Two different clinical presentations of infantile fibrosarcoma

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We describe the histological and clinical aspects of two different childhood fibrosarcomas. Infantile fibrosarcoma occurring in an extremity is usually curable with surgical excision alone. But it is generally misdiagnosed as benign hemangioma. Therefore the tumors of the extremity must be followed-up carefully during the first months of life, as these tumors are usually present at birth. The first case, presented herein, had a huge tumor but no distant metastasis and gave no response to chemotherapy and underwent amputation. The second case who had liver metastasis was treated with chemotherapy and extremity saving surgery. Both cases are alive without any tumor. [Turk J Cancer 2000;30(2):81-85]

Key words: Childhood fibrosarcoma, chemotherapy, surgery

Infantile fibrosarcoma is an uncommon tumor of childhood. It generally occurs in an extremity and is usually curable with surgical excision alone. Although local recurrence is a common problem (17-43%), metastatic spread is rare (<10%) (1). Overall survival is given as high as 90% in different series. Different chemotherapy regimens were used with success for tumors that were unresectable for total excision or amputation or for cases with extensive metastasis (1-10).

We present here two different cases of infantile fibrosarcoma: the first one is a late diagnosed case who didn’t respond to chemotherapy and amputation was performed. The second case who had liver metastasis, was successfully treated with chemotherapy and wide resection of the primary tumor.

Case 1

A one week old boy was admitted to the hospital because of mass of about 5-6 cm in his right cruris, which was present at birth. Physical examination was normal except the mass of the right leg. An abdominal MRI, taken when he was 17 days old revealed normal abdominopelvic parameters but a right crural mass...
measuring 3.2x3.8 cm without signs of infiltration. Then a biopsy from the mass was performed when the baby was 10 weeks old and in this period the mass progressed quickly becoming a huge tumor which was dense, bluish with skin ulceration and bleeding, grossly distorting the leg (extending from heel to the groin with diameters of 27x18 cm) (Figure 1). The biopsy revealed desmoplastic type of infantile fibrosarcoma (Figure 2).

Fig 1. Gross morphology of the infantile fibrosarcoma in Case 1
Fig 2. Immature appearing fibroblasts, associated with lymphocytes; hemangiopericytomatous arrangement of short fascicules of tumor cells (HEx200)

The tumor had hemangiopericytoma like areas with high cellularity and pleomorphism, high mitotic rates and necrotic areas. The MRI of the right extremity revealed a 22x16 cm mass which was destructing the tibia and fibula and giving a heterogeneous contrast imaging. No distant metastasis was found.

Laboratory data were as follows: Hb 3 gr/dl, Hct 9%, WBC 14800/mm³, Plt 296000/mm³, PT 12.8 seconds, aPTT 37.7 seconds. Serum electrolytes, urinalysis, AST and ALT were within normal limits.

Chemotherapy was the first attempt (I²VA- CWS-96: ifosfamid 3 gr/m² days 1,2; Act-D 1.5 mg/m² day 1; vincristine 1.5 mg/m² days 1,7,14) and than surgery was performed. He was given one course of I²VA regimen but as adequate shrinkage was not noted, the tumor board decided for an upper knee amputation because limb-preservation surgery would be impossible as all of the muscles and bones of the compartment were invaded. The baby was operated when he was 3.5 months old. The surgical margins of excision were free of tumor. He is now 18 months old with no complications.

Case 2

A four months-old boy was admitted to our clinic because of a purple mass found under his left foot (extending from the ankle to the sole) since birth. This mass was misdiagnosed as hemangioma and was followed up for two months till the mass progressed. At the end of two months the circumference of the mass were 10x15 cm. The mass was planned to be sclerosed when it was extirpated because of bleeding. The pathological evaluation revealed the diagnosis of infantile fibrosarcoma (Figure 3).
Fig 3. Fascicular arrangement of fibroblasts resembling an adult fibrosarcoma, intermixed with scattered round cells (HEx200)

The family refused amputation and as RT to a large area at this age would be as hazardous as surgery, chemotherapy was planned. The clinical staging before chemotherapy revealed a mass in the liver. A CT guided biopsy was taken and malignant infiltration was noted. The other laboratory findings were normal. He was treated with EVAIA regimen for 14 courses, no RT was given. He is now a 36-months old boy without any sequelae and is off therapy since 18 months.

Discussion

Fibrosarcoma constitutes approximately 10% of soft tissue sarcomas in children. It is usually regarded as a tumor of borderline or low malignant potential, like most of the fibrohistiocytic tumors of this age. Infantile fibrosarcomas (occurring during the first five years of life) are present usually at birth. Chung and Enzinger (10) reported 41 cases in whom the tumor appearing during the first year of life and 20 cases with the tumor present at birth.

The primary therapy of fibrosarcoma is surgery. Tumor generally originates in the extremities. Despite rapid growth and large size, the majority can be cured by wide local excision or amputation.

Because the nature of the tumor in newborns and young infants is more favourable and the chance of developing metastatic spread is rather small (5–8.3%), nowadays more conservative regimens are used for therapy (3). Mortality due to infantile fibrosarcoma is given as 10.6% generally occurring three months to 11 years after initial treatment (2). No prognostic clinical or histological features have been identified for recurrence but, the ones which recurred had less radical and inadequate initial treatment. Adjuvant radiotherapy and chemotherapy are only indicated for inoperable, recurrent or metastatic cases. For two decades, the
use of chemotherapy in infantile fibrosarcomas which were unable for adequate local control has been published from different institutes (4,8,9).

In the first case the diagnosis was delayed as long as 10 weeks and during this period although the tumor became a huge mass there was no distant metastasis. We decided to use regimen IVA because surgery would be hazardous as disarticulation was obligatory. After the chemotherapy we had only 2 cm shrinkage from the upper limit with little response but there was clear necrosis with demarcation areas. Desbois et al (9) got a good result with the same regimens at the 6th day and no clinical evidence of tumour at the 25th day but the tumor they published was smaller than ours. Renard et al (8) gave VAC therapy to their three weeks old girl with a tumor 6x4 cm dimension. After four courses, conservative surgery with complete excision of the mass without any nerve or vascular damage could be performed, without serious mutilation.

In our case, the tumor was not very sensitive to chemotherapy. No other course was given as the tumour was bleeding and ulceration, necrosis were prominent on the tumour surface. Although we didn't get the same marvellous response as Debois et al. the child could have knee level amputation, not disarticulation of the hip and therefore he had the chance of having a stump which would be necessary for prothesis in his later life. In the second case as distant metastasis were detected, a wide resection was performed instead of amputation. Following surgery he was given 14 courses of EVAIA supported with growth factor (G-CSF). Despite liver metastasis, an excellent result was obtained. It is important to differentiate infantile fibrosarcoma from more aggressive malignancies such as RMS, synovial sarcoma and malignant peripheral nerve sheath tumours. There are two described forms of fibrosarcoma in children, desmoplastic and medullary types. The desmoplastic type was locally aggressive and histologically similar to fibrosarcoma of adults. The medullary type was described as being less aggressive, with a benign clinical course. But some authors described that there is not a sharp division between two types (2).

The two cases that we reported were treated with surgery and combination chemotherapy. We know that the follow-up period is short and both patients carry the risk of local or distant recurrence. The treatment choice of fibrosarcoma is still complete excision of the tumor but an initial chemotherapy can convert the surgical approach from mutilating approaches to more conservative surgery, or it can be curative for distant metastasis.

References