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Malignant fibrous histiocytoma of the lung in a child

An unusual neoplasm that can mimick inflammatory pseudotumour

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Abstract We describe a 12-year-old patient with a primary pulmonary mass in the left upper lung. The diagnosis of inflammatory pseudotumour was suspected preoperatively. After pathological examination and complete clinical evaluation, a diagnosis of malignant primary pulmonary fibrous histiocytoma was established. This is a very uncommon primary neoplasm of the lung and to our knoweledge only five paediatric cases have been reported. Because of the rarity of these sarcomas and histological similarities to benign inflammatory pseudotumour, care must be taken to avoid confusion between the two disorders particularly in intra-operative frozen sections.

Conclusion Primary malignant fibrous histiocytoma of the lung is an uncommon tumour that should be considered in the differential diagnosis of pulmonary neoplasms of childhood. The histological diagnosis can be difficult due to the similarities with inflammatory pseudotumour.

Key words Child · Inflammatory pseudotumour · Malignant fibrous histiocytoma · Pulmonary neoplasms

Abbreviations *IPT* inflammatory pseudotumour · *MFH* malignant fibrous histiocytoma

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Introduction

Primary malignant fibrous histiocytomas (MFH) of the lung are uncommon tumours, described mainly in adults [5]. In recent reviews of primary pulmonary neoplasms of childhood, no case of MFH was described [4, 7, 10] and only five isolated cases of primary pulmonary MFH in children have been described [1, 2, 8, 11, 13]. The diagnosis in children of MFH of the lung is complicated by the fact that inflammatory pseudotumour (IPT) and other benign fibrous-histiocytic lesions display several overlapping histological features [3, 12]. Diagnostic problems become even greater on examination of operative frozen sections and of fine needle aspiration biopsies, where a definitive diagnosis is not always possible [5].

We describe a case of a primary MFH of the lung in a 12-year-old girl with malignant clinical and histological features.

Case report

A 12-year-old girl was admitted to our hospital with a history of chest pain, fatigue, non-productive cough and weight loss. Physical examination showed a rectal temperature of 38.7°C, disminished breath sounds and dullness to percussion over the left upper lung field. Peripheral blood smears showed 6% eosinophils. Chest X-rays demonstrated a discrete mass in the left upper lung (Fig. 1). A CT scan of the chest showed a 9 cm, solid mass in the left upper lobe (Fig. 2). A diagnosis of malignant spindle cell tumour was made from intra-operative frozen sections and a lobectomy of the left upper lobe was performed.

Pathological examination revealed a $7 \times 6 \times 5$ cm solid tumour with irregular contours that infiltrated the pleura. Histologically the tumour was highly cellular and consisted mainly of spindle cells with a fascicular and storiform growth pattern (Fig. 3a). In some areas neoplastic cells had a histiocyte-like appearance and giant multinucleated cells were also observed (Fig. 3b). The nuclei were pleomorphic, hyperchromatic, with prominent nucleoli and occasional multinucleation. The mitotic rate was high, with more than 10 mitotic figures per 10 high power fields. Atypical mitotic figures were also seen (Fig. 3a). Necrosis and occasional inflammatory cells, mainly lymphocytes and eosinophils were observed through-

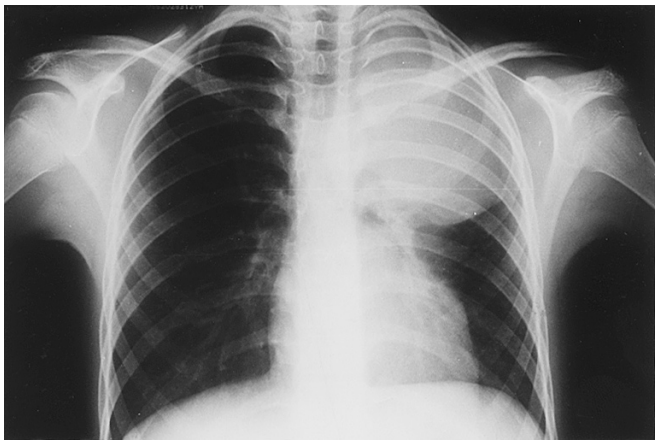

Fig. 1 Postero-anterior chest film shows a circumscribed, round mass in the left upper lung

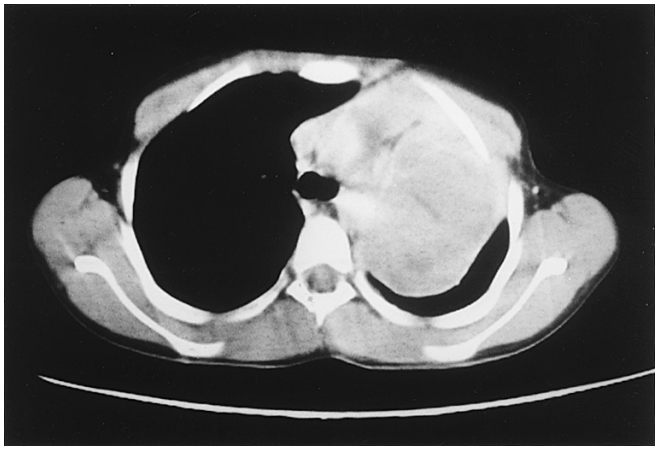

Fig. 2 CT scan of the upper chest shows a large tumour almost completely occupying the left hemithorax

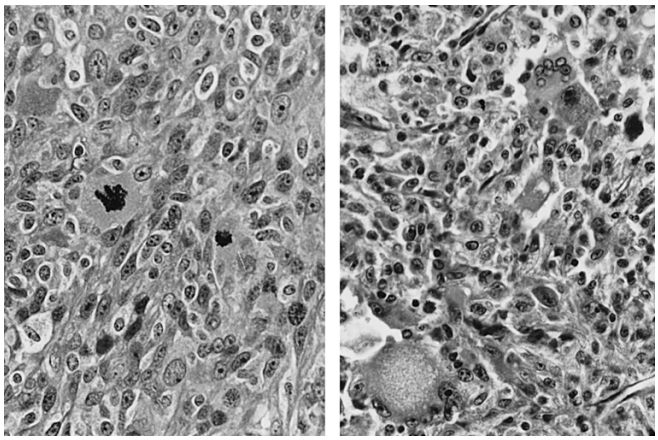

Fig. 3 a Fascicular area consisting predominantly of spindle-shaped fibroblast-like cells. Atypical mitotic figures are present (H&E original magnification × 250). **b** Giant multinucleated cells with histiocytic-like appearance and pleomorphism (H&E original magnification × 250)

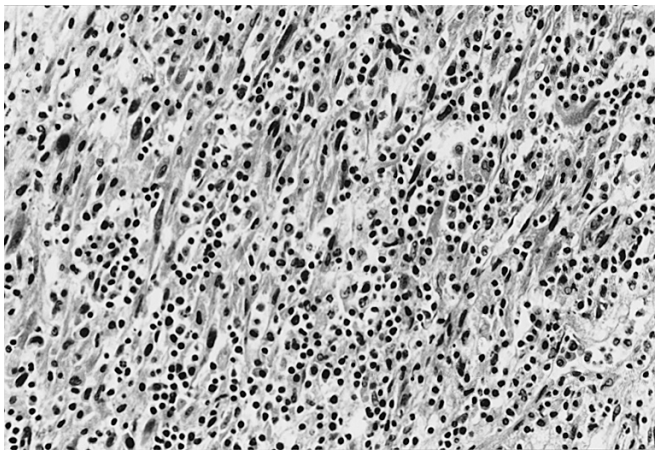

Fig. 4 Spindle cell population with multiple, chronic inflammatory cells resembling inflammatory pseudotumour (H&E original magnification × 65)

out the tumour. Some areas showed abundant chronic inflammatory cells intermixed with a population of fibroblast-like cells without atypia (Fig. 4). Infiltration of the surrounding lung parenchyma, pleura and bronchus was present. Immunohistochemical studies showed a positive reaction for vimentin, Factor XIII, alpha-1-antitrypsin and alpha-1-antichymotrypsin. There was no cellular immunoreactivity for high and low molecular weight cytokeratins, smooth muscle actin, or Factor VIII. Ultrastructural appearance of the tumour varied from regions where histiocyte-like cells predominated to those where the fibroblastic component was more evident. Intermediate forms were also observed. Basal lamina, myofilaments, tonofilaments, desmosomes, dense bodies, neurosecretory granules or melanosomes were not found.

Chemotherapy with ifosfamide, vincristine and actinomycin D according to the protocol for stage II International Society of Paediatric Oncology mesenchymal tumours was instituted. Five months after surgery a CT scan was performed and tumoural involvement of the upper left posterior pleura and costal arch was observed. Fine needle aspiration biopsy and thoracoscopy with biopsy confirmed the local recurrence of the tumour which showed the same histological features.

Discussion

Primary pulmonary neoplasms are uncommon in children. They must be differentiated from metastases and IPT. In our case, the histological diagnosis of MFH – an extremely rare pulmonary tumour – was made on the basis of morphological and immunohistochemical findings. The tumour met six of the following criteria considered for inclusion of a neoplasm into the MFH group: (1) atypical fibrous-histiocytic cellular proliferation; (2) storiform or fascicular growth pattern; (3) bizarre giant cells; (4) three or more mitosis per 50 high power fields; (5) atypical mitoses; (6) necrosis; (7) vascular invasion; and (8) metastases. According to Gal et al. [9] only four of these criteria are necessary for the diagnosis of pulmonary MFH. Local recurrence of the tumour was confirmed histologically 5 months after surgery.

Diagnosis of primary pulmonary and extrapulmonary MFH may be difficult because of the histo-

logical similarities to IPT and other benign fibrous histiocytic lesions. IPT has been misdiagnosed as sarcoma even by experienced pathologists [15]. The diagnostic difficulties are accentuated in intra-operative frozen sections. Cytological imprints and rapid immunocytochemical analysis must be used in these cases [6]. There is now general agreement that IPT is a non-neoplastic reactive lesion, although it may have angio-invasive properties [16], and malignant transformation into MFH has been reported [14].

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