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# When a Hemangioma is not a Hemangioma: Diagnostic Challenges in Infantile Soft Tissue Masses

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**ABSTRACT:** Infantile fibrosarcoma's are the most common soft tissue sarcomas in infants under the age of one, and also have an excellent survival rate. However, these tumors also have a propensity to grow rapidly, and as such, require prompt evaluation and intervention to avoid negative outcomes, such as increased local invasion, more extensive surgical resections or amputations, as well as the potential risk for distant spread. This case report describes the presentation of an infantile fibrosarcoma that was initially misdiagnosed as an infantile hemangioma. Two conditions that can present very similarly in both clinical and radiologic evaluation, but require very different treatment strategies. Through our case presentation, we aim to highlight the importance of vigilance and accuracy during diagnostic work-up to ensure swift intervention for patients with potentially sinister underlying diagnoses.

**KEYWORDS:** Infantile fibrosarcoma, Infantile hemangioma, Diagnostic vigilance, Pediatric oncology, Rural healthcare

## 1. INTRODUCTION

The infantile fibrosarcoma (IFS) is a rare malignant tumor that arises early in childhood, with an excellent overall survival rate of ~89-94%.[1] Even with its rarity, it remains the most common soft tissue sarcoma in infants under the age of one and can be present at birth.[2] These tumors often present as painless lumps that may cause pain or weakness if large enough to compress the underlying muscles or nerves. Once identified, prompt evaluation using imaging, such as x-ray, ultrasound, and/or MRI should be performed to identify the nature of the mass, and unless 100% confident that this mass is benign based on the imaging results, a biopsy should be performed. These steps are crucial in determining prompt and accurate treatment decisions.

According to the National Comprehensive Cancer Network (NCCN), the pediatric guideline for soft tissue sarcoma emphasizes the importance of diagnostic imaging being performed in the first few days after mass identification, with a core-needle biopsy being completed within days after the imaging is reviewed.[3] Therefore, the time from identifying the lesion to biopsy should typically be less than a week.

Infantile fibrosarcoma's can grow swiftly, therefore, any potential delays in diagnosis can lead to an increase in the requirement for more extensive treatments, an increase in the risk of complications, and poorer outcomes for patients.

Unfortunately, IFS can often be misdiagnosed as a benign vascular lesion and infantile hemangioma (IH). Infantile hemangiomas are the most common benign tumor of infancy, prevalent in approximately 5% of infants. They can present within the first few weeks of life, show significant growth until around 18 months, when they spontaneously involute. As these are benign in nature, they are usually not treated, unless there are specific indications;
life-threatening due to location (causing heart failure or respiratory distress), functional risks (visual obstruction, amblyopia, feeding difficulties), ulceration, or severe anatomic distortion.[4] If required, oral propranolol is the first-line treatment, often leading to rapid regression. Topical timolol may be used for small, thin, superficial hemangiomas.[5] This is in contrast to the neoadjuvant chemotherapy prior to surgery that is required for the infantile fibrosarcoma.

Through the analysis of this case, we aim to highlight the clinical and radiologic features that allow infantile fibrosarcoma to masquerade as an infantile hemangioma, contributing to delayed or missed diagnosis.

## 2. CLINICAL PRESENTATION

On 07/25/2005, a one-month-old boy was brought to the clinic by his mother for a check-up, with the complaint that he has had a

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small knot in his right wrist for the past couple of weeks. There was no history of fever, vomiting, or diarrhea, and the lesion had not been red or hot to the touch prior to today. On physical examination, there was a 3 cm x 2 ½ cm solid, raised mass on the medial right forearm. There was no redness or ulceration however, it was warm to touch.

The patient was sent for a same-day x-ray which revealed a 2.5 cm (l) x 1.4 cm (w) mass. There was no apparent bony erosion or destructive changes evident. Radiology interpretation noted that this lesion may represent a hemangioma. No calcifications were noted.

Following the x-ray, the pediatrician ordered an ultrasound to be done the next day. The ultrasound revealed a complex mass-like structure along the radial aspect of the right forearm. There was a tangle of blood flow interspersed with areas of absent blood flow. This was also interpreted as being suggestive of a hemangioma, given the rapid growth and the documented blood flow within. The differential diagnoses suggested for consideration by the radiologist included ganglion or pseudoaneurysm. However, the lack of history of any vascular penetration by any sharp objects or needle-stick effectively ruled out the pseudoaneurysm.

Unconvinced that this was simply a hemangioma, the primary care physician referred the patient to surgery for further evaluation. On 08/08/2005, the patient saw the surgeon. A physical exam revealed a bluish purple mass that was noted to be 4 cm x 3 ½ cm. There was no apparent neurovascular effect, with good distal pulses and capillary refill. The surgeon advised the mother that if this is a hemangioma as suspected, the normal course is for them to continue to grow for the first 12-18 months of life, and then they tend to spontaneously involute. The surgeon advised that because the patient is asymptomatic, a wait-and-watch approach would be best, and to follow-up in 1 month.

09/13/2005, the patient returned for follow-up with the surgeon. On a physical exam, the patient was still asymptomatic however, the mass had remarkably increased in size to 10 cm x 8 cm. Due to the rapid growth, the surgeon no longer believed observation was a viable option, and was concerned about the possible effect on the neurovascular bundle. As such, he referred the boy to a plastic surgeon to see if steroid injections would be an option to try and decrease the size of the growth.

09/26/2005, the patient had an appointment with the plastic surgeon who suggested the best treatment option would be angiography with possible embolization. The radiologists consulted disagreed and suggested that a primary investigation with MRI be carried out first.

MRI revealed a bulging soft tissue mass extending from the dorsal surface of the right forearm. There was no convincing evidence of acute bony involvement or abnormality. With the possibility of some mass effect on the dorsal surface.

Given the size of the mass, the surgeon decided to start the patient on immediate oral systemic prednisone therapy, hoping to invoke spontaneous resolution.

10/11/2005, the suspected hemangioma had continued to significantly increase in size, despite prednisone therapy. The mass continued to have fungating complications and persistent bleeding. As such, a decision was made to proceed with resection. MRI prior to surgery revealed multiple areas of radial and ulnar artery involvement. During the operation, the surgeon noted that the mass appeared more solid and less vascular in nature, and a frozen section was sent to pathology. This returned back as suspicious for a spindle cell neoplasm. The surgeon made the decision to only complete a subtotal resection, scraping the tumor off the nerves and tendons, while leaving some visible gross tumor behind. The final path report came back providing the diagnosis of a fibrosarcoma of the right forearm.

On 10/27/2005, the patient underwent a procedure for prolonged central venous access prior to beginning chemotherapy.

On 11/07/2005, prior to beginning chemotherapy, the boy was playing at home when he began “shaking all over”, his eyes rolled downward and lost eye contact. This lasted for 5-10 minutes, followed by vomiting and increased lethargy. The patient had cultures drawn and was started on ceftriaxone at an outside hospital, prior to being transferred to his tertiary care center.

11/10/2005, the patient received a neuro consult. An MRI of the head was done to assess for seizure focus or possible metastasis from his primary fibrosarcoma. No significant findings were noted on MRI. Gram negative rod sepsis was diagnosed, with three different organisms grown in 3 separate cultures. He was started on cefepime and genamicin.

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## 3. MANAGEMENT AND TREATMENT

The patient began his chemotherapy with ifosphamide and actinomycin D. He received a 5 day course with no complications. He had four total chemotherapy cycles between 11/18/2005 and 01/12/2006. No significant complications were noted during this time and his therapy was well tolerated.

On 02/15/2006, the tumor burden was reduced enough that a decision was made to resect the remainder of the tumor. The patient's mother was advised that he may lose the flexor carpi radialis tendon, as well as advising that 50% of children with this condition end up with amputation.

During the procedure, the tumor was able to be completely resected. The radial artery penetrated proximally through the mass and exited distally, as such, it was clamped and ligated. The palmaris longus tendon and a section of carpi radialis were dissected distally.

Bactrim prophylaxis was initiated and advised to be continued until 6 months post-chemotherapy. The boy was also not to receive any vaccinations until 6-months post-chemotherapy.

## 4. OUTCOME

The patient tolerated all chemotherapy and surgical procedures without complications. He is currently doing very well and has no significant issues. He has had moderate swelling and wrist pain during the past couple of years, but no tumor recurrence. He continues to remain in good health and follows with his primary care for regular check-ups.

## 5. DISCUSSION

As noted by Brockman et al., many of these infantile tumors follow a very similar clinical pattern however, they also have vastly different prognoses and treatment strategies.[6] Though IFS has a fairly low metastatic rate, they are locally aggressive and grow rapidly, so any delay in treatment allows for tumor progression and the potential for distant spread, most often to the lungs. If diagnosed early when the tumor burden is low, neoadjuvant chemotherapy with resection often has very high response rates however, diagnostic delay and increased tumor growth significantly increases the need for more aggressive surgical resection and heightened risk for amputation.[7] As such, early diagnosis and confirmation with biopsy is critical to ensure prompt intervention and prevent negative complications and outcomes.

In a review of literature, a case report identified a 6-month-old infant who was similarly worked up for an infantile lesion, which was clinically diagnosed as high probability for hemangioma. As such, a wait-and-watch approach was decided upon. As the mass continued to rapidly grow with no clinical improvement, imaging was conducted, which continued to support the diagnosis of an atypical hemangioma. It wasn't until surgery was consulted due to persistent bleeding that an excisional biopsy was completed, and congenital infantile fibrosarcoma (CIFS) was diagnosed. Though initial metastatic workup post-surgery was negative, the patient ended up having recurrent disease and pulmonary metastasis.[8] This case further emphasizes the importance of early identification and intervention for these lesions to reduce the likelihood for negative sequelae.

Infantile fibrosarcoma can potentially mimic benign vascular lesions in up to ~13% of cases.[9] On ultrasound, both masses can present with hypervascularity and both can appear with mixed echogenicity however, IFS typically has cystic components, calcifications and is usually firm and non-compressible in contrast to the infantile hemangioma.[10] The MRI characteristics between the two often heavily overlap as well. Both lesions appear isointense to skeletal muscle on T1-weighted sequences. On T2 signal, they both are hyperintense, though infantile hemangioma is slightly more so. They both show intense gadolinium enhancement, and in 83% of cases, IFS can also appear well-circumscribed, making them very difficult to distinguish from one another.[11]

It is important to keep in mind that while they do have overlapping features, IFS often show tissue invasion with infiltration of the surrounding structures and vessel encasement or displacement. Infantile fibrosarcomas will often have T1-hyperintense foci and diffusion restriction compared with the absence of these in the infantile hemangioma. While the IH typically demonstrates a homogenous enhancement pattern, a common MRI finding for IFS is the heterogeneous enhancement pattern.[12]

In our patient, no calcifications, ulcerations, or significant invasion noted on imaging. There was hypervascularity, but no distinct imaging findings that suggested malignancy, making this a convincing case for infantile hemangioma. As such, a wait-and-watch

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approach was decided upon. However, given the knowledge that these lesions can mimic each other, the NCCN recommends that unless 100% confidence can be instilled in the benign nature of the mass, that core-needle biopsy should promptly follow imaging, again, within days of identification. Our patient did not have biopsy confirmation of the fibrosarcoma until 78 days after the mass was identified. Almost 3 months after the initial presentation at the clinic, despite persistence from the primary care physician.

## 6. CONCLUSION

Although our patient's outcome was favourable and he remains tumor free, the 11-week delay in diagnosis may not have had such a positive clinical course in similar cases. While conservative management can be a great tool to prevent unnecessary invasive procedures, it is of the utmost importance that physicians remain vigilant in the workup of diseases with similar presentations, especially when the alternative is of a potentially malignant nature, with vastly different treatment strategies. This case emphasizes the importance of increased diligence during clinical evaluation and ensuring early identification and intervention are priority. Following a guideline, such as the one highlighted by the NCCN, can help ensure the frequency of misdiagnosis is reduced.

## 7. ACKNOWLEDGMENTS

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## 8. DATA AVAILABILITY

This article is freely available and may be used by all members of the scientific and medical community.

### Consent to participate

The patient is under the age of 18, therefore, verbal informed consent was obtained by the patient's parents to write and publish this case report. The informed consent was obtained by Kid Care, Department of Pediatrics.

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