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### Title

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### Permalink

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### Journal

Dermatology Online Journal, 24(2)

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### Publication Date

2018

### DOI

10.5070/D3242038167

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# Successful management of congenital/infantile fibrosarcoma presenting as large, non-healing buttock ulceration

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## Abstract

A two-year-old boy presented with a large, non-healing ulceration on his left buttock, which was originally noted as a brown patch present at birth. Punch skin biopsy was performed and histopathology revealed an atypical, pleomorphic, spindled proliferation in whorled fascicles replacing the dermis and trapping fat in the subcutis, consistent with a diagnosis of congenital/infantile fibrosarcoma. No evidence of metastatic spread was seen on imaging. The tumor was initially deemed unresectable owing to extent of local invasion. Neoadjuvant chemotherapy caused significant tumor shrinkage and the patient underwent complete resection.

*Keywords: soft tissue sarcoma, spindle cell tumor, infantile tumor, congenital tumor*

## Introduction

Congenital-infantile fibrosarcoma is the most common soft tissue sarcoma in children under the age of one. It is a malignancy of the connective fibrous tissue, which generally presents at birth or shortly after. It is most frequently found on the lower extremities and in the pelvic region. The etiology of this tumor is incompletely characterized, but chromosomal rearrangements have been implicated. It can mimic vascular neoplasms and

should be differentiated from other fibrous tumors through biopsy and histopathologic evaluation. Imaging modalities should be used to assess extent of local and distant disease as this malignancy can invade into adjacent structures, such as bone, and metastasize to distant sites. Surgical excision with wide margins is first-line treatment, but chemotherapy and radiation can also be considered.

## Case Synopsis

A two-year-old boy was referred to the dermatology clinic in Botswana for evaluation of an ulceration of the left buttock. History revealed that a brown patch had been present at birth and subsequently grew



**Figure 1.** Six-centimeter, sharply demarcated, ulcerated, round plaque with a clean base and a firm, violaceous rim on the left lateral buttock.

over the first year of life. The lesion was described as a cystic and purulent abscess when initially evaluated by a pediatrician. A series of providers managed the lesion with incision and drainage, multiple courses of antibiotics, and wound care. The lesion slowly progressed into a large, non-healing ulceration. At 18 months of age, the child was admitted to an outside hospital with a plan for surgical debridement and skin grafting, at which point a dermatology consultation was requested.

At presentation to the dermatology clinic, a six-centimeter, sharply demarcated, ulcerated, round plaque with a clean base and a firm, violaceous rim on the left lateral buttock was noted (**Figure 1**). No other skin lesions, lymphadenopathy, hepatosplenomegaly, or signs of internal involvement were observed. The child was otherwise healthy, growing and developing normally. The mother denied any history of trauma to the area. Immunizations were up to date and human immunodeficiency virus testing was negative. No significant family history was identified.

A four-millimeter punch biopsy was performed to aid in diagnosis. Histologic sections demonstrate an atypical, pleomorphic, spindled proliferation in whorled fascicles replacing the dermis and trapping fat in the subcutis (**Figure 2A**). Higher magnification highlights numerous atypical cells and mitoses (**Figure 2B**). These findings were consistent with a diagnosis of congenital/infantile fibrosarcoma. Special stains to rule out infection were negative including Ziehl-Neelsen and Periodic acid-Schiff.

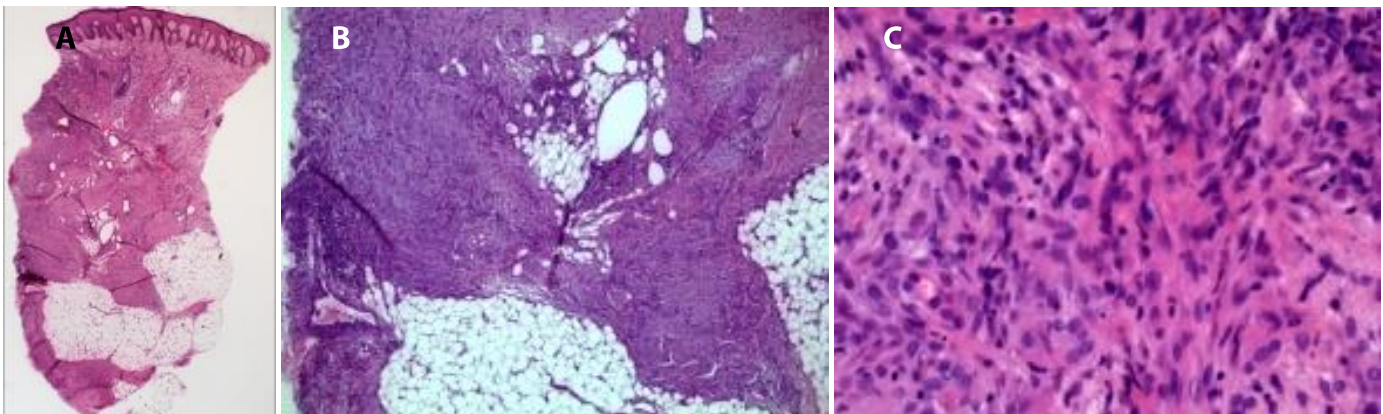
The patient underwent an abdominal ultrasound, chest X-ray, and computed tomography of the chest, abdomen, and pelvis, all of which showed no evidence of metastatic spread. Initial magnetic resonance imaging (MRI) of the left gluteal area revealed an eight-centimeter tumor infiltrating into the gluteal soft tissue and muscle without involvement of underlying bony structures. The tumor was deemed unresectable by the pediatric oncology and pediatric surgery consultants.

The patient was treated with neo-adjuvant chemotherapy of vincristine and actinomycin-D every 3 weeks for 7 cycles. At week 7, his chemotherapy regimen was modified by adding additional vincristine doses on day 8 and 15 of each cycle and by adding cyclophosphamide on day 1 of cycle 4.

Repeat MRI following cycle 7 of chemotherapy revealed significant shrinkage of the tumor and no bone involvement. The child was transferred to South Africa to undergo wide surgical excision, which achieved negative margins and was followed by skin grafting.

## Case Discussion

Congenital-infantile fibrosarcoma is a rare, rapidly growing, soft tissue malignancy that usually presents at birth or within the first year of life. According to Surveillance, Epidemiology, and End Results (SEER) Cancer Statistics Review from 1975 to 1995, the incidence of fibrosarcoma was five per one million infants, making it the most common soft tissue



**Figure 2.** Histologic sections demonstrate an atypical, pleomorphic, spindled proliferation in whorled fascicles replacing the dermis and trapping fat in the subcutis. Higher magnification highlights numerous atypical cells and mitoses. These features are consistent with a clinical diagnosis of congenital/infantile fibrosarcoma. H&E, **A**) 25x. **B**) 40x. **C**) 400x.

sarcoma presenting before the age of one year [1]. Although it is a neoplasm of the deep soft tissues, it can invade into surrounding structures, including overlying subcutaneous tissue and skin. It most commonly affects the lower extremities and pelvic area, but can also be found on the upper extremities, trunk, and the head and neck region [2].

The histopathology of fibrosarcoma is typified by a “herringbone” pattern of atypical spindle-shaped cells. Thin collagen bundles can be seen between the cells with scanty cytoplasm, elongated hyperchromatic nuclei, and frequent mitoses [2]. The genetic etiology of this malignant growth is most often a chromosomal translocation leading to the *TEL/TRKC* (also known as *ETV6/NTRK3*) fusion gene [3, 4]. However, other [TEL gene](#) rearrangements and trisomies (chromosomes 8, 11, 17, and 20) can be found within this tumor [3].

Congenital-infantile fibrosarcomas have a distinct cytogenetic profile and a much better prognosis than adult fibrosarcomas. Although congenital-infantile fibrosarcomas can be locally invasive and recurrent in up to 40-50% of cases, metastatic disease is uncommon (<10%) and survival is considered high (>90%), [4, 5]. In contrast, adult-onset fibrosarcomas have a much poorer five-year recurrence rate of 42%, a five-year distant metastases rate of 63%, and a five-year survival as low as 39% [6].

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When possible, wide surgical excision is the most favorable treatment approach [2]. However, chemotherapy also plays an important role in treatment, especially when tumor is deemed inoperable or complete excision is not achieved [3]. Radiation therapy can also be considered with caution in young children owing to retardation of growth potential [3].

## Conclusion

It is important for clinicians to be aware of the diagnosis of congenital-infantile fibrosarcomas because they can mimic vascular neoplasms, such as congenital hemangiomas, infantile hemangiomas, tufted hemangiomas, Kaposi hemangioendotheliomas, and/or vascular malformations [7-9]. Benign and malignant infantile fibrous tumors, such as infantile myofibromatosis, rhabdomyosarcoma, and dermatofibrosarcoma protuberans, may also be considered in the differential diagnosis. Histopathology is necessary to confirm the diagnosis of congenital-infantile fibrosarcoma and biopsy should be considered whenever a lesion that has developed within the first year of life displays atypical features, such as rapid growth, fixation, ulceration, and poor response to standard therapy [7].