Large Congenital Infantile Fibrosarcoma (CIF): Early Excision Versus Neo-Adjuvant Chemotherapy?

Mohd Khalid S, Abd Rashid AH, Sapuan J

Department of Orthopaedics and Traumatology, Faculty of Medicine, Hospital Canselor Tuanku Muhriz (UKMMC), Jalan Yaacob Latif, Bandar Tun Razak, 56000, Cheras, Kuala Lumpur, Malaysia.

INTRODUCTION:

We described a rare case of Congenital infantile fibrosarcoma (CIF) with consumptive coagulopathy in a 2 months old baby who presented with right arm swelling since birth. The lesion at birth measured around 2x2cm, however, at 2 months old, it rapidly enlarged to 7x7cm in size. The mass was firm, deep-seated, non-pulsatile, with vascular markings over the posterior aspect. Circulation and neurology of right hand was intact. MRI done showed lobulated intramuscular vascular lesion. The histopathological result was consistent with infantile fibrosarcoma.

The infant received preoperative vincristine and actinomycin D chemotherapy. After her second chemotherapy, she developed disseminated intravascular coagulopathy (DIVC) and bleeding from the tumour. Urgent resection of the tumour was done after pre-operative embolization of feeder vessels. Subtotal resection revealed a huge 14x14x12cm poorly circumscribed mass infiltrating the subcutaneous tissue and skin with massive intralesional hematoma. The radial nerve was entrapped inside the tumour and intra-lesion excision done to preserve the radial nerve. The child recovered well from the operation with preserved hand function.

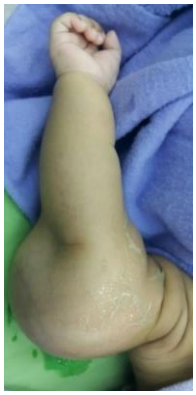

Figure 1: Huge mass involving middle and distal right arm at presentation.

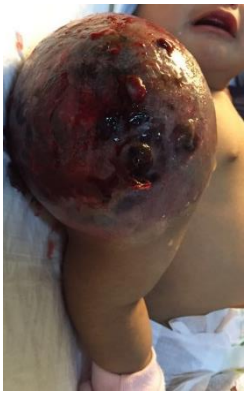

Figure 2: Huge arm mass with ulceration and bleeding during coagulopathy.

DISCUSSIONS:

CIF is the most common soft tissue sarcoma under 2 year of age and is identified by the t(12;15) translocation leading to gene fusion ETV6-NTRK3[1]. CIF is histologically indistinguishable from the adult fibrosarcoma, however, CIF has better prognosis, a lower rate of metastasis, and a high 10 year survival rate (89-90%) as compared to the adult patients whose 5-year survival rate does not exceed 50%[2]. CIF incidence is very low and is estimated to be 5 per million infants[2].

Surgery is the mainstay of treatment, and wide resection represents adequate treatment strategy in most patients. However, as CIF is generally regarded as a chemosensitive tumour (complete remission could be achieved with chemotherapy alone), surgery need to be proposed only if can be done simply without mutilation[3]. Chemotherapy is the initial treatment in cases of inoperable tumours (preoperatively) to permit tumour shrinkage and the subsequent surgery.

Salman M. et al[4] reviewed 21 reported published cases of patients with anaemia, bleeding, or coagulopathy upon presentation of CIF. They found that all the reported cases were diagnosed within the first 2 months of life and 4 presented perinatally with hydrops fetalis and died. Anaemia was present in all patients with 6 patients had coagulopathy resulting in death in 2 patients due to bleeding complications. The remaining 15 children did well : 1 patient received only chemotherapy while 14 underwent surgical resection of the tumour, and 6 of the 14 also received chemotherapy. Of note, 6 of the 14 surgical procedures consisted of amputation.

CONCLUSION:

The primary treatment is surgical resection, and chemotherapy is used in non-resectable tumours [2].

REFERENCES:

1. 1. **Knezevich SR**, McFadden DE, Tao W et al: A novel ETV6-NTRK3 gene