

Prenatal Sonographic Appearance of Congenital Fibrosarcoma

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ABSTRACT: Congenital fibrosarcoma is a rare soft tissue sarcoma. A 22-year-old woman in the 22nd week of her first pregnancy underwent sonographic examination, which revealed a soft tissue swelling of the fetus's left thigh. The pregnancy was terminated, and congenital fibrosarcoma was diagnosed by pathologic examination. To our knowledge, this is the first published report of the intrauterine sonographic observation of this tumor in a fetal extremity. © 1998 John Wiley & Sons, Inc. *J Clin Ultrasound* 26:276–279, 1998.

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Congenital infantile fibrosarcoma (CIFS) is a relatively rare disease. Over the past 40 years, about 150 cases have been reported in the literature. In 1951, Andersen¹ reported 5 (3%) fibrosarcomas in her series of 175 malignant tumors in infants and children. Stout,² reviewing the literature on this subject in 1962, was able to collect 42 cases of CIFS that developed in patients 5 years or younger, including 4 (10%) cases in which CIFS was present at birth. In 1976, Chung and Enzinger³ reported 53 cases of CIFS, of which 20 (38%) were present at birth. In 1991, Coffin and Dehner⁴ reported 14 (2%) cases of CIFS among more than 900 soft tissue tumors in children and adolescents. Coffin et al⁵ later reported a series of 26 cases, of which 21 (81%) were considered congenital.

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The widespread use of sonography during pregnancy has resulted in a few reports of prenatally diagnosed CIFS. Dolkart et al⁶ reported the prenatal sonographic diagnosis of CIFS involving the mediastinum. Recently, Michigami et al⁷ reported on a fetus in which prenatal sonography showed a huge hypervascular tumor of the fetal neck, later proven to be CIFS. We report the prenatal sonographic observation of CIFS in a fetal extremity and discuss the differential diagnosis.

CASE REPORT

A 22-year-old woman, gravida 1 para 0, who had received standard prenatal care was referred to our unit for routine ultrasound examination in her 22nd week of pregnancy. An ultrasound examination at 14 weeks had been unremarkable. The examination at 22 weeks revealed localized swelling of the fetus's left thigh (Figure 1A). Further detailed examination revealed a soft tissue growth extending from the greater trochanter to the lateral epicondyle of the femur, with a maximal width 3 times that of the opposite leg (Figure 1B). The femoral shaft of the abnormal leg was slightly more hyperechoic than was that of the unaffected leg. The mass was covered by redundant skin. Color Doppler flow mapping revealed normal blood flow in both legs. Targeted organ screening revealed no other fetal anomalies.

The parents elected to terminate the pregnancy, and following oxytocin induction, a male

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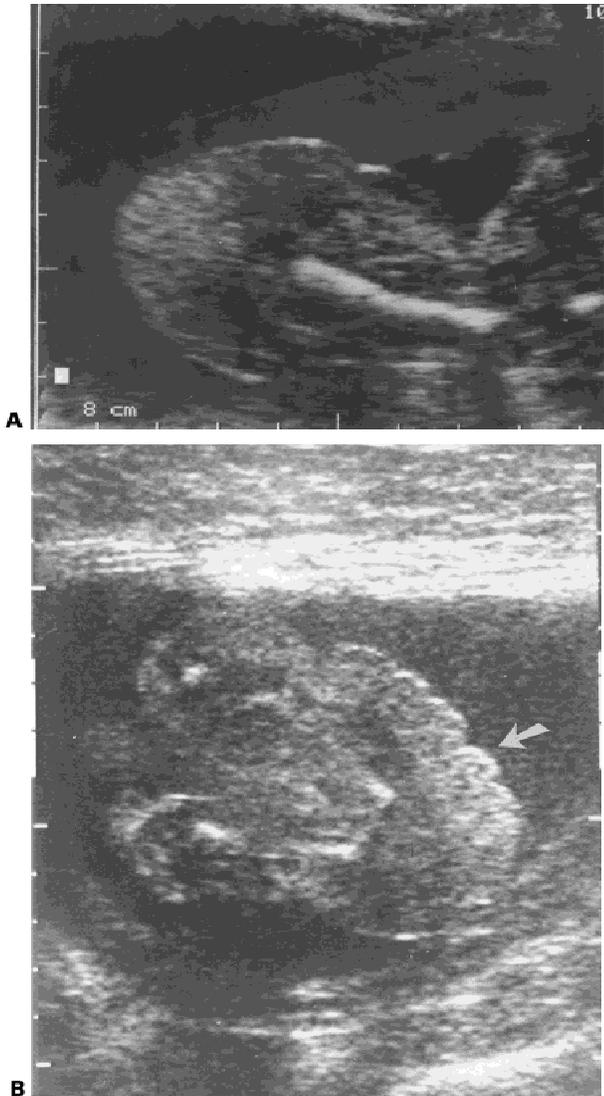


FIGURE 1. Intrauterine sonographic appearance of congenital fibrosarcoma. (A) Sonogram shows a soft tissue growth involving the left thigh. (B) Another scan of the left thigh shows swelling with redundant overlying skin (arrow).

fetus weighing 435 g was aborted. On the lateral aspect of the left leg was a 7-cm, poorly circumscribed mass (Figure 2) with a pale gray and pink cut surface. Light microscopy (Figure 3) revealed small, solidly packed, spindle-shaped cells that were separated by variable amounts of interstitial collagen. Ultrastructural examination by electron microscopy revealed fibroblast-like cells with large irregular nuclei, 1 or 2 nucleoli, free ribosomes, a well-developed Golgi complex, and a prominent and often dilated endoplasmic reticulum. The extracellular space contained scattered collagen fibers and abundant electron-dense material. The pathologic diagnosis was congenital fibrosarcoma.



FIGURE 2. Appearance of the fetus after delivery. Note extreme swelling of the left leg.

DISCUSSION

Swelling in a fetal limb may be due to any of a number of conditions, including Klippel-Trénaunay-Weber syndrome, lymphangioma, lymphedema,

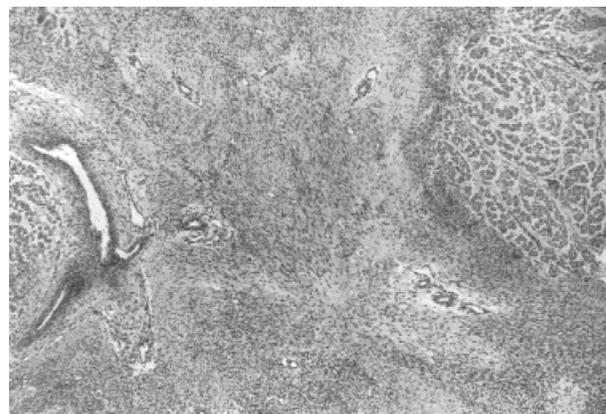


FIGURE 3. Photomicrograph of the tumor showing small spindle-shaped cells separated by interstitial collagen.

or CIFS. In Klippel-Trénaunay-Weber syndrome, also called asymmetric limb hypertrophy, the major abnormality is congenital or early childhood hypertrophy of usually 1, but occasionally more than 1, limb. The hypertrophy may coincide with an area of hemangiomas involvement.^{8,9} This syndrome could not have accounted for the findings in our case because the swelling in asymmetric limb hypertrophy involves the entire limb.

Lymphangiomas, which are benign tumors of lymphatic origin, are malformations consisting of dilated lymph channels of various sizes lined by endothelium. Generally, these tumors consist of fluid-filled vesicles or cysts of various sizes. Lymphangioma has been diagnosed in utero,¹⁰ but the characteristic appearance of hypoechoic fluid-filled areas in lymphangioma could not be confused with the sonographic findings in our case.

Congenital lymphedema occurs in 2 forms: primary lymphedema and the hereditary form, called Milroy's disease. The edema is confined to the lower extremities and is firm and easily pitted. The overlying skin is warmer than usual. Lymphedema of the legs also occurs in Turner's (XO) syndrome. Infants with congenital lymphedema may develop chylothorax and chylous ascites, which may be seen on prenatal sonography. In contrast to our case, in congenital lymphedema both extremities are affected equally.

We could find no reference in the medical literature (English and other common languages) to CIFS in a limb being diagnosed prenatally. Puzey et al reported a case of perinatal death caused by shoulder dystocia with tumor rupture and massive hemorrhage upon delivery; the tumor was subsequently diagnosed as CIFS.¹¹ Dolkart et al⁶ reported on the prenatal sonographic observation of CIFS involving the mediastinum; sonography demonstrated a homogeneous, echogenic mass posterior to the sternum. Recently, Michigami et al⁷ reported on a fetus with CIFS, which on prenatal sonography presented as a huge hypervascular tumor on the fetal neck. Because other cases of CIFS involving an extremity have been diagnosed after birth, our intrauterine sonographic observation of CIFS involving a fetal extremity is of special interest. In contrast to the findings in postnatally diagnosed cases, we observed a soft tissue growth covered by redundant skin on prenatal sonograms.

The principal manifestation of CIFS is a nontender or painless swelling or mass that can range in size from 1 to 20 cm. The most common sites of the masses are the extremities, especially

the foot, ankle, lower leg, hand, wrist, and forearm.^{2,3} Roentgenographic examination may show, in addition to the soft tissue mass, cortical thickening and curvature of the bones.

Fibrosarcoma in newborns, infants, and young children differs little histologically from that occurring in adults but must be considered as a separate entity owing to its more aggressive clinical behavior. CIFS must also be distinguished from the richly cellular, but benign, forms of fibromatosis and especially from other types of childhood sarcoma, such as embryonal rhabdomyosarcoma, which behave more aggressively.

The microscopic picture of CIFS may be confused with that of other mesenchymal neoplasms, but in most cases the uniformity of the spindle-shaped tumor cells, the solid growth pattern, and the fascicular arrangement permit a reliable diagnosis. In cases of doubt, reticulin preparations should be obtained to demonstrate the capacity of the tumor cells to produce collagen.¹²

Despite rapid growth and a high degree of cellularity, most CIFSs are cured by wide local excision or by amputation (if the large size of the tumor leaves no alternative).¹³ In the case we describe, following extensive discussion, the parents requested termination of the pregnancy based on the sonographically detected abnormalities.

To our knowledge, this is the first published report of the intrauterine sonographic observation of CIFS involving an extremity. We suggest that the possibility of fibrosarcoma be considered in the differential diagnosis of any soft tissue mass noted during fetal sonography.

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