Rapidly growing intrathoracic extraskeletal Ewing’s sarcoma

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Abstract

There are few reported cases of intrathoracic Ewing’s sarcoma, a very rare malignant neoplasm. We report a surgical case of extraskeletal Ewing’s sarcoma that had been followed-up as a stable sized tumour for many years, which then grew rapidly within a year. A 27-year old female patient with a rapidly growing abnormal shadow on chest roentgenogram was admitted to our department. She had undergone periodic examinations including chest computed tomography (CT) scans for 6 years since a small nodule in her chest had been pointed out by chest roentgenogram. The initial CT demonstrated a solitary nodule with a diameter of 20 mm on the parietal pleura that covered the V rib of the posterior chest wall. For 5 years the tumour’s size did not change noticeably but it suddenly grew to about 90 mm diameter in a year. The tumour volume doubling time was calculated to be 17 days.

Keywords: Extraskeletal Ewing’s sarcoma · Thoracic · Primitive neuroectodermal tumour · Volume doubling time

INTRODUCTION

Extraskeletal Ewing’s sarcoma (EES)/primitive neuroectodermal tumours (PNET) are uncommon primary neoplasms of the bone that primarily affects children and young adults. EES commonly affects the extremities (especially the lower extremities), soft tissues of the trunk such as paravertebral and intercostal regions, head and neck, pelvis, mediastinum [1] and peritoneum. A few reported intrathoracic EES cases had huge or multiple tumours associated with mediastinal shift [2], but there were no data on the growing speed. We present a very rare case of intrathoracic EES, including tumour volume doubling time (VDT), which can be followed-up over a long term.

CASE REPORT

A 27-year old female patient with a rapidly growing abnormal shadow on chest roentgenogram was admitted to our department (Fig. 1a, c). She had undergone periodic examinations including chest computed tomography (CT) scan for 6 years since the first detection of a small nodule by the chest roentgenogram. The initial CT demonstrated a 23 × 20 × 20 mm solitary nodule with an extrapleural sign in the left side of the posterior chest wall (Fig. 1b). The nodule was round and present on the parietal pleura over the V rib. T2-weighted magnetic resonance (MR) imaging was very high in the tumour (Fig. 2a). The tumour was diagnosed as a benign tumour like a neurogenic tumour with internal necrosis. F-18 fluorodeoxyglucose (FDG)-positron emission tomography (PET) did not show active uptake (Fig. 2b). She refused surgical expiration of the tumour, but agreed with the physician’s proposal to undergo annual examination. During the following 6 years, the size of the tumour did not change but our CT revealed a 91 × 81 × 67 mm round mass-like shadow (Fig. 1d). The tumour VDT, calculated using the Schwartz formula [3], was about 17 days. FDG-PET showed active uptake (standardized uptake value: SUV max = 4.97: early phase, 5.42: delayed phase) in the tumour (Fig. 1). Signal intensity on both T1- and T2-weighted MR imaging was high in the tumour, and the findings suggested necrosis with bleeding. The intensity on the peripheral side of the tumour was muscle intensity by T1-weighted. Thoracoscopically, the tumour was removed combined with partial lung and parietal pleura. On histopathological examinations, a diffuse proliferation of primitive small round blue cells with round nuclei and scanty cytoplasm was observed. Mitotic activity was low. The tumour cell had invaded the visceral and parietal pleura but lung invasion was not obvious. Immunohistochemical analysis revealed positive staining for CD56, bcl-2, NSE, S100 and CD99, and negative staining for alpha-SMA, desmin, neurofilament, LCA, TTF-1, CAM5.2, EMA, CD34 and calretinin, suggesting an Ewing’s sarcoma (ES). MIB-1 index was about 10%. Fluorescence in situ hybridization using a break-apart ES region 1 probe set showed a split signal in about 60% of the interphases, indicating a chromosomal translocation involving the ES/PNET.

Postoperatively, the patient made an uneventful recovery. She was not treated with chemotherapy or radiotherapy by her choice but no recurrence was detected 1 year later.

DISCUSSION

PNET and EES have a similar neural phenotype and can therefore be considered as the same entity. EES is a rare malignant

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soft tissue ES that is regarded as a member of the family of small, round cell neoplasms of bone and soft tissue, including PNET and neuroblastoma, which commonly affects the lower extremities, paravertebral and intercostal regions, head, neck, pelvis and peritoneum. Differing locations of primary sites have been reported including the duodenum, neck and kidney. In the few cases of primary intrathoracic EES that have been reported, the locations were the lung, oesophagus, posterior mediastinum and chest wall. The clinical diagnosis of EES is difficult and many EES occur in young patients; therefore, those tumours can reach 10 cm by the time of diagnosis [4] in many cases.

The VDT of lung nodules has been widely accepted as an index of tumour growth rate. Steele and Buell [5] suggested that a VDT of 30 ± 490 days represents a malignant zone.
indicative of malignancy, while a VDT outside the above range was referred to as benign zone. In cases of primary lung cancers, VDT are 100–200 days. Band et al. [6] reported that the median VDT of spherical pulmonary metastases in a study of 15 patients with osseous and soft tissue sarcoma was 25 days. Judging from the extremely short VDT of the present case, the malignancy of the tumour is considered to be high grade. However, the size of the tumour had not changed for 5 years since detection. Because of these features, it is not an overestimate to assume that the tumour was benign during the initial 5 years of follow-up. This change in the tumour progression rate might be explained as neoplastic cell transformation or malignant transformation. The present case proves the necessity that a benign tumour should be resected or strictly followed-up over a long time.

Usually, clinical or imaging findings of EES are non-specific and diagnosis is based on histology. However, reported CT findings of intrathoracic EES were huge masses with a central low density area within the mass, representing necrosis. The CT findings of the present case had peripheral enhancement heterogeneous density and a wide-ranging low-density area in the centre of the tumour. The CT and the MRI findings suggested that the necrosis and bleeding in the tumour accelerated it to grow rapidly on its own.

In conclusion, it is difficult to diagnose small-sized intrathoracic peripleural tumour, and thus those tumours should be resected to obtain a histological diagnosis even if the tumour does not enlarge for several years.

Conflict of interest: none declared.

REFERENCES