MALIGNANT MESENCHYMOMA - AN UNUSUAL PRESENTATION IN A CHILD (A Case Report)

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Introduction

Malignant mesenchymoma are small group of malignant soft tissue tumours rarely encountered in children. Stout in 1948 defined this histological subtype of soft tissue sarcoma as that composed of two or more unrelated tissue elements sufficiently differentiated to permit clear recognition of its histogenetic type amalgamated within a tumour in addition to fibrosarcomatous element [1]. Enzinger and Weiss [2] recommend that only those tumours that meet Stout's original criteria for malignant mesenchymoma be designated as such thus avoiding the use of the designation for 'wastebasket' category. The common sites are extremities and retroperitoneum. We report such a case in a child for its rarity and unusual presentation.

Case Report

A 12-year-old boy was hospitalised with complaint of cough, intermittent fever, weight loss, distension of abdomen and gradually increasing breathlessness of one year duration. On examination he had tachycardia, tachypnoea and facial congestion. Right side of the chest had spaced out ribs with deviation of trachea to the left. Radiograph of the chest revealed areas of calcification in the right hemithorax. The patient was put on antitubercular therapy but showed no response. Computerised tomogram scan chest and abdomen (Fig 1 and 2) showed soft tissue mass in the right adrenal region with calcified areas reaching up to right hemithorax. A clinical impression of neuroblastoma right hemithorax, paraspinal region with ascitis and bilateral pleural effusion was made. Thoracotomy was performed for debulking tumour mass in the right hemithorax. The tumour mass was filling the entire right chest cavity pushing the lung medially and was densely adherent to the chest wall, mediastinum and the diaphragm. The tumour could not be mobilized due to considerable bleeding and only few bits of tissue were taken to establish the diagnosis.

The largest biopsy tissue measured 2 x 1 cm, yellowish white, oval in shape with smooth bossolated surface. Cut section showed homogeneous surface with tiny foci of necrosis. Another firm irregular reddish brown tissue with foci of haemorrhage and two calcified tissue masses were also received. On microscopic exami-

nation only two lines of differentiation were identified in addition to fibrosarcomatous element, the pleomorphic areas showing muscle differentiation (Fig 3) and chondroid areas merging with fibrosarcoma component comprising of atypical chondrocytes embedded in chondroid matrix classifiable as chondrosarcoma (Fig 4). Electron microscopic (EM) study revealed spindle cell lesion with primitive mesenchymal cells, fibroblastic and myofibroblastic differentiation. A few focal areas showing chondroblastic differentiation were present. Based on light and EM studies the tissue expressed three distinct mesenchymal lines of differentiation in the tumour mass and thereby qualified to label as malignant mesenchymoma.

Discussion

The term malignant mesenchymoma was introduced by Stout who simplified the classification of sarcomas composed of multiple mesenchymal ele-

Fig. 1 : CECT Scan chest and abdomen showing areas of calcification in right hemithorax and upper abdomen.

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Fig. 2 : CECT scan thoracoabdomen showing right large suprarenal mass with dense central calcification and associated pleural effusion (right).

Fig. 3 : Photomicrograph showing muscle differentiation in addition to fibrosarcomatous elements. Fascicles of spindle cells with large hyperchromatic nuclei and eosinophilic cytoplasm (x20).

ments in various stages of differentiation [1]. Stout recommended naming tumours composed of two or more malignant mesenchymal elements any of which taken by itself might be considered a primary malignant neoplasm in fibrosarcomatous background as malignant mesenchymoma. The histological diagnosis of the tumour is not always simple on light microscopy since the differentiation of these differentiating primitive cells is not complete. Areas of undifferentiation need immunohistochemical and ultrastructural studies. However many tumours under this category can be specifically described depending upon the prominent component and designated as leiomyosarcoma, rhabdomyosarcoma or chondrosarcoma etc., within the

Fig. 4 : Photomicrograph showing chondroid differentiation. Neoplastic chondrocyte with large hyperchromatic uni or multinucleated nuclei in lacunar spaces embedded in chondroid matrix (x20).

same tumour.

Mesenchyme is an extremely versatile tissue. Almost all known combinations of various mesenchymal elements have been reported in the literature from various sites [1,2,5,6]. The most common sites are retroperitoneum and extremities especially the deeper tissue [4]. The rarity of the tumour has been widely accepted by various authors more so in children. Middle aged adults and old patients are usually affected. In children the largest series of 42 cases of malignant mesenchymoma was reported by Nash and Stout [7].

We reported this case for its rarity and unusual site of occurrence involving almost the entire hemithorax and retroperitoneal tissue in a child.

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