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Chest Wall Rhabdomyosarcoma in Newborn- The Rarest of Rare Site in a Newborn: A Case Report

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Abstract: Soft tissue tumours are rare in childhood with most common being benign low flow vascular malformation (Heamangiomas). Neonatal soft tissue malignancy is rare. Chest wall as the primary site has been rarely reported. The aim objective is to report and to add to the existing literature a case of Neonatal chest wall RMS. A neonate born with chest wall tumour was successfully treated with in toto excision. Last follow up appeared after 8 years. The child was disease free. The conflict between alveolar Rhabdomyosarcoma (RMS) and Malignant Mesenchymal Tumour (MMT) was resolved by immunohistochemistry with vimentine and desmin being positive and cytokeratin and S100 being negative. Hereby, rarest of the rare case report of chest wall RMS is being reported.
Keywords: Neonatal chest wall tumour, Neonatal soft tissue tumours, Neonatal malignant meseenchymal tumours, Neonatal rhabdomyosarcoma.

INTRODUCTION

Soft tissue tumours of newborn seen commonly are benign, vascular [1]. The malignant tumours of soft tissue described range from fibromatosis, myofibromatosis, rhabdomyosarcoma and neuroblastoma [2]. In the pediatric age group Rhabdomyosarcoma (RMS) has been reported as the most common soft tissue sarcoma. Primary tumor site RMS is an important prognostic determinant [3]. The other prognostic determinant includes age of the patient, size of the tumour and histologic finding [4].

The most typical sites of RMS are head and neck region and the genitourinary system [5]. Chest wall rhabdomyosarcoma is a rare entity [6]. Cases of chest wall RMS have been reported in literature [3, 4, 6].

The aim and objective is to report and to add to the existing literature a case of Neonatal chest wall RMS.

CASE REPORT

A full term otherwise healthy newborn was referred immediately after birth with a chest wall tumour. Child weighed 3kgs and had all normal neonatal reflexes with good cry and no respiratory distress. Examination revealed a horizontally oblong ,oval, pyriform shaped mass over the anterior chest wall 2cm below the manubrium extending both sides of chest wall. It was firm in consistency, mobile over the chest wall,skin was stretched with visible redness and cutaneous telangiectasia at one place on surface (Fig. 1a). The tumour was bilobular, right component very close skin (Fig.1b). There were no satellite nodules, no palpable lymph nodes in both the axillae and on both sides of the neck. No other organ was palpable on abdominal examination. Examination of chest was unremarkable. Chest radiograph revealed soft tissue mass without involvement of ribs, no pulmonary metastasis and no evidence of pleural effusion (Fig. 1c, d) An ultrasonography of chest wall (Fig. 1e, f)showed mixed echogenic lesion with scattered areas of reduced echogenecity. Serum alpha fetoproteins were raised to more than 1750 ng/ ml. The tumour was excised in situ, in toto along with ellipse of skin and skin was closed with local advancement flaps (Fig. 2a). Post-operative chest radiograph was normal (Fig. 2b). Post-operative period was uneventful. Excised specimen showed solid nature of tumour (Fig. 2c, d). Child was lost to follow up after first visit after a month. 8yrs later child showed up with a scar showing no evidence of disease.

Histopathology report revealed a predominantly small cell tumour with occasional spindle cells (Fig. 2d).

Differential diagnosis for a long time was between rhabdomyosarcoma of alveolar type and undifferentiated fibromatosis. Immunohistochemistry done for vimentin, desmin were highly positive. Cytokeratin was negative. S100 was negative (Fig. 2f, g). Thus the diagnosis of rhabdomyosarcoma in newborn on chest wall was confirmed by consensus.

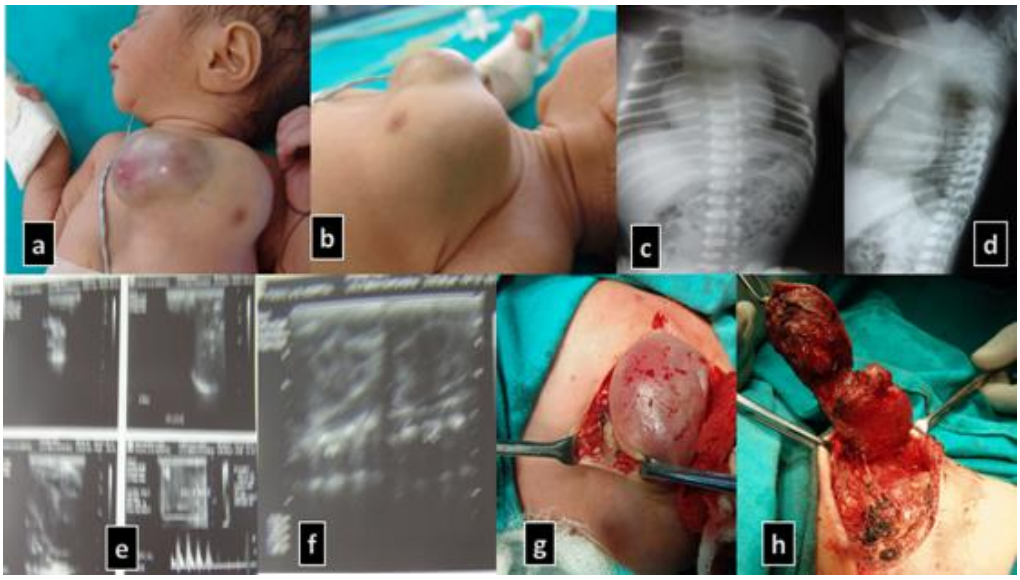

Fig. 1: a) Clinical photograph showing tumor with localized redness and cutaneous telangiectasia, b) Photograph showing bi lobed nature, c) Chest radiograph showing sift tissue mass, normal lungs, d) Lateral radiograph showing mass without calcification, e) Ultra sonogram showing the mixed echoic lesion, f) Longitudinal USG and Doppler, g) Planned excision along with ellipse of skin, h) Showing a feeder vessel from inter costal artery

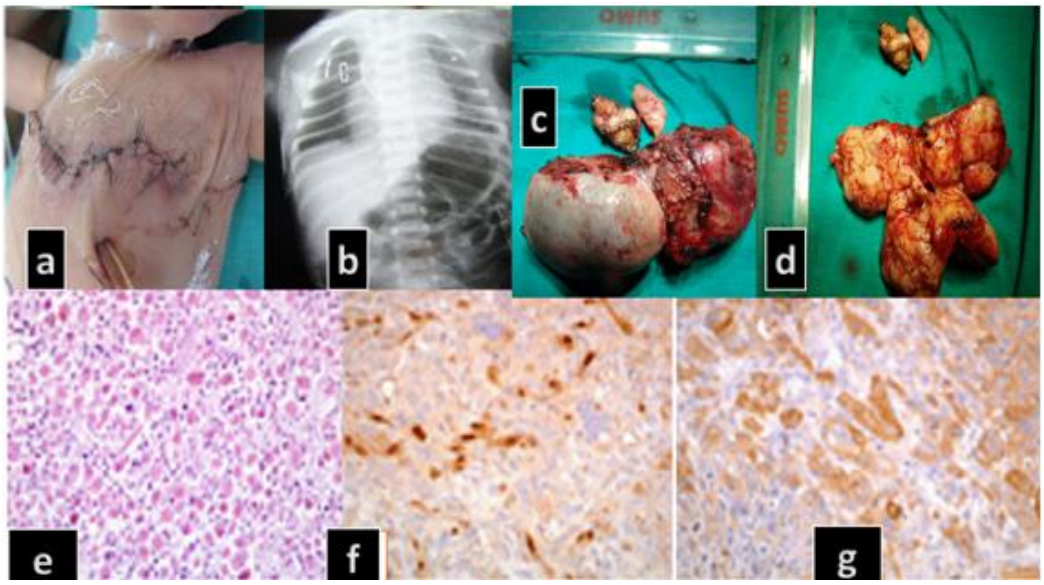

Fig. 2: a) Final closurewith local advancement flaps, b) Post OP chest radiograph, c) Excised tumor with eclipse of skin, d) Cut surface of tumour showing solid nature, e) Microphotograph showing round cells, f) Desminand vimentin positive, g) Cytokeratin and S100 negative

DISCUSSION

Soft tissue tumors have been reported to account for approximately 25% of neonatal tumors. Most of the soft tissue tumours seen were benign [1]. Soft tissue vascular malformations are the most frequent benign tumors, called as Haemangiomas [1, 7].

The two main types of RMS in kids are reported: Embryonal RMS (usually develops in the head and neck area, genitals, or urinary tract, occurs in kids younger than 6) and Alveolar RMS (often affects the arms or legs, chest, or abdomen; occur during the teen years) [8].

In the index case, the tumour was involving the chest wall primarily and was present since birth without signs of this syndrome.

R.D Spicer classified neonatal soft tissue tumours in 5 clinical groups [9].

A: Excellent prognosis with no treatment or simple surgical excision.
B: Good prognosis treatment depends on anatomical site.
C: Good prognosis treatment usually surgical but chemotherapy may be indicated in certain situations.
D: Intermediate prognosis treatment as for older child usually surgery or chemotherapy.
E: Poor prognosis.

Controversies exist in histogenesis of soft tissue sarcoma [10]. The histological type of sarcomas does not provide sufficient information in the prediction of the clinical course and thus plan for therapy. Grading of the tumour on the basis of histological parameters only, evaluates the degree of malignancy and the probability of distant metastasis. The staging of the tumours on the basis of both clinical and histological parameters gives information on the extent of the tumour [11].

Small number of experience as seen in the recent SIOP report of 102 cases in 2005 [2], indicate difficulty of building consensus. Malignant undifferiented fibromatosis (MMT) versus alveolar rhabdomyosarcoma always exist.

Chest radiograph revealed soft tissue mass without involvement of ribs, no pulmonary metastasis and no evidence of pleural effusion. The point of conflict revolves around the description of small round cell with big nucleus. Immunohisto chemistry does help to differentiate the undifferentiated masses. SIOP group [2] showed 64 cases of rhabdomyosarcoma and 30 cases of non RMS MMT. The histology of the index case was conflicting. Immunohistochemistry being positive for vimentin and desmin, and negative for S100 supported RMS.

Treatment requires multidisciplinary approach. Both surgery and chemotherapy have their own specific role. Complete resection of chest wall rhabdomyosarcoma is recommended [12].

Review of patient after 8years without any local or distant disease confirmed that this case belongs to R.D Spicer's group A RMS. Thus this case suggests a good wide local excision forms the important pillar of success in soft tissue tumours in newborn.

CONCLUSION

A rarest of the rare and first case of alveolar RMS of chest wall in a newborn successfully treated with surgical excision is being reported.

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