Fibrous Hamartoma of the Chest Wall: A Benign Tumour that Mimics a Malignant Entity

The Editor,

Sir,

Fibrous hamartoma (FH) is a rare and benign fibrous proliferation of soft tissue and is usually diagnosed during the first two years of life. Only less than 10% of cases have been diagnosed during the second year of life (1). These rare lesions occur two to three times more commonly in males (2, 3), and recurrence is noted in 10–15% of cases (1). To date, fewer than 200 cases have been reported in the medical literature, and very little is known about its histiogenesis (4). We report a case of FH diagnosed in a two-year-old girl from Malaysia because of its extreme rarity.

A two-year-old female presented with a two-month history of progressive enlarging, painless swelling over the right upper anterior chest wall. There was no history of trauma, fever or discharge from the swelling. General examination revealed that the lump (12 x 10 cm) was warm, non-tender and hemispherical in shape and extended over to the right upper anterior chest wall. It was overlaid by dilated superficial capillaries and was not attached to the skin (Fig. 1). Ultrasound findings revealed the possibility of subcutaneous lipoma. Additionally, the lump was warm and had dilated superficial capillaries. Therefore, haemangioma and lipofibromatosis were ruled out. Computed tomography (CT) scan showed a hypodense lesion, suggestive of a fatty component with multiple septate (Fig. 2). The lesion was located beneath the pectoralis major muscle which was stretched and thin without any bony involvement, suggesting the presence of lipoblastoma. The patient was operated under general anaesthesia, and the lump was excised. The patient showed no sign of recurrence six months after surgery (Fig. 3). Histologically, the lump was composed of matured adipocytes separated by

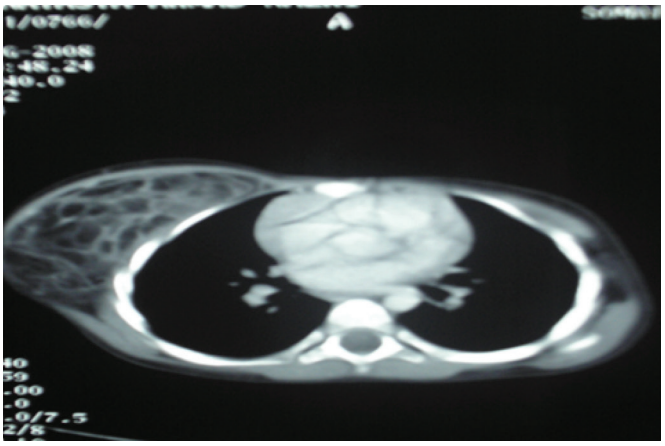

Fig. 2: Computed tomography scan showed a hypodense lesion suggesting presence of fatty components with multiple septate, suggestive of lipoblastoma.

Fig. 1: Swelling at the right upper anterior chest wall.

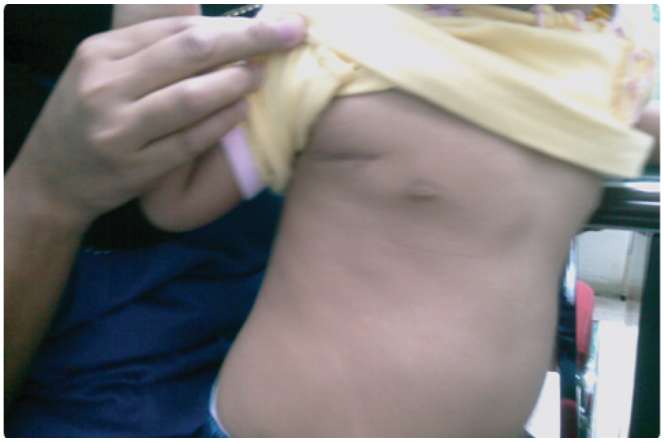

Fig. 3: Healing at postoperative follow-up.

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fibrocollagenous stroma. Furthermore, the presence of immature mesenchymal cells suggested FH of infancy with benign features (Fig. 4).

Clinically, the lesions of FH are characterized as a firm, non-tender, painless solitary mass and poorly circumscribed with ill-defined margins and rapidly growing nodule. An occurrence of more than one lesion has been reported (3). Despite its infiltrative appearance and tendency to recur, the clinical course of the tumour is benign. Sometimes, the tumour is misdiagnosed as a malignant tumour due to the high degree of cellularity and the presence of immature cells leading to overtreatment (3).

Pathological gross appearance showed a firm circumscribed and poorly defined fibro-fatty tissue that is usually present in the deep dermis and subcutis. The cut surface of tumour has a glistening grey-white appearance interspersed with fatty tissue. Microscopically, there are three main components: (a) interlacing trabeculae of dense fibrocollageneous tissue, finger-like projections of fibrous tissue extending into the fatty tissue; (b) 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 (c) interspersed with mature adipocytes. The presence of fatty tissue varies in different cases and is more abundant than normally present in the subcutis. Extension into the underlying tissue (such as muscle or fascia) is a known feature, but visceral involvement has not been reported. Routine haematoxylin-eosin stained sections are generally sufficient for a diagnosis, but immunochemistry may be useful. The primitive mesenchymal cells, fibroblastic components, smooth muscles and adipocytes are vimentin, desmin, actin and S100 protein positive respectively (5).

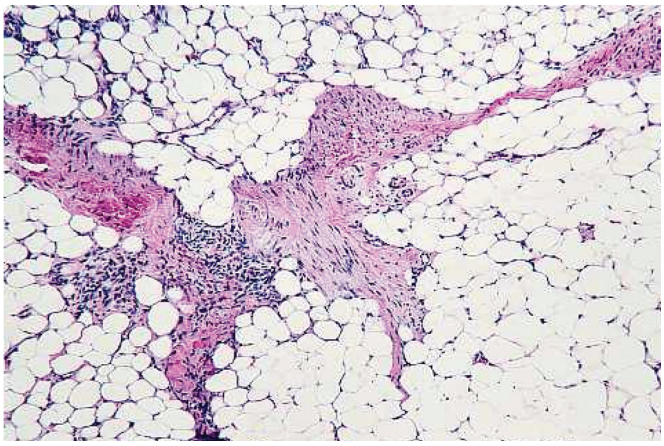

Fig. 4: Low power view of part of the subcutaneous component showing fibrous septa formed bridges of varying width and also having jagged borders and cell clusters.

Pathological diagnosis can sometimes be challenging for pathologists because FH has a mixture of fibrous and adipose tissues and nests of immature mesenchyme in different proportions, nearly all of which show lymphocytes and thick patent capillaries in the mesenchyme. However, the fibrous component varies considerably in amount, pattern and cellularity that differentiates these lesions from others closely resembling collagenizing vascular granulation tissue, deep fibrous histiocytoma and fibromatosis. In cases where adipose tissues predominate, FH is distinguished from fibrolipoma by foci of immature mesenchyme and from lipoblastoma by its lack of capsule and lobular pattern (3).

Mesenchymal hamartoma is another differential diagnosis. Radiographic findings revealed a large expansile rib lesion and an associated extrapleural soft-tissue mass. Mineralization is seen in 64% of the lesions on plain radiography and in 100% of the lesions on CT scan. Haemorrhagic 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 FH (6).

The current case was longstanding. Hence, the tumour capsule was more well-defined with distinct cleavage planes. This made surgical excision less difficult.

It is imperative not to overdiagnose and overtreat these benign lesions with radical and ablative procedures. Wide local excision taking judicious margins with tissue reconstruction is a treatment of choice for FH (4).

In conclusion, FHs of the chest wall are unusual subcutaneous lesions affecting infants. The clinical manifestation and radiographic appearance may suggest a more aggressive malignant process unless one is familiar with the diagnosis. Computed tomography scan and magnetic resonance imaging may not clearly reflect the underlying histopathological characteristics of these lesions. A strong clinical acumen coupled with prompt wide local excision and subsequent histopathological confirmation is crucial for accurate diagnosis and treatment.

Keywords: benign fibrous proliferation, benign tumour, chest wall, fibrous hamartoma, surgery

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