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CASE REPORT

# Thymic carcinoma initially presented with geographic destruction of scapula in a child

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**Abstract** As the conventional histopathologic examination of thymic carcinoma (TC) is nonspecific, immunohistochemical studies along with correlative radiographic investigations are needed for its correct diagnosis. TC commonly occurs in the late 5th to early 6th decades of life but is extremely rare in childhood. It may be incidentally detected from chest radiographs taken as routine or for other reasons. However, most patients present with symptoms such as chest pain, cough, shortness of breath, dysphagia and hoarseness, which are directly attributable to the mediastinal mass. Although TC frequently invades the neighboring organs, pleura and pericardium and metastasizes to the lymph nodes, liver and lung at the time of the first diagnosis, initial or late metastasis to the bone has been seldom reported in adults. Indeed, the English literature revealed no earlier report on initial bony metastasis in a child to date. We report a case of TC in a 12-year-old boy who initially presented with scapular osteolysis masquerading as a primary bone tumor to emphasize the usefulness of combined imaging for staging and histologic studies, particularly for such an unexpected case.

**Keywords** Thymic carcinoma · Metastasis · Primary bone tumor · Scapula · Child

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## Introduction

The occurrence of a mass lesion in the mediastinum is not rare in childhood, and most are inflammatory or infective lymphadenopathy and neoplasms. Most malignant tumors are lymphomas, germ cell tumors and neuroblastoma. Thymic carcinoma (TC), a highly aggressive malignant tumor, is exceedingly rare in this age group.

For many decades, TC has been considered as metastasis from occult primary carcinoma of another organ rather than carcinoma of the thymus itself because of the histologic similarity [1]. However, Shimosato et al. [2] first recognized it as squamous cell carcinoma of the thymus per se in 1977. The non-specific histology of TC makes its diagnosis by exclusion of metastasis from other organs using clinical and imaging evaluations. Although TC is age immune, it occurs most commonly in adults between 30 and 60 years [3]. TC often invades the neighboring pleura and pericardium and the lymph node at the time of the first diagnosis. Although several cases have been reported to metastasize to bones, most cases occurred in the late phase of the clinical course, and all of the patients were middle-aged adults [4–7]. We were unable to find the earlier publication on TC, which clinically presented as a primary scapular disease or unusual metastasis in a young school boy. Here, we describe a unique case of TC in a child who complained of scapular pain and initially presented with scapular osteolysis mimicking a primary bone tumor. An ultrastructural examination was performed to confirm carcinoma in addition to immunohistochemical and imaging studies.

## Case report

A 12-year-old boy presented with pain in his left scapular area for 1 month, which became significantly aggravated 5 days

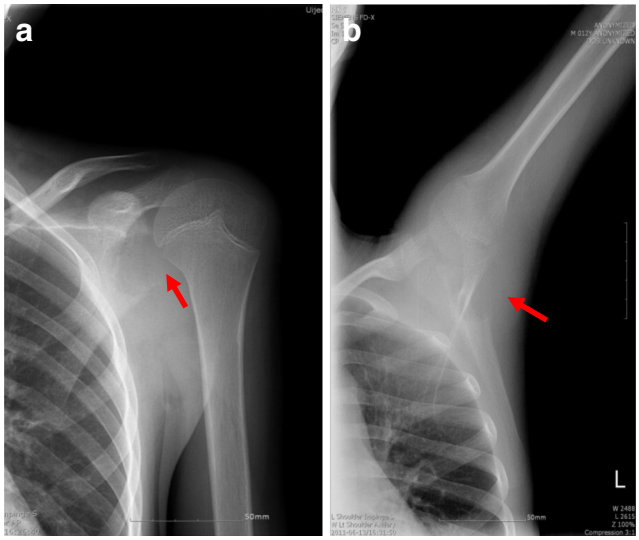


**Fig. 1** Plain radiographs show ill-defined geographic destruction with a hazy soft tissue mass (*arrows*) in the scapular neck and glenoid

before visiting our hospital. He and his parents denied any other symptoms or past illness. Plain radiographs showed a poorly defined geographic destruction with hazy soft tissue mass formation in the glenoid and neck area of the left scapula (Fig. 1). Magnetic resonance images (MRI) showed an ill-defined lesion with posterior cortical destruction and soft tissue mass formation in the same anatomic area. The signal intensities were homogeneously low on the T1-weighted image and intermediate to high on proton density and T2-weighted images (Fig. 2). The lesions in the bone and soft tissue were irregularly enhanced with a non-enhancing portion. Considering the patient's young age and clinical symptoms of scapular pain, a locally aggressive benign bone tumor or primary malignant bone tumor was first considered.

Incision biopsy was performed in the posterior aspect of the scapula to secure a specimen directly from the soft tissue mass. Low-power light microscopy revealed highly cellular nests embedded in the desmoplatic stroma characterized by the sheets and islands of "primitive small round tumor cells" (Fig. 3a). Higher power microscopic examination showed primitive-appearing round to oval tumor cells with prominent nucleoli and smudged features (Fig. 3b). A presumptive histologic diagnosis was a desmoplastic small cell tumor or other small round cell tumors. Metastatic carcinoma was not considered at this time.

For staging, a Tc-99m HDP bone scan, CT scan and positron emission tomography-computed tomography (PET-CT) were performed. The bone scan showed intense tracer uptake in the left scapula and right 6th rib. Chest CT demonstrated a large lobulated mass with positive enhancement (3.5 × 6.3 cm) in the anterior mediastinum closely abutting the sternum and great vessels (Fig. 4). PET-CT revealed inhomogeneous high FDG uptake in the subglenoid of the left scapula (SUVmax = 7.1) and the right 6th rib (SUVmax = 5.8). In addition, there was a large lobulated mass with slightly increased FDG uptake (SUVmax = 4.9) closely attached to the aortic arch in the left mediastinum (Fig. 5). These imaging studies strongly suggested a mediastinal malignant tumor with scapular and rib metastases.

Immunohistochemical stains were performed. Negative stains for LCA, CD99, actin, desmin and vimentin and positive stains for CK Pan and CK 5/6,7 suggested that the scapular lesion is a carcinoma rather than a sarcoma (Fig. 6a–c). Additional positive stains for CD5, C-kit (CD117) and epithelial membrane antigen (EMA) strongly indicated the lesion was a TC (Fig. 6d–f). Finally, an ultra-structural examination was carried out for full determination, and it confirmed that the lesion was a carcinoma by disclosing polygonal tumor cells with oval nuclei, a clear nuclear membrane and prominent nucleolus in association with perinuclear tonofilaments, abundant intracellular desmosomes and microvilli-like protrusions. Based on a correlation of imaging studies and microscopic examinations, a final diagnosis of TC metastasizing to the scapula was made. Chemotherapy with cisplatin, doxorubicin, vincristine and cyclophosphamide was performed. He

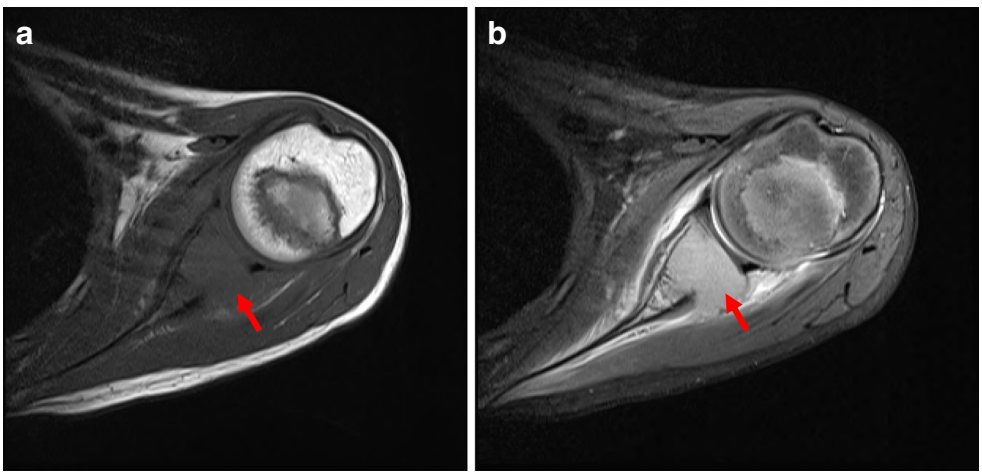


**Fig. 2** MR images show an ill-defined destructive lesion with a definite soft-tissue mass formation (*arrows*) in the same anatomic area. The lesion shows homogeneously low signal intensity on the T1-weighted image (**a**) and high signal on the T2-weighted image (**b**). No evidence of intra-articular involvement is noted

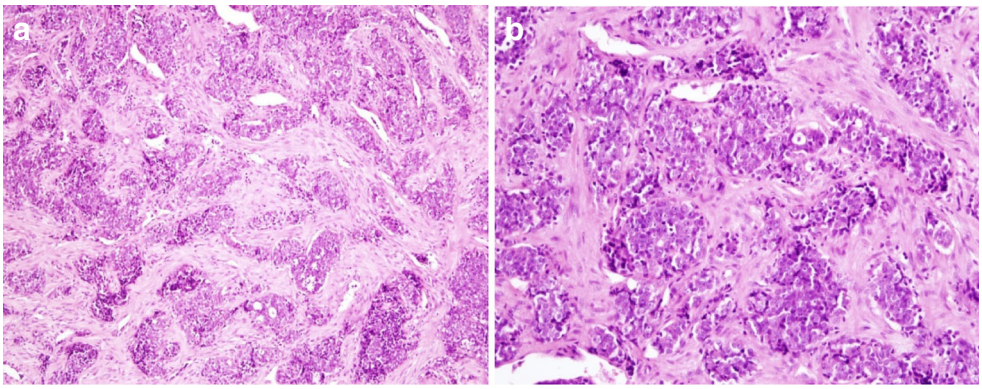


**Fig. 3** (**a**) Low-power microscopic examination (×100) shows numerous multiple high cellular nests embedded in abundant desmoplatic stroma. (**b**) Higher power examination (×200) shows that the nests have primitive-appearing round to oval tumor cells with prominent nucleoli and some smudged features

tolerated two cycles of chemotherapy but succumbed to spreading disease during the third chemotherapy.

## Discussion

Although TC may be incidentally detected from chest radiographs taken as routine or for other reasons, the majority of patients present with symptoms directly attributable to the anterior mediastinal localization of a mass. Symptoms include chest pain, cough, shortness of breath, dysphagia, hoarseness and superior vena cava syndrome. These symptoms could be followed by weight loss, fatigue, fever and anorexia [3, 8]. TC is very rare in childhood. In a clinico-pathologic study of 60 patients, the patient ages ranged from 10 to 76 years with a mean age of 46. Only one patient was aged 10 years [8]. Of 305 patients compiled in 19 studies from the MEDLINE source, 2 patients were aged under 10 years [9]. Another English literature search carried out between 1982 and 2006 found 282 patients, of whom 14 were under the age of 16 years [10].

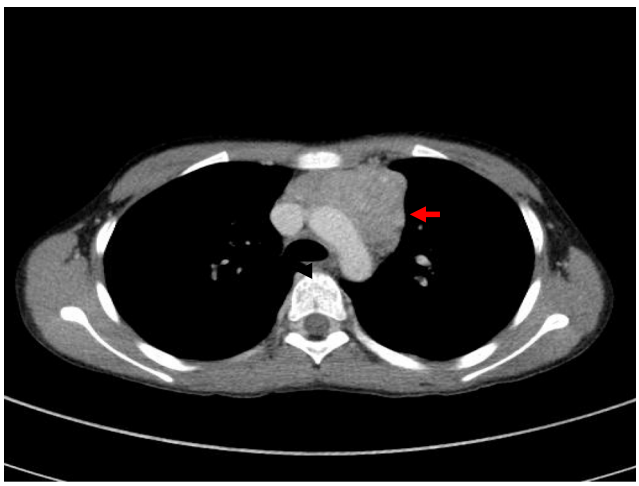

**Fig. 4** CT scan shows a 3.5 × 6.3-cm lobulated and well-marginated contrast-enhanced mass (*red arrow*) in the upper anterior mediastinum abutting the sternum anteriorly and great vessels posteriorly

Radiologically, TC frequently presents as a hazy mass with calcification in the upper anterior mediastinum that may have invaded the adjacent lung or pleura. The CT scan revealed a mass with an irregular lobulation and fuzzy contagious area with an effaced surrounding fat plane [11]. Additionally, the obvious invasion of the neighboring organ may be seen along with suggestive lymphadenopathy [12, 13]. MRI demonstrates typical lesions of intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images [14]. Inoue et al. [15] described foci of low signal intensity within the mass on T2-weighted images along with mediastinal lymphadenopathy. PET-CT presents a large multilobular mass with areas of necrosis and calcification with the SUVmax being 7 or more [16, 17]. A recent pictorial review described that the combination of a SUVmax over 7, mediastinal lymph adenopathy and distant metastasis is indicative of TC [18]. Our patient initially presented with

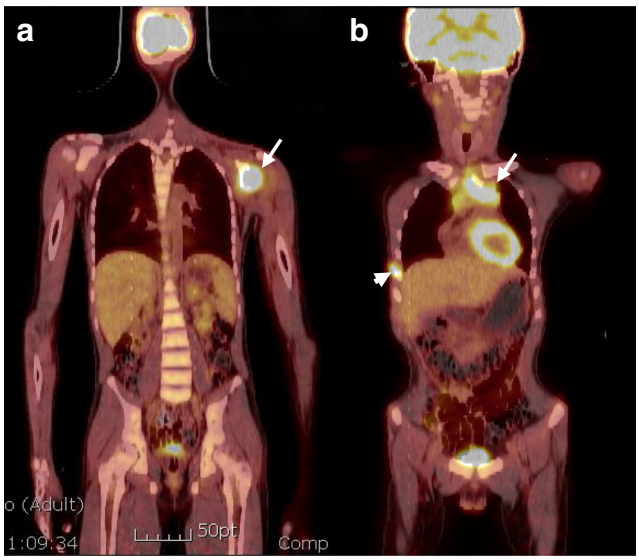


**Fig. 5** PET-CT scan shows intense FDG uptake (**a**) in the left scapula (SUVmax = 7.1) (arrow), (**b**) anterior medistinum (SUVmax = 4.9) (arrow) and right 6th rib (SUVmax = 5.8) (arrowhead). The imaging studies of the CT scan and PET CT scan strongly suggest the lesions are a mediastinal malignant tumor with scapular and rib metastases rather than a primary bone sarcoma metastasizing to the mediastinum

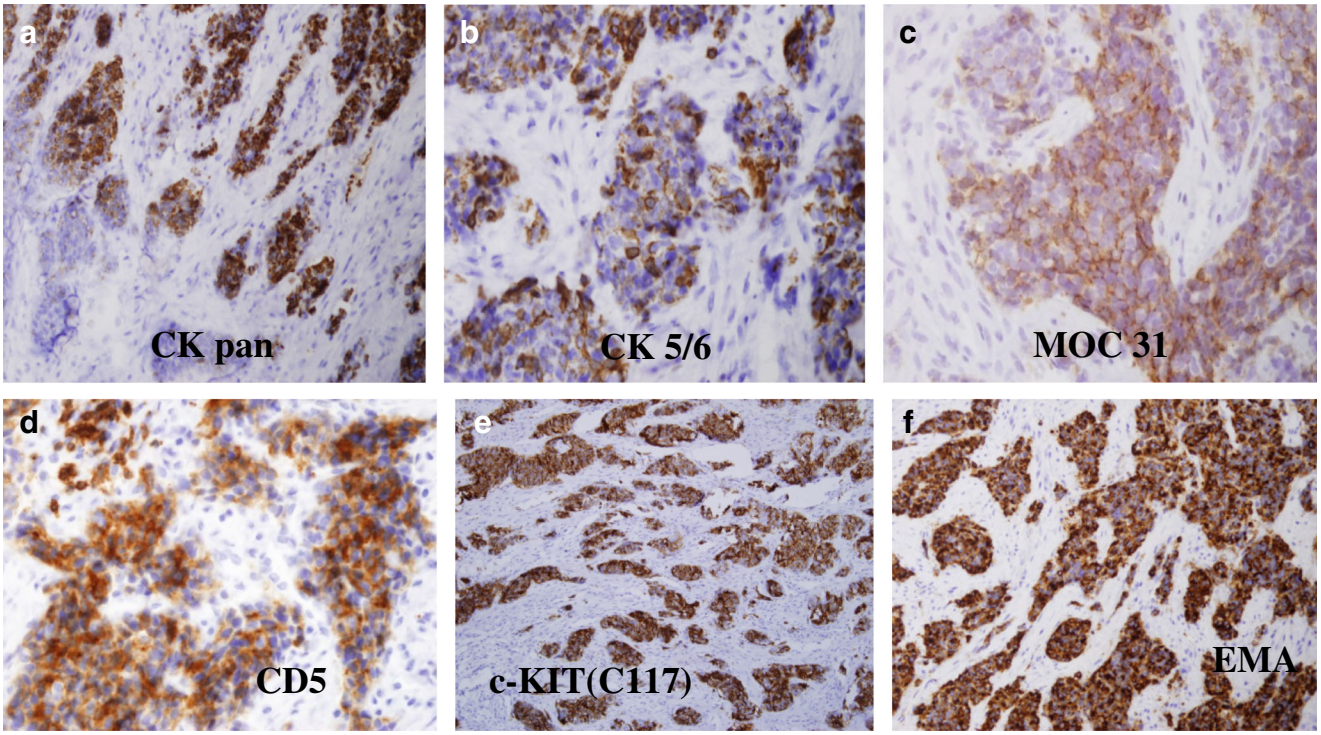


**Fig. 6** Positive immununo-histochemical stains for CK Pan (**a**), CK 5/6 (**b**) and MOC 31 (**c**) suggest carcinoma. Additionally, positive stains for CD5 (**d**) as well as c-KIT (CD117) (**e**) and EMA (**f**) strongly indicate the lesion is a thymic carcinoma

geographic destruction of the scapula that mimicked a primary bone tumor. The differential diagnosis on the plain radiograph and MRI could include a locally aggressive benign bone tumor such as Langerhans cell histiocytosis, and a primary malignant bone tumor including osteosarcoma or Ewing's sarcoma, when considering young age and the clinical symptom of scapular pain. Hematologic malignancies such leukemia or lymphoma might also be suspected to be less likely. During imaging studies for staging, a large, lobulated, enhancing mass was, however, observed in the anterior mediastinum on the CT scan as well as high FDG uptake in the mediastinum (SUVmax = 4.9), scapula (SUVmax = 7.1) and rib (SUVmax = 5.8) on PET CT. These findings strongly suggested a mediastinal malignant tumor with scapular and rib metastases rather than a primary malignant bone tumor metastasizing to the mediastinum.

Histologically, Hishima et al. [19] suggested that CD5 might be valuable in differentiating between primary and metastatic carcinomas in the anterior mediastinum. Epithelial membrane antigen (EMA) was reported to be useful for determining the grade of malignancy among thymic epithelial neoplasms [20]. TC has also been reported to be associated with increased expression of c-KIT (CD117) [21–23]. In our case, immunohistochemical stains for CD5, C-kit and EMA were all positive, which strongly indicated the lesion is a TC. Finally, the diagnosis of TC metastasizing to the scapula was reached by a combination of the imaging studies and immunohistochemical examination despite the patient's young age.

In conclusion, we report an exceptionally rare case of TC in a child that initially masqueraded as a primary tumor of the scapula and emphasized the well-known importance of the correlation of imaging and histologic studies, particularly for such an unexpected case.

#### Compliance with ethical standards

**Conflict of interest** The authors declare that they have no conflict of interest.

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