CASE REPORT

Chest Wall Infantile Fibrosarcomas- A Rare Presentation

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Abstract Infantile fibrosarcoma is rare and represents less than 1 % of all childhood cancers. Commonly, it arises in the limbs followed by the trunk and head and neck. We present a rare case of infantile fibrosarcoma of trunk at two different sites in a newborn with brief review of the relevant literature.

Keywords Fibrosarcoma · Childhood cancers · Infantile fibrosarcoma · Chest wall tumors

Introduction

Infantile fibrosarcoma (IF) is a rare malignant soft tissue neoplasm composed of uniform, cytologically malignant spindled fibroblasts [1, 2]. It represents less than 1 % of all childhood cancers, and it is considered the most common soft tissue sarcoma in infants [3–5]. Most of these tumors are discovered at birth (30 %) or in the first year of life, and presents a diagnostic and therapeutic challenge. It often arises in the arms or legs but may also arise in the head, neck, trunk, and rarely at other sites [3–10]. This report describes an unusual case of IF of the chest wall chest in a newborn with brief review of the relevant literature.

Case Report

A 25-day-old male neonate weighing 3.7 kg was referred to the hospital with a progressively increasing mass over chest since birth. Initially, it was covered by skin, which sloughed away gradually 10 days after birth. On examination, a chest wall mass measuring 10 × 8 cm in size was present at right mid axillary line, which was extending both anteriorly and posteriorly. The overlying skin was ulcerated through which fleshy mass was visible (Fig. 1). The mass was of normal temperature, nontender, solid, firm in consistency, and mobile over the chest wall. The surrounding veins over the chest wall were dilated. Another tumor, measuring 4 × 3 cm, was present on left side in the mid axillary line. It was having the same clinical features as described above except for the presence of intact overlying skin.

Contrast enhanced computerized tomographic (CECT) scan of thorax revealed nonenhancing, tumor of size 10 × 8 × 5, confined to the chest wall without intrathoracic extension or ribs infiltration (Fig. 2).

The patient was operated upon under general anesthesia. Intraoperatively, a large tumor was present in the pectoral area, which was extending from midclavicular line to posterior axillary line, from fifth intercostals space (ICS) to eighth ICS. The tumor was mobilized and excised in toto, along with part of the pectoralis major muscle. The surrounding skin was mobilized carefully and primary wound closure was accomplished. In the postoperative period, there was superficial wound dehiscence, which was managed conservatively. Patient was discharged with proper advice and follow up. The histology showed malignant proliferation of spindle-shaped cells, which were arranged in fascicles. Immunohistochemistry showed the tumor strongly positive for vimentin, but it was negative for smooth muscle actin, myosin, and desmin (Fig. 3). The skeletal muscles were invaded by the tumor but margins

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Fig. 1 Clinical picture showing tumor of the right side. The overlying skin is eroded and engorged veins are surrounding it

were free. The features were consistent with infantile fibrosarcoma.

The tumor of the left side was operated after one month. It was fixed to the lateral border of the scapula. The border was excised along with the mass. It was followed by the muscular reconstruction. Postoperative period was again uneventful. Histopathology showed the same characteristics as described above. The patient has been in regular follow up for last eight months and has not developed any recurrence up till now.

Discussion

IF, also called congenital fibrosarcoma, is a tumor that arises from mesenchymal cells. It is composed of malignant fibroblasts in a collagen background [1, 2, 4]. In contrast to the adult fibrosarcoma, IF is considered to be a rare and low grade malignant neoplasm. It is locally aggressive with an increased rate of local recurrence but systemic metastasis is very rare. Most IFs are diagnosed postnatally, but there are increasing reports of antenatally diagnosed cases [11, 12]. There appear to be no advantage to preterm delivery of these patients except

if there is associated hydrops. Delivery by cesarean section is indicated only in those with large tumors that are likely to obstruct delivery.

Fibrosarcomas are usually located in the subcutaneous tissues and develop in different parts of the body, but they are most commonly seen in the extremities. Other less common sites include trunk, head and neck. IF, arising in the head and neck, is commonly seen in older children. Other rare sites of origin include the mesenteries, retroperitoneum, mouth, presacral region, intestines, and lungs [3–10]. Its principal manifestation is a painless swelling with steady, sometimes rapid growth. Males are affected slightly more than females. Histopathologically, it should be differentiated from other tumors including infantile fibromatosis and myofibromatosis [1, 2]. It is a highly cellular tumor, composed of spindle cells, which are arranged in bundles or fascicles. The tumor cells are diffusely positive for vimentin and may show focal positivity for actin but are negative for S-100, desmin, and myoglobin. The diagnosis of soft tissue tumors poses a challenge to the pathologist and sometimes necessitates the use of further tests including histochemical staining, immunohistochemistry, and electron microscopy. Cytogenetic analysis of IF shows trisomy 11 being the most important finding, followed by trisomy 20. In another study, cytogenetic analysis identified a novel t (12;15) (p13, q25) translocation involving the ETV6/NTRK3 gene [13–15].

IF is a slow growing tumor having good prognosis, which differentiates it from adult fibrosarcoma. Metastasis is rare and occur mostly in the lungs, bones, and sometimes in lymph nodes; however, local recurrence is rather high and occurs in 20 % to 40 % of cases [4–7]. These patients should be followed up closely and monitored for local recurrence, which can occur as early as 6 weeks postoperatively and as late as 10 years after the initial excision. Although there are reports of spontaneous regression and disappearance of infantile fibrosarcoma, the accepted treatment is wide local excision. Adjuvant chemotherapy is recommended for those patients with unresectable tumors of axial skeleton and proximal extremities. The wide local excision appears to be the treatment of choice unless the size of the tumor and its anatomical location require amputation. As late recurrent and metastatic lesions may be encountered, long-term follow-up is necessary before one can safely assume that the patient has been cured.

On clinico-radiological basis, wide local excision appeared to be feasible; hence, it was performed. This also saved the time in process of incisional biopsy and waiting for the report for definitive surgery. We could have performed the simultaneous excision of both masses; however, given the age of the patient and the nature of the operation, we believed it to be more hazardous. Also, as the overlying skin was intact, it was not an immediate concern in comparison to the side with skin necrosis. Besides, simultaneous excision may have compromised the skin covering for both the defects.

Fig. 2 CT scan showing tumors of both right and left side

Fig. 3 Photomicrograph showing **a** Sheets of plump cells showing high nucleo-cytoplasmic ratio ($\times 125$). **b** Strong expression of vimentin ($\times 400$). **c** Negative expression of desmin ($\times 400$). **d** Negative expression of s100 ($\times 400$)

To the best of our knowledge, this is the first report where IF presented as two separate masses over the trunk in the neonatal period.

In conclusion, although IF is a rare soft tissue tumor in the pediatric group, it should be included in the differential diagnosis of a large mass presenting at birth. Imaging provides good information about the extent of the tumor. Wide local excision is the treatment of choice, and these patients should be followed up closely for the possibility of local recurrence.

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