A Case Report

FIBROUS HAMARTOMA; A BENIGN TUMOUR THAT MIMICS A MALIGNANT ENTITY, CASE REPORT

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ABSTRACT: Fibrous hamartoma is a rare tumor located subcutaneously in infants and children. The tumor was previously known as 'subdermal fibromatous tumor of infancy'. Two years old Malay girl presented with history of progressive painless swelling over her right upper anterior chest wall over two months duration prior to being attended at the paediatric surgical clinic. On general examination, the child was active and pink. There was a hemispherical lump, measuring 12x 10 cm over the right upper anterior chest wall. Computed tomography showed a hypodense lesion suggestive of fatty component with multiple septate within. Post operatively, patient was well. On her follow up, her wound healed well. Accurate diagnosis allows appropriate treatment, which may consist of either close clinical follow-up or surgical resection in symptomatic cases.

Key words: Fibrous hamartoma, chest wall, tumour, peadiatric

1. INTRODUCTION

Fibrous hamartoma is a rare tumor located subcutaneously in infants and children. The tumor was previously known as 'subdermal fibromatous tumor of infancy'. The term 'fibrous hamartoma of infancy' was coined by Enzinger in 1965. It usually occurs in the first two years of life. The tumor may be present since birth or in some cases, present in older children and is usually located at shoulder, axilla, neck and upper arm. Cases have been reported that it occurs in the perianal area, scrotum and the inguinal region. Fibrous hamartoma presents as a painless solitary, at times rapidly growing nodule. Multiple lesions have been reported. There is a male predominance of 3:1. The clinical course is benign, despite its infiltrative appearance and tendency to recur. The tumor shows no evidence of malignant transformation.

The current manuscript presents a case of a 2 years old child who had a progressive anterior chest wall tumor. Histopathological examination denoted fibrous hamartoma of the chest wall.

2. CASE REPORT

A 2 years old Malay girl presented with history of progressive painless swelling over her right upper anterior chest wall over two months duration prior to being attended at the paediatric surgical clinic. There was no similar lump elsewhere. No history of trauma, fever or discharge was observed from swelling.

On general examination, the child was active and pink. There was a hemispherical lump, measuring 12x 10 cm over the right upper anterior chest wall. The lump was warm but non tender, and there were dilated superficial capillaries overlying the lump. It was not attached to the skin and was free from the chest wall.

Ultrasound denoted possibility of subcutaneous lipoma. Since the lump was warm and had dilated superficial capillaries, a CT scan was done with a hemangioma in mind. Computed tomography showed a hypodense lesion suggestive of fatty component with multiple septate within (Figure 1).

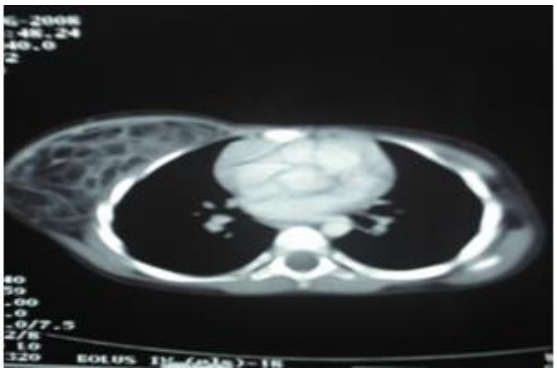

Figure 1: CT scan showed a hypodense lesion suggestive of fatty component with multiple septate within, suggestive of lipoblastoma.

The lesion was beneath the pectolaris major muscle which was stretched and thin. There was no bony involvement. The impression was lipoblastoma. Excision was done under general anesthesia. Post operatively, patient was well. On her follow up, her wound healed well.

Histopathological examination showed matured adipocytes separated by fibrocollagenous stroma. There were also immature mesenchymal cells. It was interpreted as fibrous hamartoma of infancy with benign features.

3. DISCUSSION

Children with chest wall subcutaneous lumps are frequently brought to the clinic for consultation. Among the differential diagnosis are lipoma, lipoblastoma, fibromatosis, cystic hygroma and pilomatrixoma. Not many would ever think about fibrous hamartomas. In fact, diagnosing fibrous hamartoma is usually not easy and it is rare.

The tumor was previously known as 'subdermal fibromatous tumour of infancy'. Fibrous hamartoma of infancy was first

described by Reye in 1954 and later coined its current name 'fibrous hamartoma of infancy' by Enzinger in 1965. It usually occurs in the first two years of life. The tumor may be present since birth or in some cases present in older children. Common site are at the shoulder, axilla, neck and upper arm. There are cases that have been reported in the perianal area, scrotum and the inguinal region.

Pathological gross appearance shows a firm circumscribed poorly defined fibro-fatty tissue usually present in the deep dermis and subcutis. Cut surface has a glistening gray-white appearance interspersed with fatty tissue. Microscopically, there are three main components, interlacing trabeculae of dense fibrocollageneos tissue. Finger like projections of fibrous tissue extend into the fatty tissue, Small round to ovoid nests of undifferentiated spindle or stellate cells set in a myxoid stroma containing delicate vessels. The mucoid matrix is Alcian blue positive. Sparse lymphocytes may be present in the stroma and finally Interspersed mature adipocytes. The fat tissue varies from case to case and is much greater than normally present in the subcutis.

Extension into underlying tissue, like muscle or fascia, is a known feature, but visceral involvement has not been reported. Routine hematoxylin-eosin stained sections are generally sufficient for a diagnosis, but immunochemistry may be useful. The primitive mesenchymal cells are vimentin positive, the fibroblastic component is desmin and smooth muscle actin positive and the adipocytes are S100 protein positive [1].

Pathological diagnosis can sometimes be a challenge to the pathologist. All fibrous hamartoma has the characteristic mixture of fibrous and adipose tissue and nests of immature mesenchyme in different proportions, and nearly all shows lymphocytes and thick patent capillaries in the mesenchyme. However, the fibrous component varies considerably in amount, pattern, and cellularity. This is what differentiates the lesions that is typical in some areas, in others resembles collagenizing vascular granulation tissue, deep fibrous histiocytoma, or fibromatosis. Those in which adipose tissue predominates are distinguished from fibrolipoma by foci of immature mesenchyme and from lipoblastoma by their lack of a capsule and of a lobular pattern [2].

Mesenchymal hamartoma is another differential diagnosis. Although it is rare, it could be easily considered be mistaken for fibrous hamartoma. Radiography in all cases revealed a large expansile rib lesion and an associated extrapleural soft-tissue mass. Mineralization is seen in 64% of lesions on plain radiography and in 100% of lesions on CT Scan. Hemorrhagic cavities from a secondary aneurysmal bone cyst are common. Mesenchymal hamartoma of the chest wall may be recognized by its characteristic occurrence in infancy and cross-sectional imaging is needed to differentiate it from fibrous hamartoma [3].

Local recurrence rate is approximately 16% according to a study by Enzinger and may have been explained by incomplete excision of the lesion. The clinical course of FHI is benign, as demonstrated by a study of untreated FHI [4]. In this study with protracted and untreated FHI, the tumor capsule was more complete and well defined which made surgical excision less difficult secondary to more distinct cleavage planes. Additionally, this study demonstrated that untreated FHI continues to grow without regression. Local excision is the treatment of choice for FHI; however, delayed surgery is not associated with an increased risk of operative complications [5].

4. CONCLUSION

Fibrous hamartomas of the chest wall are unusual subcutaneous lesions most commonly affecting infants. The clinical manifestation and radiographic appearance may suggest a more aggressive malignant process unless one is familiar with this diagnosis. CT scan and MR imaging may not clearly reflect the underlying histopathologic characteristics of these lesions.

5. REFERENCES

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