

## Case Report

# Giant multinodular infantile fibrosarcoma: a case report

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### ABSTRACT

Infantile fibrosarcoma is a rare type of soft tissue sarcoma seen in children usually less than 2 years of age. Few cases of giant infantile fibrosarcoma have been reported in literature. We report a rare case of a giant multi-nodular infantile fibrosarcoma in the left anterolateral chest wall in a 7-year-old boy. The tumour was said to have been recurrent twice for about 6 years. At presentation patient was evaluated and was commenced on 6 cycles of neoadjuvant chemotherapy with vincristine, adriamycin and cyclophosphamide (VAC) regimen to alternate with Ifosfamide/Etoposide (IE) regimen with very good response. Thereafter, had a wide local excision of the tumour and then had 4 more cycles of adjuvant chemotherapy. His 6 months follow up showed no evidence of tumour recurrence. Infantile fibrosarcoma is said to be chemo-sensitive tumour with very good response, though surgical excision is the main treatment of choice and overall it is said to have a good prognosis.

**Keywords:** Chemotherapy, Fibrosarcoma, Giant, Infantile, Radiotherapy

## INTRODUCTION

Infantile fibrosarcoma belongs to the group of tumours classified as soft tissue sarcomas.<sup>1</sup> They can arise from anywhere in the body. Infantile fibrosarcoma is a rare tumour that is usually congenital and most cases are seen before the age of two years though few cases can be seen between the ages of two to ten years.<sup>2,3</sup> Reports of giant infantile sarcomas are very rare and even rarer are the gross nodular presentations of such tumours.<sup>4-6</sup> Giant infantile fibrosarcomas have been reported to occur in the scalp but it is said to be commoner in the extremities and head and neck regions.<sup>6,7</sup>

A biopsy is necessary to confirm the diagnosis. Microscopically, the tumor is composed of sheets of spindle cells showing staghorn blood vessels. They are positive for vimentin, focally positive for smooth muscle

actin and negative for EMA, CD34, S100, myogenin, HMB-45, ALK-1, and desmin immunostains.<sup>2,8</sup>

CT scan and/or an MRI scan be used for imaging the tumour mass to assess the extent of tumour spread and possible distant metastasis though said to be unusual.<sup>9</sup> A wide local resection is the mainstay of treatment.<sup>10</sup> However, if initial surgery cannot be done without extensive mutilation or is not possible, preoperative chemotherapy should be given. Infantile fibrosarcoma is chemosensitive as such re-evaluation after neoadjuvant chemotherapy should be done for possible complete excision.<sup>7</sup>

## CASE REPORT

A report a 7-year-old boy, who presented to the oncology clinic with a recurrent swelling of the left chest wall of

6years duration, which was said to have gradually increased in size. It had been excised twice in the preceding years with subsequent re-growth of the swelling. There was no other swelling in any other part of the body. There was associated ulceration of the skin overlying the swelling with occasional bleeding and purulent discharge and mild pain.



**Figure 1: Picture showing patient with multinodular mass on the left chest wall, with lesion showing some ulcerative surfaces.**

On examination, there was a huge multi-nodular swelling on the left chest wall just below the left axilla, extending about 2cm centimeters from the midline to the mid-axillary line. It measured about 25x20cm in widest dimensions, firm with some cystic consistency, mildly tender and fixed to the chest wall. Other systemic examinations were essentially normal.

He was worked up and was commenced on neoadjuvant chemotherapy as a review by the plastic surgeons deemed tumour as inoperable at that time. Chemotherapy regimen was VAC (vincristine, adriamycin, and cyclophosphamide) to alternate with IE (ifosfamide, etoposide) was commenced cyclically every 3weeks for 6 courses.



**Figure 2: Picture shows patient 4 weeks post-surgery.**

After 6 courses he had a remarkable response with a significant reduction in tumour size. Chemotherapy was also well tolerated. He was evaluated and had a wide local excision with a split-thickness skin graft over the left chest wall area where the excision was done. Biopsy of resection specimen showed involved margin and reconfirmed fibrosarcoma. He had a 100% graft take with good wound healing.

He was thereafter commenced after 4weeks post-surgery with 4 more courses of adjuvant chemotherapy using the same regimen (VAC-IE). He was thereafter placed on follow up. His 6months post-treatment follow up evaluation showed no evidence of recurrence.

## DISCUSSION

Infantile fibrosarcoma is a rare tumour seen in about 1% of the population, though statistics in Africa are lacking.<sup>11</sup> It is most commonly seen in the upper or lower extremities and in the head and neck regions. The patient reported had a tumour located on the left chest wall, which is a rare site of involvement.

It is commonest among ages less than 5years old.<sup>6</sup> Though the patient was 7years of age, patient symptoms started 6years prior to presentation when he was about a year old in keeping with literature. Very few giant fibrosarcomas have been reported in literature of more than 15cm in widest dimension.<sup>12,13</sup>

Local recurrence is said to be common in patients with infantile fibrosarcoma as seen in this patient who has had a recurrence of tumour of over 6years.

Infantile fibrosarcoma is said to be chemosensitive with a good response rate to pediatric sarcoma regimen. The patient had 6 courses of cyclical chemotherapy with VAC (vincristine, adriamycin, and cyclophosphamide) to alternate with IE (ifosfamide, etoposide) regimen, with very good clinical response evidenced by a marked reduction in tumour size.<sup>14,15</sup>

Surgery is said to be the main treatment modality for the patient to achieve a cure. Thus, Patient had a wide local excision plus split-thickness skin graft following neoadjuvant chemotherapy.

Due to the positive margins noted on the microscopic review of the histological report, he was commenced on adjuvant chemotherapy (4 more courses), since he had a very good response in the neoadjuvant setting with the same chemotherapy regimen.<sup>16</sup>

The patient was not commenced on radiation therapy because of good disease control achieve with chemotherapy and also the site and extent of the tumour, the feasibility of radiotherapeutic techniques available was noted to be faltered without severe side effects.

The prognosis of the patient is good as the five-year survival of children diagnosed with infantile fibrosarcoma is about 80-100%.<sup>1,17</sup> Also, recurrence of the tumour locally is usually not associated with distant metastasis.<sup>10,18-20</sup>

## CONCLUSION

Children with infantile fibrosarcoma, a rare malignancy can be managed with surgery, which is said to be the main treatment and a harbinger for cure. The use of chemotherapy in the neoadjuvant setting to shrink tumour not amenable for excision is apt due to its good chemosensitivity. However, patients should be followed up closely due to its high rate of recurrence.

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