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Case Report

Abdominal wall congenital infantile fibrosarcoma: A rare case report and literature review ☆

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ABSTRACT

Congenital infantile fibrosarcoma is a rare malignant soft tissue tumor, accounting for less than 1%-2% of childhood cancers. Although it can arise from any body part, an abdominal wall origin is exceptionally rare. This case report presents a case of congenital infantile fibrosarcoma originating from the abdominal wall. To the best of our knowledge, this is only the second reported case of abdominal wall congenital infantile fibrosarcoma. Unlike adult fibrosarcoma, infantile fibrosarcoma has a good prognosis with less metastasis. Early diagnosis and management are critical for improving outcomes in such rare cases.

Introduction

Soft tissue sarcomas account for 7.4% of all cancer cases in patients under 20 years of age. Among these, infantile fibrosarcoma is the most prevalent type of non-rhabdomyosarcoma soft tissue tumor, making up 24.5% of such sarcomas diagnosed within the first year of life [1,2]. Typically, it presents as a well-defined, hypervascular soft tissue mass in the extremities. However, it has also been identified in less common sites, such as the cervical region and abdominal wall [3,4]. Diagnosing infantile fibrosarcoma can be challenging due to its resemblance to more common pediatric neoplasms and its nonspecific imaging characteristics. This necessitates a high index of suspicion, especially when the tumor presents in unusual locations. Early identification is crucial for its unique management strategies and generally favorable prognosis [5,6].

There are few case reports of infantile fibrosarcoma in the literature; however, there has been only 1 report so far of an infantile fibrosarcoma arising from the abdominal wall [7]. Here

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we report a biopsy-proven case of abdominal wall fibrosarcoma in an infant who presented with progressive abdominal wall swelling since birth.

Case presentation

A 25-day-old full-term male infant presented with progressive left-sided abdominal wall swelling which was observed since birth. The mother had a regular antenatal follow-up with an uneventful delivery. Other than a small left flank mass noted at birth, which rapidly increased in size, the infant was apparently healthy.

During the physical examination, the infant displayed no dysmorphic features and all vital signs were normal. Anthropometric measurements fell within the normal range. The abdominal examination revealed a non-tender, firm, and immobile mass measuring 7 × 6 cm, protruding from the left flank covered by normal skin. Initial tests, including a complete blood count, organ function tests, and urinalysis, reveled normal results.

Ultrasound examination identified a well-circumscribed hypervascular mass in the left abdominal wall, necessitating further evaluation with computed tomography (CT). Pre- and post-contrast abdominal CT scans (Fig. 1) revealed a well-defined, homogenous mass measuring 4.3 × 4.5 × 3.9 cm with heterogeneous avid enhancement at post-contrast image. The mass caused displacement of adjacent bowel structures and splaying of mesenteric vessels but showed no clear signs of invasion. No abnormalities were detected in the liver, kidneys, or spleen.

Imaging diagnosis of infantile hemangioma with possible differential diagnosis of fibrosarcoma was considered. The

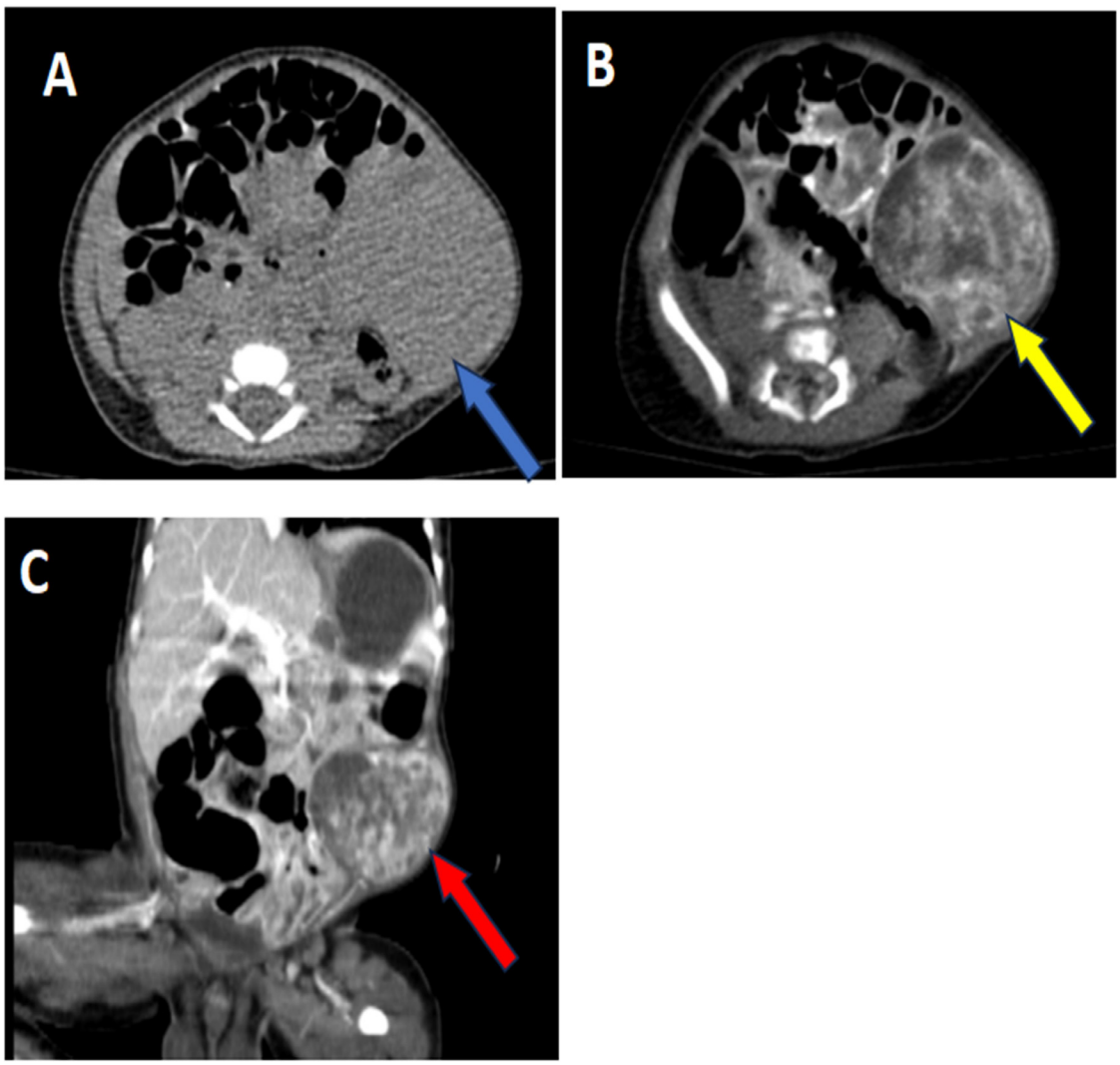

Fig 1 – Axial pre-contrast abdominal CT (A) shows homogenous mass in the left abdominal wall having soft tissue attenuation (blue arrow). Axial post-contrast image (B) shows heterogenous avid enhancement of the mass (yellow arrow). Coronal postcontrast CT (C) depicts the relationship of this mass with adjacent structures (red arrow), having mass effect on the bowel and no signs of clear invasion.

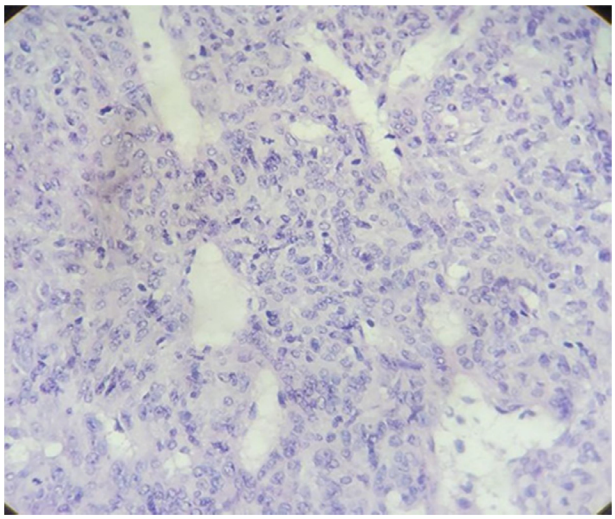

Fig 2 – Pathologic section shows tumoral tissue composed of moderately pleomorphic spindle cell proliferation with fascicle and herringbone arrangement with increased mitotic activity (8/10 HPF) along with hemangiopericytoma-like blood vessels and areas of necrosis suggestive of spindle cell sarcoma.

lesion was present from birth and the mass showed rapid growth, raising the possibility of fibrosarcoma. Following these imaging results, a consultation with the surgical team led to an incisional biopsy. It showed spindle cell proliferation with a herringbone arrangement and staghorn vessels with frequent mitosis, which confirmed a diagnosis of fibrosarcoma (Fig. 2).

The patient then began chemotherapy with vincristine and actinomycin, resulting in a reduction in mass size after 2 cycles. The plan was to refer him to a tertiary hospital for further surgical management, which was deferred by the family, and the treatment was ultimately discontinued.

Discussion

Fibrosarcoma is an intermediate-grade locally aggressive mesenchymal neoplasm of proliferating fibroblasts [5,8]. It primarily affects adults, with over 80% of the patients diagnosed above 20 years of age [9]. However, it also manifests in children under 2 years, with approximately one-third of these cases being congenital [10,11]. Fibrosarcoma accounts for 1%-2% of childhood malignancies and 13% of pediatric fibroblastic and myofibroblastic tumors. Infantile fibrosarcoma differs from its adult counterpart in pathology, cytogenetics, and clinical outcomes, despite similar histology. Prognostically, infantile fibrosarcoma is favorable, with a potential cure rate above 80% and a reported 10-year survival of 90% [12].

Clinically, fibrosarcoma is typically characterized by a rapidly growing firm mass. It most commonly appears in the extremities, accounting for about 70% of cases, and shows a predilection for the upper limbs and trunk [5,11]. Regarding gender distribution, there are inconsistencies in the literature; while some studies suggest a slight male predominance, others indicate an equal distribution [6]. Occurrences in the abdominal wall, such as in the presented case, are exceptionally rare, with only 1 other case reported thus far [7].

Various imaging modalities are employed to assess pediatric soft tissue masses, with sonography being the initial approach. Sonographic studies of infantile fibrosarcoma often reveal a heterogeneous echogenic mass, with Doppler imaging showing hypervascularity resembling high-outflow soft tissue tumors like hemangioma. Cross-sectional imaging further aids in defining these tumors and their relationship to adjacent organs, thereby guiding management decisions [8,9,11]. Magnetic resonant imaging is considered superior to CT and is the preferred modality [8,13]. It depicts a well-circumscribed, heterogeneous mass that is predominantly isointense on T1-weighted images and hyperintense on T2-weighted images, demonstrating strong post-contrast enhancement [5,8,13]. Angiography may reveal intense angiogenesis characterized by a disorganized branching pattern [7,14]. However, these findings are not specific enough to definitively diagnose congenital infantile fibrosarcoma, making biopsy confirmation essential for an accurate diagnosis [10,11]. In pathologic specimens, this tumor is characterized by a densely packed spindle cell formation with infiltrative margins. The tumor forms fascicles, displaying a distinctive herringbone pattern [6].

Infantile fibrosarcoma can often resemble vascular tumors, with differential diagnoses including rapidly involuting hemangiomas, kaposiform hemangioendotheliomas, hemangiopericytomas, lymphatic malformations, rhabdomyosarcomas, and other forms of fibromatosis. However, there are distinct clinical differences that aid in distinguishing fibrosarcoma from these vascular tumors. Unlike infantile hemangiomas, which typically appear 2 to 4 weeks post-birth and rapidly grow within the first 1 to 2 years before gradually involuting, infantile fibrosarcoma is present at birth and progressively enlarges [5,10]. On the other hand, arteriovenous malformations, like infantile fibrosarcoma, are present at birth but tend to grow proportionally with the child's growth. The growth pattern seen in our patient aligns with the description provided by Ainsworth KE et al. [6], where the mass was present since birth and exhibited rapid growth thereafter.

Surgical resection remains the primary treatment approach for patients with infantile fibrosarcoma. According to the European Pediatric Soft Tissue Sarcoma Study Group (EPSSG), chemotherapy is not necessary for approximately 70% of children diagnosed with infantile fibrosarcoma. Nonetheless, given its chemosensitivity, preoperative chemotherapy is advised for patients with inoperable tumors (Group III/R2 patients). Vincristine and actinomycin are the preferred chemotherapy agents, chosen to minimize long-term side effects [15]. Additionally, the Children Oncology Group, EPSSG, and Cooperative Weichteilsarkom Studiengruppe recommend using tropomyosin receptor kinase inhibitors for patients with metastatic disease or when conventional chemotherapy is ineffective, particularly before undertaking disfiguring surgery [16]. Generally, patients with infantile fibrosarcoma have a favorable prognosis [6].

Conclusion

This case highlights the necessity of a multidisciplinary approach and vigilant assessment in infants presenting with unusual soft tissue masses. Different imaging methods employed in evaluating infantile fibrosarcoma typically reveal features like a heterogeneous mass and marked angiogenesis. However, due to the non-specific nature of these imaging results, a biopsy is essential for a definitive diagnosis. Promptly diagnosing congenital infantile fibrosarcoma is vital, as it enables timely treatment and enhances the chances of a favorable long-term outcome.

Patient consent

We have received written informed consent from the patient's parents to publish this case report and related images.

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