Congenital-Infantile Fibrosarcoma of Hand – A Case Report

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Abstract
Congenital infantile fibrosarcoma is a rare malignant tumor of the soft tissue in the infant primarily developed in the soft tissue of distal extremities and occasionally in unusual location such as the lungs and retroperitoneum. It occurs mainly in children below the age of 5 years. About 250 cases have been reported in the literature so far, very few of them in newborns. Its clinical course differs from conventional fibrosarcoma, the prognosis is relatively good compared to adult forms and the 5-year survival rate can be more than 80%. We report a case of congenital infantile fibrosarcoma of the hand treated successfully by below elbow amputation. The child is now 5 years old with no recurrence.

Keywords
Fibrosarcoma; Congenital; Infantile

Introduction
Congenital infantile fibrosarcoma is an uncommon soft tissue malignancy. It usually affects the extremities [1]. Few cases involving the hand were reported and a limited number were presents during neonatal period. It represents less than 1% of all childhood malignancy and about 10% of all sarcomas [2]. Clinically there may be rapid growth of the tumor so the initial clinical diagnosis is often erroneous [3]. Although histologically similar to fibrosarcomas occurring in adults, congenital-infantile fibrosarcomas differs in their clinical behavior, metastasis is rare and local recurrence is common. The prognosis is good, much better than that for histological similar fibrosarcoma seen in adults. In one series, the 5-year survival rate for congenital-infantile fibrosarcoma was more than 80%, which is in sharp contrast to fibrosarcoma in adults, where 5-year survival rate is only 35% [4]. We report a case of congenital infantile fibrosarcoma involving the left hand in a 25 days old male infant, managed by forearm amputation and followed up for 5 years without recurrence or metastasis.

Case Report
A 25 days old baby present with painless swelling of the left hand. Parents had noticed a nodule on the palm of the left hand since birth, which enlarged rapidly. The child was treated with different antibiotic and homeopathic medicine locally. Because of rapid enlargement of the swelling, the child was referred to our hospital when he was about 1 month old.

On physical examination he was a normal healthy baby of 3 kg weight and a mass in the left hand which was poorly circumcised, measuring about 8cm x 8cm, firm in consistency, skin was tense and shiny. Other physical examination reveal no abnormality.

Plain x-ray of the left hand showed a huge soft tissue mass. The laboratory tests (CBC, PT, APPT, S. Creatinine), x-ray chest, abdominal Ultrasonography was normal. Fine needle aspiration cytology (FNAC) of the mass was done. It showed malignant spindle cell tumor. The size of tumor was rapidly increasing doubling its size.
within 2 weeks. Open biopsy was done which suggested soft tissue sarcoma. The management was an below elbow amputation and histopathological examination confirmed the diagnosis. Gross pathological specimen showed skin with underlying tumor tissue which infiltrating fat, skeletal muscle, bone and neuro-vascular bundle. It was not involving the resected margins. Cut surface was gray white with area of necrosis. Microscopic examination confirmed features of infantile fibrosarcoma with Herring bone pattern. Postoperatively the baby was stable. The child was followed up for 5 years of age without evidence of recurrence or metastasis.

Discussion

The congenital infantile fibrosarcoma is a rare soft tissue malignant tumor. Fibrosarcoma is primarily an adult malignancy and histologically similar to those seen in children. The congenital infantile fibrosarcoma usually occurs at birth or in the neonatal period, mostly presenting within the first six months of life [4]. No significant sex predilection has been found [3].

These tumors originate from the primitive mesenchymal tissue. There is no evidence of increased familial incidence but a recent study showed that all patients with congenital infantile fibrosarcoma under the age of two years exhibited some type of chromosomal gain. Most had trisomies of chromosome 8, 11, 17 and/or 20 and an abnormal karyotype 48 xy, 11 and 20 [5, 6].

Clinically, they present with a rapidly enlarging swelling in the soft tissue most commonly in the extremities (71%) but may present axial locations (29%) as well. When the limbs are involved, the tumor tends to affect the leg more than the arm and the distal extremity more than the proximal one [1]. A limited number however developed primarily in the bones [7]. Therefore the initial clinical diagnosis is often erroneous [2]. The diagnosis is sometimes made base on a fast growing soft tissue swelling. At times it is discovered late before one year [8]. On plain x-ray showed soft tissue swelling that may obliterate normal fat planes and deform or destroy the adjacent bone. CT scan is useful in demonstrating the extent of the tumor and the amount of bony involvement, where as MRI is particularly useful in showing the disruption of the soft tissue planes. The MRI findings of congenital infantile fibrosarcoma usually include a mixed cystic and solid tumor of heterogeneous density which typically has in homogenous enhancement [6]. Although the imaging findings are non specific, congenital infantile fibrosarcoma should also be considered in cases of a soft tissue mass in infants. A biopsy should be done to avoid delayed and incorrect management [9]. Antenatal and postnatal ultrasound usually shows a poorly circumscribed, heterogeneous and vascularize soft tissue mass, which grows very rapidly and cause deformity of the anatomical region involved, sometimes it causes polyhydromnious. Rest of the fetus is usually normal.

The diagnosis is done by the histological examination having anaplastic spindle shaped cells arranged in a herring bone pattern, however, some of the striking features are uniform, well oriented fibroblast, scattered round cells and chronic inflammatory cells like lymphocytes. Multinucleated giant cells are rare. Mitotic figures are common features. Rich vascular areas may be seen [10].

Although histologically similar to fibrosarcoma in adults, congenital-infantile fibrosarcoma differs in its clinical behavior compared with the adult type. Metastases are rare, however, local recurrence is common. The prognosis for congenital-infantile fibrosarcoma is extremely good with a 5 year survival rate of 84% in series of 53 cases [2]. Treatment is wide local excision or amputation. Chemotherapy and radiotherapy are reserved for unresectable tumour and for recurrence or metastasis. Amputation is indicated only in the cases of local recurrence, giant tumor involving bone and neurovascular structures [8].

In the case of our child the presence of diffuse soft tissue involvement of the hand and the bony lesions incite us to do a biopsy which revealed a well differentiated congenital fibrosarcoma. The extent of the tumor growth made us to perform an below elbow amputation of the hand. That gave us a good result because no metastasis or local recurrence were noted after 3 years however follow up should be longer to look for these evolution aspects [11].

There are high metastatic and mortality rate for axial lesions, indicating a more aggressive behaviors. The relatively benign course of such tumors may be due to a significantly lower proliferative index coupled with enhanced apoptosis. The favorable clinical course and biological features of congenital-infantile fibrosarcomas have raised a question about its nomenclature as sarcoma rather than a borderline tumor [5, 10].

In summary, although congenital-infantile fibrosarcoma is a rare soft tissue tumor in children, it should be included in the differential diagnosis of a large
extremity mass presenting at birth because the prognosis is much better than adult type. CT provides the information about bony involvement but shows poor demarcation of the extent of the tumor. On the other hand, MRI is particularly useful in delineating the extent of the tumour. It also provides multiplanar views so that diagnosis and planning for treatment can be reinforced.

References


Figure 1: Presentation

Figure 2: X-ray

Figure 3: Resected Tumor

Figure 4: Appearance of left limb after below elbow amputation