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Clinical management of infantile fibrosarcoma: a retrospective single-institution review

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Abstract

Background—Infantile fibrosarcoma (IFS) is an uncommon soft-tissue sarcoma. Here we review our experience treating this tumor.

Patients and methods—We retrospectively reviewed records of patients with IFS treated at St. Jude Children's Research Hospital between 1980 and 2009.

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Conflict of interest Nothing to declare.

Results—We identified 15 patients, 8 girls and 7 boys; 13 white and 2 black. Median age at diagnosis was 3 months. Primary sites included the leg ($n = 3$), chest wall ($n = 2$), foot ($n = 2$), and one each in the tongue, occipital region, axilla, parascapular region, arm, forearm, retroperitoneum, and thigh. All patients underwent resection; 11 upfront surgery, and 4 delayed. Complications included loss of the posterior tibial nerve and artery, axillary vein, biceps, pectoralis major, gallbladder, and transverse/sigmoid sinus. Eight received chemotherapy and three radiotherapy. Seven experienced local recurrence and three lung metastasis. Median follow-up was 65 months. At the time of the review, 12 patients were alive and 3 had died. All deaths were in patients older than 1 year at diagnosis with an axial primary site.

Conclusions—Non-mutilating surgery should be the primary treatment for IFS. Neoadjuvant chemotherapy is indicated when upfront resection is unfeasible. Patients with positive surgical margins should receive adjuvant chemotherapy. Radiotherapy is indicated for axial primary sites where complete resection is impossible.

Keywords

Fibrosarcoma; Infant; Retrospective study; Surgery

Introduction

Soft tissue sarcomas account for 7.4 % of all malignancies in people younger than 20 years [1]. Infantile fibrosarcoma (IFS) is the most common non-rhabdomyosarcoma soft tissue tumor with an incidence of 24.5 % of all soft tissue sarcomas seen in the first year of life [2]. It can be present at birth or can develop during the first 5 years, particularly in infants and toddlers younger than 2 years; 80 % of cases are diagnosed during the first year of life [3–5]. IFS most commonly presents as a non-tender, poorly circumscribed mass varying in size and consistency [6, 7]. Compared with the adult form of fibrosarcoma, IFS is characterized by a lower incidence of metastasis (<10 %), a higher probability of long-term survival (90 % at 5 years), and greater chemosensitivity [7–10].

Surgical excision has been the mainstay of treatment over the last few decades, but a few patients receive additional chemotherapy or radiotherapy [3, 7–9, 11]. Primary excision can entail a radical and even mutilating procedure that results in high morbidity. Recent studies have continued to advocate surgery and have added chemotherapy and radiotherapy in select conditions [3, 12]. Loh et al. [13] reviewed 11 patients with IFS and concluded that initial chemotherapy combined with surgery is the most successful treatment. A recent multi-institutional European study concluded that conservative surgery is the mainstay of treatment, and alkylating agent and anthracycline-free first-line chemotherapy should be used to treat unresectable tumors [14]. Here, we describe our experience in the clinical management of IFS at St. Jude Children's Research Hospital.

Materials and methods

The medical records of patients with IFS treated at St. Jude from 1980 to 2009 were retrospectively reviewed. This study was approved by the St. Jude institutional review board.

All patients with a histopathologic diagnosis of IFS were included. Records were analyzed for demographic details, procedural details, pathologic details, and outcome.

Results

Patient demographics and characteristics

Fifteen patients were included in our study; 8 were girls and 7 were boys; 13 were white and 2 were black (Table 1). The median age at diagnosis was 3 months (range 3 days to 36 months). The primary sites of lesions included the leg ($n = 3$), chest wall ($n = 2$), foot ($n = 2$), and one each in the tongue, occipital region, axilla, parascapular region, arm, forearm, retroperitoneum, and thigh. The primary lesion was located on the left side in seven patients and on the right side in eight patients. The size of the primary lesion was noted in 11 cases; the median size was 5.6 cm (range 2–12 cm). Signs of disease were first noticed at a median age of 2 months (range 0–28 months). All patients presented with a painless swelling.

Clinical management

Diagnostic imaging varied with the availability of modalities at the time of diagnosis. Primary lesions were imaged by plain radiograph, computed tomography, or magnetic resonance. Metastasis was detected by chest radiograph, chest computed tomography, and bone scan imaging. Metastasis was present in one patient at the time of diagnosis. An initial biopsy was done in all patients. All 15 patients underwent surgical resection; 11 had upfront surgery and 4 had delayed surgical resection. Local control included wide local excision ($n = 12$), debulking ($n = 1$), partial resection ($n = 1$), and amputation ($n = 1$). Five patients underwent re-excision of their primary lesions and two underwent a second biopsy during therapy. Of the five patients who underwent re-excision, only one with a chest wall lesion needed adjuvant chemotherapy and radiation therapy as further treatment.

For eight patients, no major nerve, vessel, muscle, or organ was sacrificed during surgical excision. The following tissues were lost in the remaining seven patients: posterior tibial nerve and artery ($n = 2$), axillary vein ($n = 1$), biceps ($n = 1$), pectoralis major ($n = 1$), gallbladder ($n = 1$), and transverse/sigmoid sinus ($n = 1$). Surgical margins were positive in six patients (patients 10 through 15); these were treated with further adjuvant chemotherapy ($n = 3$), adjuvant chemotherapy and radiotherapy ($n = 1$), adjuvant chemotherapy and re-excision ($n = 1$), or re-excision alone ($n = 1$).

Patients were treated on an institutional protocol with adjuvant or neoadjuvant multiagent chemotherapy. Four patients received neoadjuvant chemotherapy and eight received adjuvant chemotherapy. Three tumors decreased in size in response to neoadjuvant chemotherapy, as indicated by imaging studies. Neoadjuvant chemotherapy consisted of the VAC regimen (vincristine, actinomycin-D, and cyclophosphamide) for patients 7, 11, 14, and 15, and doxorubicin and dacarbazine were added for patient 7. Eight patients (patients 5, 7, 9, 10, 11, 12, 14, and 15) received the VAC regimen as adjuvant chemotherapy. Patient 5 also received doxorubicin and dacarbazine. Patient 10 received the VAC regimen alternating with vincristine, doxorubicin, and cyclophosphamide and subsequently received vincristine, ifosfamide, and etoposide. Patients 7 and 10 received initial adjuvant radiation and patient 9

received radiation for metastasis. Patient 7 received abdominal radiation (45 Gy), patient 10 received brachy-therapy (24.7 Gy) and external beam radiation (45 Gy), and patient 9 received pulmonary radiation (12 Gy).

Outcome

No recurrence of IFS tumor was reported in seven patients. Five patients (patients 1, 3, 4, 6, and 7) had only local recurrence. Two patients (patients 5 and 10) had local recurrence and lung metastasis. Patient 9 had only lung metastasis. Local recurrence was treated by amputation (patients 3 and 4), re-excision (patients 1, 5, and 6), chemotherapy and radiotherapy (patient 7), or excision, chemotherapy, and radiotherapy (patient 10).

The median follow-up period for the entire group was 65 months (range 6 months to 22 years). At last follow-up, 12 (80 %) of the patients were alive. 7 (58.3 %) had been treated by surgery alone, 4 (33.3 %) by surgery and chemotherapy, and 1 (8.3 %) by surgery, chemotherapy, and radiotherapy. None were treated with chemotherapy alone. Of the three patients who died, the causes of death were progressive lung disease ($n = 2$), progressive abdominal disease, and biliary obstruction ($n = 1$). The median duration of follow-up of the IFS survivors was 79 months.

Nine patients had no late effects of treatment. The late effects of treatment in the remaining six patients included leg-length discrepancy and leg and foot hypoplasia ($n = 1$), calcaneal valgus deformity ($n = 1$), encephalomalacia ($n = 1$), radiation pericarditis ($n = 1$), scoliosis ($n = 1$), and splenic involution ($n = 1$).

Discussion

Wide local excision, rather than radical ablative surgery, should be the initial choice of surgery in patients with IFS, as it was in 12 of 15 of our patients. A complete resection with negative surgical margins is the goal, but it is often limited by the fact that these tumors surround and invade neurovascular bundles and can replace muscle groups [15]. In this situation, delayed surgery is advocated. Amputation or disarticulation used to be indicated when obtaining negative surgical margins was not possible by a less aggressive approach [11], but it should be avoided, especially if the tumor does not involve the neurovascular bundle. The primary and secondary amputation rates have been reported to be approximately 50 % [16]. In our study, an initial conservative complete resection was possible in 4 of 15 patients. Another study showed a rate of 21 % [14]. Lymphadenectomy is usually not warranted, because IFS seldom metastasizes to lymph nodes, but two case reports have described regional lymph node metastasis [17, 18]. We did not observe any regional lymph node metastasis, except for positive omental lymph nodes in the patient with a retroperitoneal lesion.

The outcome for patients who needed only surgery as their treatment was favorable in our study. All seven patients who needed only surgery were alive at the end of the study period. This group of patients had a median age of 3 months (range 5 days to 6 months) at diagnosis. Four of these seven patients had a local recurrence that was managed with a second surgery, and none of them had a distant metastasis. Only one of these seven patients

had a positive surgical margin after an initial excision, but negative margins were achieved in this patient after a second wide local excision. It has been reported that children whose tumors are completely resected have a 100 % probability of 5-year survival, but those with residual disease after resection who receive adjuvant therapy still fare well (5-year survival, 76 %) [14].

It was previously reported that the presence of positive postsurgical margins is an important prognostic factor [19]. Positive margins were noted in six of our patients. Of these, one patient died of lung metastasis and five were alive with no evidence of disease (median follow-up, 38 months). According to our study, a positive postsurgical margin did not always indicate a poor outcome. Local recurrence or systemic metastasis was observed in one of the six patients with a positive margin. Whether chemotherapy is an effective treatment for microscopic positive margins after surgical resection is still not well defined [13]. In our study, five of six patients with a positive margin received adjuvant chemotherapy. Positive surgical margins after resection following neoadjuvant chemotherapy may not indicate a poor prognosis [20]. In unresectable cases or in cases in which major ablative surgery is the only possibility, we suggest preoperative chemotherapy to decrease tumor size, followed by resection to achieve complete resection or resection with microscopic residual disease, especially where margins involve the neurovascular bundle. Three patients with positive margins in the current study received neoadjuvant chemotherapy and all were alive at the time of the review. A recent study concluded that most patients with positive margins after resection treated without adjuvant chemotherapy and those with unresectable tumors treated with neoadjuvant chemotherapy were long-term survivors [14].

The role of chemotherapy has been described in previous reports [14, 16, 21–23]. Patients have been treated with neoadjuvant chemotherapy [13, 23] or chemotherapy alone [18, 21, 22, 24]. Neoadjuvant chemotherapy should be used for extensive lesions when complete surgical excision during an upfront procedure is not possible or would result in amputation [20, 25]. Local recurrence was observed in one of four patients in the current study who received neoadjuvant chemotherapy, but no systemic metastasis was observed in the four patients. Neoadjuvant chemotherapy has been advocated to reduce the risk of both local recurrence and metastasis [14, 26]. In five patients who received adjuvant chemotherapy for positive surgical margins in the current study, local recurrence was observed in one and systemic metastasis occurred in one. The role of adjuvant chemotherapy after initial surgery is less well defined [13, 14, 19, 23]. A previous review of the literature revealed 22 cases of IFS treated with neoadjuvant chemotherapy or chemotherapy alone. The most commonly used chemo-therapeutic drugs in that review were vincristine, cyclo-phosphamide, actinomycin-D, and doxorubicin; ifosfamide and etoposide were used to treat infants [27].

Radiotherapy of the primary site in our study was limited to partially resected tumors (chest wall and retroperitoneum); however, this modality was also used to treat lung metastasis. Radiation was administered to only one patient with a positive surgical margin, one who had a chest wall lesion, in the current study. Two groups have reported that local control was significantly better in patients with microscopic residual disease who received postoperative radiotherapy [19, 25]. Cecchetto et al. [3] administered radiotherapy to patients who were 3

years or older and had microscopic or macroscopic residual tumor. Radiotherapy is usually not advocated because it has the potential to impair growth [28].

Local relapse is more common in children younger than 5 years at diagnosis; whereas distant metastasis is more common in children older than 10 years at diagnosis [9], and the lung is the most common site of metastasis of IFS [8]. The incidence of distant metastasis in our study was 20 % and all were in the lung. The probability of local recurrence can be as high as 43 %; this usually occurs within 1 year of previous surgery, but may occur 5–31 years after the initial resection [7, 9, 11, 29–31]. Local recurrence was observed in 7 of 15 patients in the current study. It has been reported that factors associated with local recurrence included postoperative microscopic residual disease, intra-abdominal primary tumor, and the omission of adjuvant radiotherapy [19]. However, only one of seven patients in our study with a local recurrence had a positive surgical margin. Survival after isolated local recurrence is significantly better than that after distant or combined local and distant recurrence, and in most cases, death after local recurrence is caused by subsequent metastatic disease [19].

Although fibrosarcoma most commonly occurs in adults in the fourth through sixth decades, it does occur in the first two decades. Congenital or infantile fibrosarcoma, despite its similarity to the adult variant, tends to be much less aggressive with limited potential for metastasis. Children younger than 5 years at the time of diagnosis of IFS have a 7.3 % mortality rate that is due to either the primary tumor or metastasis [9]. Mortality was observed in 3 of 15 of our patients. The three patients who died in our series had axially located tumors and were older than 1 year at diagnosis (median age, 29 months). Two of these patients died from progressive metastatic pulmonary disease and one died due to progressive abdominal disease and biliary obstruction. Age is an important prognostic factor and lesions detected during infancy have the best prognosis [32]. In our study, 5 of 12 of the survivors (median age, 7 weeks) had a positive surgical margin and 4 of 12 of the survivors had a local recurrence.

It is acknowledged that chemotherapy, surgical techniques, and radiation modalities have evolved during the 30-year period covered by the current study. Moreover, this study involves a small number of patients and is retrospective in nature. However, all these patients were managed by a single surgeon over this 30-year study period. The conclusions drawn from these 15 cases and the subsequent recommendations are, therefore, tempered and incorporate the experience of others from recent literature. We recommend non-mutilating surgery as the primary modality of treatment for IFS, and neoadjuvant chemotherapy is indicated in cases in which upfront resection is not feasible. Patients with positive surgical margins should receive adjuvant chemotherapy. Radiotherapy is indicated for axial primary sites where complete resection is not possible. Due to the deleterious effects on infants, we suggest at present to closely watch them after resection and institute brachytherapy if needed. Mortality is high in children older than 1 year at the time of diagnosis of IFS and in those with axial primary sites.

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Table 1

Demographics of 15 patients with infantile fibrosarcoma

Patient number	Age, sex	Years	Location, laterality	Treatment at presentation			Mets at diagnosis	Comments, late effects	Status, duration of follow-up (months from diagnosis)
				Surgery, structure lost, surgical margins (+/-)	Chemotherapy	Radiation			
1	5 mo, M	1980	Axilla, left	WLE, left axillary vein, (-)	No	No	No	Local recurrence (excised)	NED, 122
2	3 mo, F	1980	Leg, left	WLE, posterior tibial artery/nerve, posterior compartment muscles, (-)	No	No	No	Calcaneal valgus deformity	NED, 264
3	5 days, M	1983	Foot, right	WLE, (-)	No	No	No	Local recurrence (disarticulation at ankle)	NED, 65
4	2 mo, F	1983	Arm, right	WLE, biceps, (-)	No	No	No	Right scapula recurrence (forequarter amputation)	NED, 224
5	27 mo, M	1984	Chest wall, left	WLE, (-)	Yes	No	No	Local recurrence (WLE), lung mets (chemo)	Died, 18
6	1 mo, F	1985	Tongue	Debulking, (-)	No	No	No	Local recurrence (midline glossectomy)	NED, 6
7	29 mo, F	1986	Retroperitoneum	Partial resection, gall bladder, (-)	Yes	Yes	Yes (omental LN)	Local recurrence (radioactive implants), biliary obstruction	Died, 52
8	6 mo, F	1987	Occipital region	WLE with dural graft, transverse/sigmoid sinus, (-)	No	No	No	Encephalomalacia	NED, 240
9	3 days, M	1993	Leg, right	Above-knee amputation, (-)	Yes	No	No	Bilateral lung mets (thoracotomy, chemotherapy, radiation)	NED, 240
10	36 mo, F	1994	Chest wall, left	WLE, pectoralis major/minor, (+)	Yes	Yes	No	Local recurrence (excision, chemotherapy, radiation), lung/mediastinum mets	Died, 69
11	21 days, F	2001	Leg, left	WLE, posterior tibial nerve/artery, posterior compartment muscles, (+)	Yes	No	No	Limb length discrepancy	NED, 93
12	23 mo, M	2003	Foot, right	WLE, (+)	Yes	No	No	Below-knee amputation	NED, 40
13	6 mo, M	2006	Parascapular region	WLE, (+)	No	No	No		NED, 36
14	35 days, F	2008	Forearm, right	WLE, (+)	Yes	No	No		NED, 13
15	42 days, M	2009	Thigh, left	WLE, (+)	Yes	No	No		NED, 11

Mets metastasis, WLE wide local excision, LN lymph node, NED no evidence of disease, +/- positive/negative