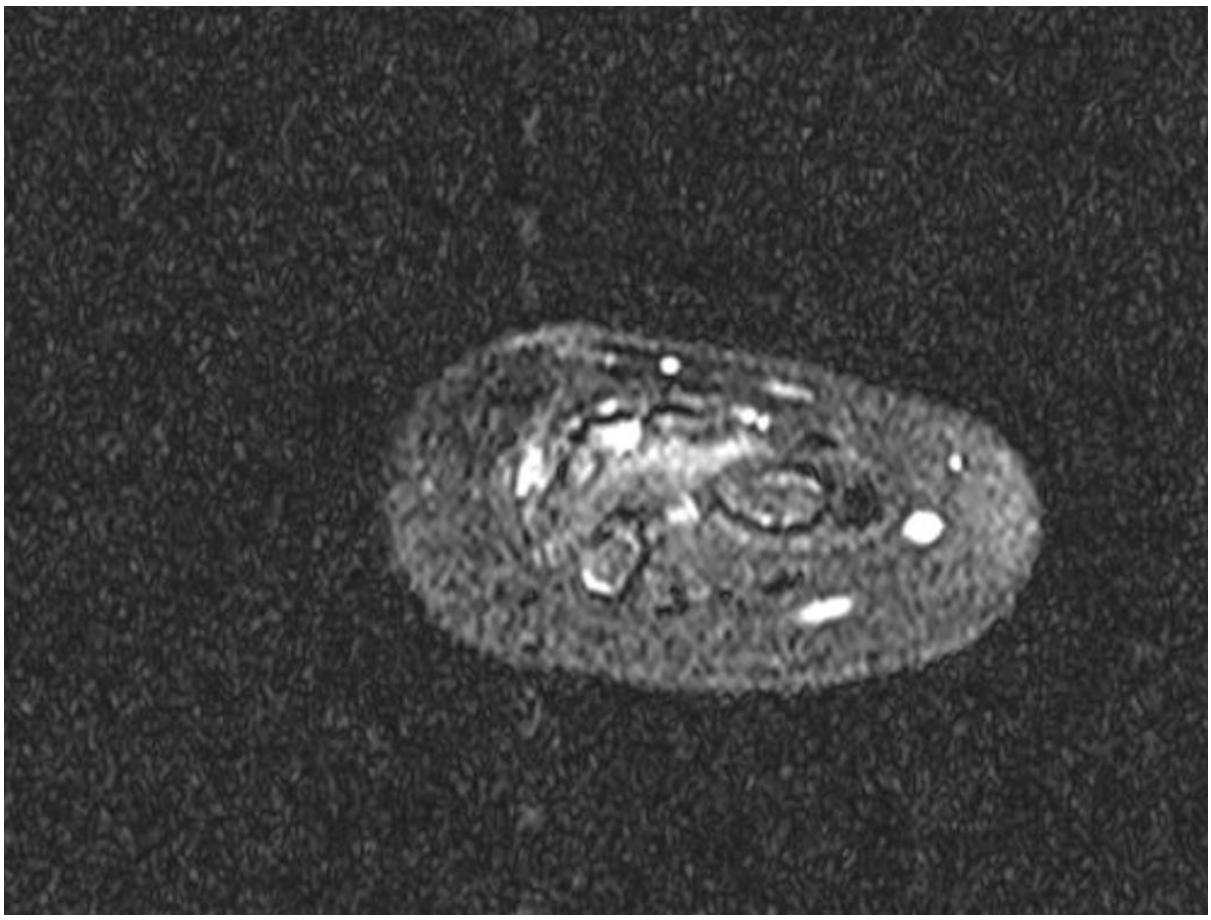


Figure 6 MRI wrist.

3. Discussion:

IFS is a rare tumor which is difficult to diagnose based on only histologic findings [2]. Clinical characteristics, age of onset, and site are also of great importance to the diagnosis of IFS [13]. The precise definition of IFS is much disputed; however, some authors suggest that it is best defined based on histologic or biologic findings. In molecular biology, the presence of *ETV6-NTRK3* is sometimes—though not always—suggestive of IFS [14]. However, in this case, a genetic analysis of the patient was not carried out.

Although the standard treatment for the last few years has been surgical excision, some patients may additionally receive chemotherapy or radiotherapy [6, 15, 16]. Similar to this case, a study of 11 patients reported that initial chemotherapy followed by surgical excision showed significant success as a treatment [17]. Furthermore, a multicenter European study suggested that chemotherapy (alkylating and anthracycline-free agent) is effective and can be used as

a first-line treatment for unresectable tumors, yet conservative surgery remains the prevailing form of treatment [16]. However, in the same study, it was stated that the initial surgical procedure is only feasible in 21% of the cases [16]. Another recent study reported an overall ~90% survival rate after chemotherapy and surgical resection [18].

The surgical approach has progressed over the past years from the primary modality to a multidisciplinary strategy and from mutilating to a more conservative approach [14]. The reported amputation rate is 50%, and it is used in cases where obtaining clear and clean surgical margins is impossible [12]. However, in locally advanced cases, surgical resection may be mutilating and can cause functional damage; in such circumstances, surgery can be planned after the reduction of the tumor size [14]. In the case of post-surgical positive margins, the first treatment is adjuvant chemotherapy, but the effectiveness is still not well-defined [17]. In a study they reported that individuals with positive post-surgical margins and not received adjuvant chemotherapy, whereas patient with unresectable tumors and received neoadjuvant chemotherapy showed good signs of long-term survival [7]. Radiotherapy is given when complete resection is impossible and is specified for axial primary sites [2].

Nevertheless, IFS has shown an overall satisfactory good prognosis, and 80% of patients are usually cured [6]. There is an estimated 90% survival rate of 10 years when using a conservative approach [14]. Children younger than five years old have been shown to be more often recurrent when compared to children who are older than ten years of age, in which case the higher risk is metastasis [19]. The local recurrence probability can be as high as 43% and usually occurs within one year after surgical excision—although it may occur at any point during one's lifetime [20]. Another study, which included 27 IFS patients, reported a 26% local recurrence rate [2].

4. Conclusion

Although it is a rare tumor among infants, it should more commonly be included in the differential diagnosis of masses that appear in a patient's first year of life, especially at birth. The overall prognosis of an IFS patient is usually satisfactory. The management of IFS mainly consists of chemotherapy (adjuvant and neoadjuvant) and conservative surgery with an estimated survival rate of 10 years. The patient should follow-up regularly for the early recognition of local recurrence.

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