Journal of Pediatric Sciences

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Journal of Pediatric Sciences; 2010; 4; e58

How to cite this article:

Bhuyan, P., Mahapatra, S., Mahapatra S., Sethy, S., Das, A. Congenital infantile fibrosarcoma: a rare case report. Journal of Pediatric Sciences 2010;4: e58

CASE REPORT

Congenital infantile fibrosarcoma: a rare case report

Pallavi Bhuyan, Sitaram Mahapatra, Smita Mahapatra, Sudha Sethy, Ajit Das

Abstract:
Congenital infantile fibrosarcoma is a rare soft tissue neoplasm most commonly involving the extremities and are usually present at birth. These tumors are notoriously misdiagnosed at birth as either hemangiomas or lymphatic malformations. Although histologically similar to fibrosarcomas occurring in adults, the congenital lesions differ in their clinical behavior; metastases are rare, local recurrence is common, and prognosis is good with wide local excision combined with chemotherapy. We herewith report such a rare case with review of the recent literatures.

Key words: infantil fibrosarcoma
Received: *28/08/2010; Accepted: 22/09/2010*

Introduction

Paediatric soft tissue sarcomas constitute 7% of all the childhood tumors [1]. The comprehensive statistical analysis of the Asian and the world medical literature shows that CIFS is an extremely rare soft tissue sarcoma seen is early childhood. Only four cases of childhood fibrosarcoma are reported from the Indian subcontinent [2]. We report herewith such a rare case of 6 weeks old female infant.

Case Report

A 6 weeks old female infant was noticed to have a painless swelling over the right leg since birth. The mother had a prolonged labour due to the mass and the baby failed to respond to antibiotics. The baby was brought to the hospital due to the rapid growth of the mass.

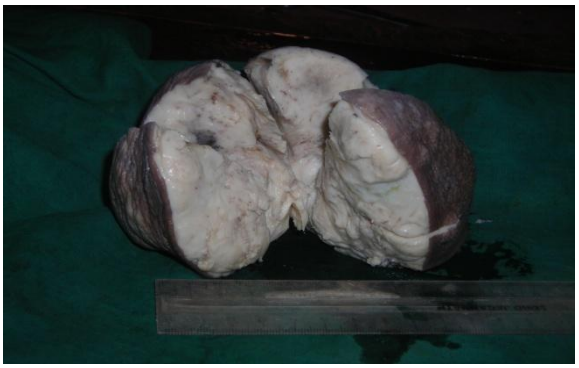
On examination, a firm to hard mass of 12×10cm, with shining skin, prominent blood vessel was seen on the antero medial aspect of right thigh. X-ray of leg showed a soft tissue mass. Doppler USG revealed a solid mass with gross vascularity. Excision of the mass was undertaken with ligation of the feeding vessels. Post operative recovery was uneventful with primary healing and vascularity of the foot was not compromised. Grossly the tumour was a poorly circumscribed mass (m) 8×7×6 cm, soft to firm in consistency with greyish white cut surface with areas of hemorrhage and necrosis (Figure 1). Histopathological picture revealed spindle shaped cells arranged in long fascicles and herring bone pattern (Figure 2) and showed minimal pleomorphism with frequent mitotic figures along with scattered lymphocytes (Figure 3). The reticulin stain showed fibres around individual tumour cells. Immunohistochemically cells are strongly positive for vimentin (tumor marker for cells of mesenchymal origin) and negative for desmin (tumor marker for smooth muscles) supporting the diagnosis of fibro sarcoma (Figure 4).

Pallavi Bhuyan, Sitaram Mahapatra, Smita Mahapatra, Sudha Sethy, Ajit Das

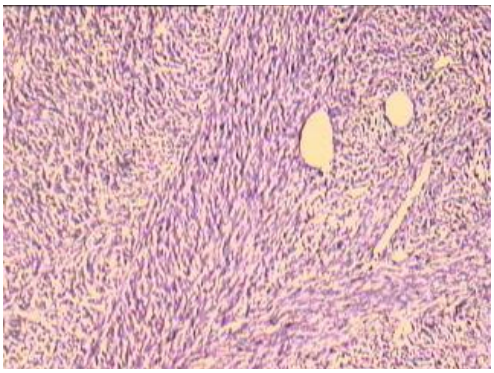
Department of Pathology, S.C.B. Medical College, Cuttack, INDIA

Corresponding Author:
Pallavi Bhuyan, MD

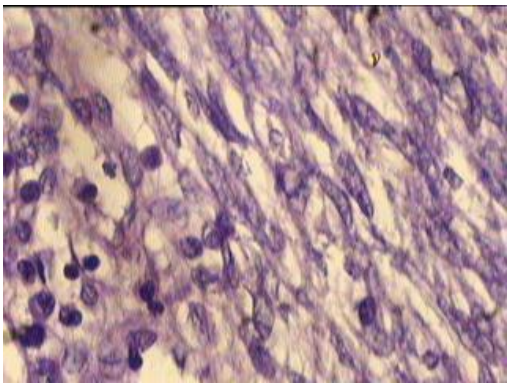
Department of Pathology, S.C.B. Medical College, Cuttack, 753007, Orissa, INDIA
E-mail: pallavibhuyan@sify.com



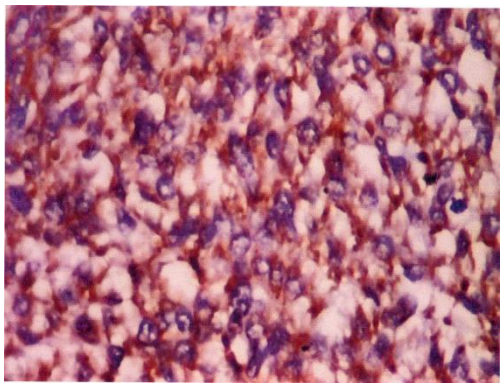
(Figure 1)



(Figure 2)



(Figure 3)



(Figure 4)

Figures 1-4. Gross Photograph showing solid soft tissue grey white tumour (fig 1), Photomicrograph reveals spindle shaped cells arranged in long fascicles and herring bone pattern. (HE x100) (fig 2), Photomicrograph showing minimal pleomorphism of the spindle cells along with scattered lymphocytes. (HE x400) (fig 3) and Immunohistochemically cells are strongly positive for vimentin (fig 4).

Discussion

Paediatric soft tissue sarcomas account for 7% of all childhood tumours. CIFS is a rare soft tissue malignancy with 300 reported cases in world literature. CIFS is a relatively rare tumour first described by Stout in 1962 [3]. This tumour mainly involves the extremities (72%), head and neck region and orbit [4-6]. Rarely retroperitoneum & mesentery may be involved. The age groups affected are infants & young children under the age of five (congenital variety), and children between 10 and 15 years of age.[7] A characteristic chromosomal translocation of t (12,15) (p13,q25) and the fusion transcript ETV 6-NTRK3 in the primitive mesenchymal cells results into the genesis of CIFS. RT-PCR analysis with positive ETV6-NTRK3 fusion gene differentiates all other spindle well lesions from CIFS. Additional chromosomal mutations like monosomy 19 & 22, del(2), t (2:11) (q37:13) may also be noticed in few cases. Histologically, the tumour bears a close resemblance to fibrosarcoma in adults, having anaplastic spindled shaped cells arranged in herring bone pattern, with areas of uniform, well oriented fibroblasts, scattered round cells and lymphocytes. Multinucleated giant cells are rare. Mitotic figures are common. This tumor needs to be distinguished from embryonal RMS and aggressive infantile fibromatosis. CIFS has an excellent prognosis (5yr survival 84%)? This benign behavior may be due to the combination of lower proliferative index coupled with enhanced apoptosis. In CIFS flow cytometry reveals a moderate to high proliferation activity and a diploid DNA content. Treatment of choice is wide local excision. Chemotherapy and radiotherapy are reserved for unrespectable aggressive tumours and for tumours having metastasis or recurrences. Amputation should be reserved for chemoresistant

patients in whom the involvement of neurovascular structures by the tumour make a limb-sparing aggressive excision impossible. CIFS is usually a deep seated soft tissue lesion showing herring bone pattern of arrangement of spindle cells in absence of whirling pattern which distinguishes this from infantile aggressive fibromatosis. Desmin negativity differentiates these lesions from embryonal RMS, infantile hemangiopericytoma, lipoblastomatosis teratoma, lymphangiomatosis and fibrous histiocytomas. Few centers have also advocated a pre-operative Chemotherapy (VAC regimen) before undertaking the conservative surgery. Regardless of its histologic grade metastatic spread is uncommon. Regional lymph node spread is rare but hematogenous spread can occur with metastatic deposits occurring commonly in the lungs, bone marrow or abdominal organs. The tendency to local recurrence is high when initial treatment is not radical or when vital structures are involved.[9,10] Recurrence up to 17 years after initial surgery is known.

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