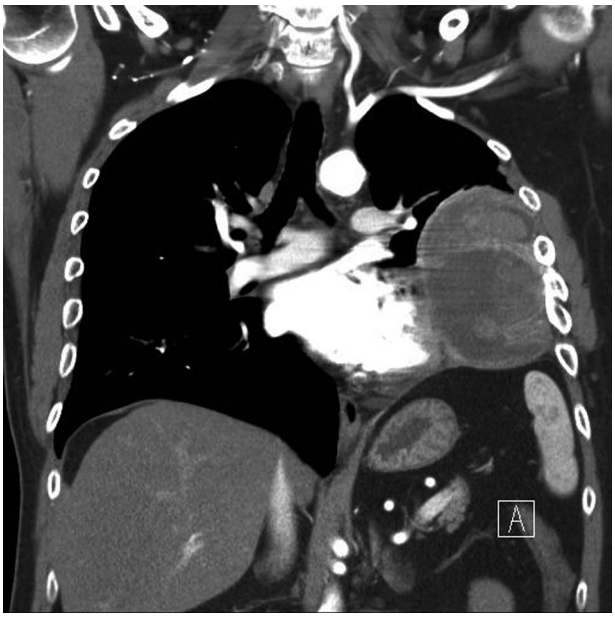

Figure 1 Coronal CT scan upon referral depicting inhomogeneous mass in left thoracic cavity.

Mesenchymal cystic hamartoma? A revised diagnosis after 23 years

In 1994, we published a rare case report of a left sided pneumothorax caused by a mesenchymal cystic hamartoma (MCH) in this journal (*Thorax* 1994;49:1175–6).[1]

After a follow-up of 23 years, the same patient was referred for a left sided pain and thoracic mass at age 36 years. At age 14 years, a left sided thoracotomy was performed for recurrent pneumothorax followed by lobectomy of the left lower lobe in which a MCH was diagnosed.[1] Now his symptoms started almost 1 year before referral. Analysis showed an inhomogeneous mass in the lower left thoracic cavity with metabolic activity on the F-18-fluorodeoxyglucose positron emission tomography-PET scan. Follow up CT-scans showed slow progression (figure 1) and CT-guided biopsy showed a monophasic malignant tumour consisting of spindle cells, immunohistochemically positive for vimentin and epithelial membrane antigen (EMA) (figure 2). Additional fusion gene analysis confirmed synovial sarcoma (SS18-SSX1, t(X;18)(p11;q11): positive). Tissue from the original resection was still available and showed an identical histological and immunohistochemical pattern. However, the RNA was degraded and could not be analysed. Given these findings we diagnosed the present mass as a late recurrence of synovial sarcoma and rejected the original diagnosis MCH. He was treated with two cycles of doxorubicin and ifosfamide chemotherapy followed by surgical resection. A necrotic cystic tumour and left upper lobe were resected and followed by two adjuvant cycles of chemotherapy.

MCH was primarily described by Mark in 1986 as a rare clinicopathological entity characterised by nodules of primitive mesenchymal cells that gradually increase in size and then become cystic.[2] Since 1986, only few case reports including ours have been published. Often these cases refer to our case in association of pneumothorax as a presenting symptom. Before a diagnosis of MCH can be made alternative diagnoses like pulmonary blastoma, cystic adenomatoid malformation, lymphangioleiomyomatosis, metastatic solid tumours and sarcomas should be addressed. Besides morphological characterisation, immunohistochemistry and proliferation markers can be used but also genetic analysis of soft tissue tumours has become available.[3] Molecular analysis has become essential for an accurate diagnosis in mesenchymal tumours. However, in the presented MCH cases such a genetic analysis is seldom reported.[4] Our case shows that a rare diagnosis of MCH must be made with caution and only after thorough genetic analysis.

The original diagnosis of MCH must now be rejected after 23 years, and changed into a final diagnosis of synovial sarcoma with a very late local recurrence. Given this new finding, the case report on this patient published in 1994 must therefore be revised and retracted.

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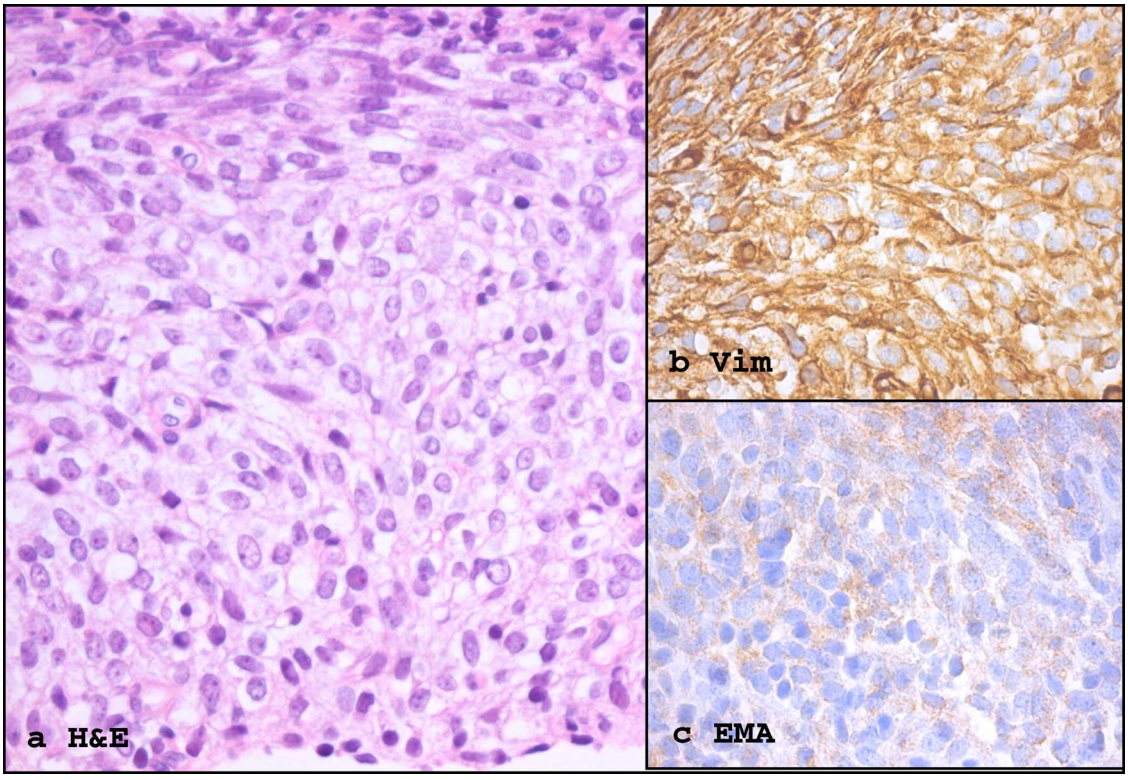

Figure 2 Microscopic findings of recent transthoracic biopsy on which reverse transcription polymerase chain reaction revealed SYT-SSX1 fusion transcript. (A) H&E stain: Sheets of ovoid to fusiform tumour cells with vaguely storiform growth pattern and relatively bland nuclei; (B) diffuse and strong expression of vimentin and (C) weak but distinct expression of epithelial membrane antigen (EMA).

critical revision of the manuscript. He approved the final version of the manuscript and he has no competing interests. He is guarantor of the manuscript taking responsibility for the integrity of the work as a whole from the inception to its publication. SEJK: responsible for interpretation of clinical data, clinical treatment and critical revision and final approval of the manuscript. She has no competing interests. HHMH: critical revision of the manuscript. He approved the final version of the manuscript and has no competing interests. AdFTMV: responsible for interpretation of clinical data, clinical treatment and critical revision and final approval of the manuscript. He approved the final version of the manuscript and has no competing interests. ML-S: responsible for interpretation of clinical data, and critical revision and final approval of the manuscript, she provided the photomicrograph image. She approved the final version of the manuscript and has no competing interests.

Competing interests None.

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