Case Report

Giant multinodular infantile fibrosarcoma: a case report

Aliyu M. Usman1, Okuofo C. Ehiosa1*, Okwonna O. Charles1, Abdullahi Adamu2

1Department of Radiotherapy and Oncology, Usmanu Danfodiyo University Teaching Hospital, Sokoto, Nigeria
2Department of Radiotherapy and Oncology, Ahmadu Bello University Teaching Hospital, Zaria-Shika, Kaduna State, Nigeria

Received: 01 December 2017
Accepted: 30 December 2017

*Correspondence:
Dr. Okuofo C. Ehiosa,
E-mail: docehiosa@yahoo.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

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. 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. Reports of giant infantile sarcomas are very rare and even rarer are the gross nodular presentations of such tumours. 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.

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.

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. A wide local resection is the mainstay of treatment. However, if initial surgery cannot be done without extensive mutilation or is not possible, preoperative chemotherapy should be given. Infantile fibrosarcoma is chemo-sensitive as such re-evaluation after neoadjuvant chemotherapy should be done for possible complete excision.

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...
6 years 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.

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 4 weeks post-surgery with 4 more courses of adjuvant chemotherapy using the same regimen (VAC-IE). He was thereafter placed on follow up. His 6 months 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.\(^{11}\) 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.

Though the patient was 7 years of age, patient symptoms started 6 years 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 15 cm in widest dimension.\(^{12,13}\)

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 6 years.

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.\(^{14,15}\)

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.\(^{16}\)

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%. Also, recurrence of the tumour locally is usually not associated with distant metastasis.

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.

ACKNOWLEDGEMENTS

Authors would like to thank all the members of the multidisciplinary tumour board team for their efforts in the treatment of this patient.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. The WHO Classification of Tumours of Soft Tissue and Bone (Sarcomas). Liddy Shriver Sarcoma Initiative. Available at http://sarcomahelp.org/reviews/who-classification-sarcomas.html. Accessed on Dec 1, 2017.

2. Arora K. Pathology Outlines - Fibrosarcoma of soft tissue-infantile. Available at http://www.pathologyoutlines.com/topic/softtissuefibrosarcoma infantile.html. Accessed on December 1, 2017.

3. Tarik E, Abdelouahed A, Tarik M, Hassan G, Anouar DM. Unusual case of congenital/infantile fibrosarcoma in a newborn. Afr J Paediatric Surg. 2013;10(2):185.

4. Duan S, Zhang X, Wang G, Zhong J, Yang Z, Jiang X, et al. Primary giant congenital infantile fibrosarcoma of the left forearern. Chirurgie de la Main. 2013;32(4):265-7.

5. Muzumdar D, Michaud J, Ventureyra ECG. Primary giant congenital infantile fibrosarcoma of the scalp: case report and review of literature. Childs Nerv Syst ChNS Off J Int Soc Pediatr Neurosurg. 2006;22(3):300-4.

6. Chung EB, Enzinger FM. Infantile fibrosarcoma. Cancer. 1976;38(2):729-39.

7. Orbach D, Rey A, CECCHETTO G, Oberlin O, Casanova M, Thebault E, et al. Infantile fibrosarcoma: management based on the European experience. J Clin Oncol Off J Am Soc Clin Oncol. 2010;28(2):318-23.

8. Hayek SN, Janom HH, Ibrahim A, Moran SL. Infantile fibrosarcoma misdiagnosed as vascular tumors. Hand N Y N. 2013;8(4):464-8.

9. Aga P, Singh R, Parihar A, Parashari U. Imaging Spectrum in Soft Tissue Sarcomas. Indian J Surg Oncol. 2011;2(4):271-9.

10. Ferguson WS. Advances in the adjuvant treatment of infantile fibrosarcoma. Expert Rev Anticancer Ther. 2003;3(2):185-91.

11. National Cancer Institute. Childhood Soft Tissue Sarcoma Treatment. Available at https://www.cancer.gov/types/soft-tissue-sarcoma/hp/child-soft-tissue-treatment-pdq. Accessed on 1 December 2017.

12. Vishnoi JR, Kori C, Shukla S, Gupta S, Kumar V, Rajan S. Giant fibrosarcoma of anterior abdominal wall: a rare case report and review literature. Int J Res Med Sci. 2017;3(9):2488-91.

13. Arman DM, Ekramullah SM, Mukherjee SK. A Giant Soft Tissue Fibrosarcoma of Posterior Head and Neck: A Case Report. J Natl Inst Neurosci Bang. 2017;1(2):65-8.

14. Orbach D, Brennan B, De Paoli A, Gallego S, Mudry P, Francotte N, et al. Conservative strategy in infantile fibrosarcoma is possible: The European paediatric Soft tissue sarcoma Study Group experience. Euro J Cancer. 2016;57:1-9.

15. Russell H, Hicks MJ, Bertuch AA, Chintagumpala M. Infantile fibrosarcoma: clinical and histologic responses to cytotoxic chemotherapy. Pediatr Blood Cancer. 2009;53(1):23-7.

16. Loh ML, Ahn P, Perez-Atayde AR, Gebhardt MC, Shamberger RC, Grier HE. Treatment of infantile fibrosarcoma with chemotherapy and surgery: results from the Dana-Farber Cancer Institute and Children’s Hospital, Boston. J Pediatr Hematol Oncol. 2002;24(9):722-6.

17. Orbach D, Rey A, CECCHETTO G, Oberlin O, Casanova M, Thebault E, et al. Infantile fibrosarcoma: management based on the European experience. J Clin Oncol. 2010;28(2):318-23.

Cite this article as: Usman AM, Ehiosa OC, Charles OO, Adamu A. Giant multinodular infantile fibrosarcoma: a case report. Int J Res Med Sci 2018;6:701-3.