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Sarcomatoid carcinoma of the thymus – a case report

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A 20-year-old female with a sarcomatoid carcinoma of the thymus invading the left upper lobe of the lung was treated with surgical resection and adjuvant radiotherapy. We report a case of this rare histologic variant of thymic carcinoma and review the literature.

Key words: thymus neoplasms; carcinosarcoma; sarcomatoid carcinoma.

Introduction

Sarcomatoid carcinoma of the thymus is a rare histologic variant of thymic carcinoma, which was named by Snover et al in 1982.[1] That group also suggested that a thymic carcinoma should fulfill the following criteria: (1) anterior mediastinal location and (2) absence of another primary tumor. We have reported 20 consecutive cases of thymic carcinoma in a 10-year period at our institute.[2] Among these 20 cases, no histologic variant of sarcomatoid carcinoma has been disclosed. We hereby describe a case od sarcomatoid carcinoma of the thymus that, microscopically, contains both a malignant epithelial component and a sarcomatoid component. The expression of cytokeratins and epithelial membrane antigen (EMA) in tumor cells could differentiate it from true sarcomas which do not stain for these markers.[3]

Case report

A 20 year old female presented with a six month history of increasing dyspnoea and left chest pain. On admission, physical examination revealed decreased breath sounds in her left upper chest. No lymphadenopathy was found. The full blood count revealed a haemoglobin of 13.7 g/dl, a white cell count of 7.8 x 10^9/l (neutrophils 7.2, eosinophils 0.2, lymphocyte 1.7), and a platelet count of 371 x 10^9/l. A chest radiograph demonstrated a big mass in the anterior aspect of the left lung. A computed tomographic (CT) scan of the chest showed a big necrotic tumour, measuring 14 x 12 x 12 cm in size, arising from the anterior mediastinum and invading to the left upper lung field (Figure 1). The serum titre of beta-choriogonadotropin (beta-HCG), alfa feto proteim (AFP) and carcinoembrionic antigen

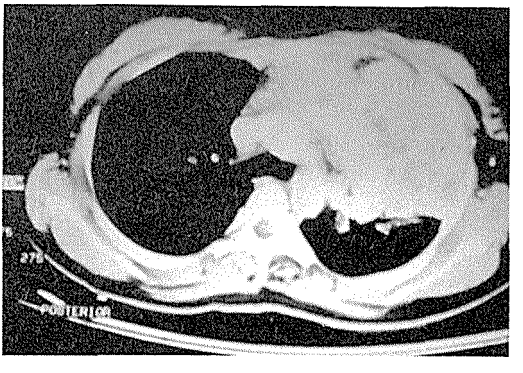

Figure 1. CT scan of the chest demonstrating a big mass arising from the anterior mediastinum and invading the left upper lung field.

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(CEA) were within normal limit. Sono-guided aspiration of the tumour was performed, and a cytological examination showed spindle cell tumour. 99m Tc-MDP whole body bone scanning and liver sonography showed no evidence of metastatic foci.

An operation was performed via standard posterolateral thoracotomy. While the tumour occupied the whole anterior mediastinum, its left lateral site invaded the left upper lobe of the lung. Removal of the mediastinal tumour with a left upper lobectomy of lung was performed. The postoperative course was uneventful, and the intercostal drain was removed on the fifth postoperative day.

Histopathological examination of the tumour revealed a custers of epithelial cells mixed with the strap-like spindle cells (Figure 2). An immunohistochemical study showed a positive staining for cytokeratin in the epithelial area and in some spindle cells (Figure 3). The patient then received radiotherapy with a 6000 Gy tumour dose. There was subjective improvement of dyspnoea and chest pain, and the patient is currently alive 10 months after surgery with no evidence of tumour recurrence or metastasis.

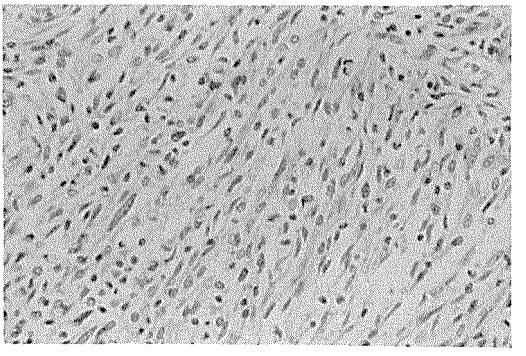

Figure 2. Cluster of thymic epithelial cells mixed with strap-like spindle cells (hematoxylin-eosin, x400).

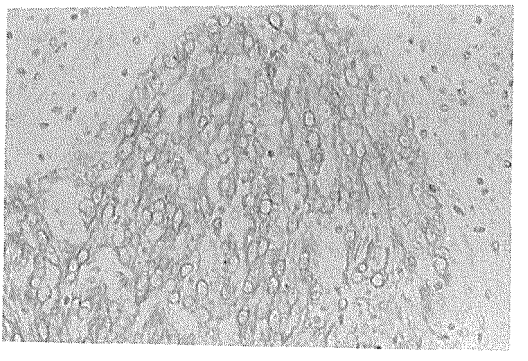

Figure 3. Sarcomatoid carcinoma of the thymus showing dark colouration in the epithelial area and in some spindle cells (peroxidase-antiperoxidase [PAP] staining with cytokeratin) (original magnification x400).

Discussion

Thymic carcinoma per se is a relatively rare tumour, with distinct pathological and clinical characteristics. There were eight histological variants of thymic carcinoma, reported in the literatures with sarcomatoid type among them.[1, 4] Various tumours showing the histological features of sarcomatoid carcinoma are seen also in other organs such as: lung,[5] pancreas,[6] kidney,[7] breast,[8] and urinary bladder.[9] However, sarcomatoid carcinoma of the thymus, as one of the histological variant of thymic carcinoma, has seldom been reported. The clinicopathologic features of the reported cases are summarized in Table 1. Clinically, this tumour mostly occurs in middle or in old age, similarly to the other variants of thymic carcinoma. To our knowledge, this is the youngest case reported in the literature.

In our previous study of 20 cases of thymic carcinoma, we found that invasion of the mediastinal stuctures is almost always present, including the inominate vein, mediastinal pleura, pericardium, and lung.[2] As compared with thymoma, thymic carcinoma has a more invasive tendency on computed tomographic scan examinations, and most of the patients have clinical symptoms caused by tumour compression of the mediastinal vital structures.[2, 4]

In general, thymic carcinoma are immunoreactive to EMA and cytokeratin, but not reactive to AFP, beta-HCG, placental alkaline phosphatase, or common leukocyte antigens.[11–13] Snover et al suggest that the presence of keratin within the spindle cell component can justify the use of the term "sarcomatoid carcinoma".[2]

In one case, initially, germ cell tumour was highly suspected, but a subsequent study of a series of tumour markers disclosed no elevation serum titre of beta-HCG, AFP and CEA.

During operation, we found that the space-occupied mediastinal tumour invaded the left upper lobe of the lung, but fortunately, the hilar structures such as the left upper lobar bronchus, superior pulmonary vein, and pulmonary artery branches to left upper lobe of lung were pushed laterally by the tumour, and total removal of the tumour with a lobectomy could be performed without difficulty.

Table 1. Reported cases of sarcomatoid carcinoma of the thymus.

Year/Author	Age/Sex	Symptoms	Location/Size/Invasion	Therapy	Follow-up
1982/Snover et al[Ref. 1]	64/M	Asymptoms	Ant. mediastinal/ 6x5x4.4 cm/–	Excision	died with metastasis at 13 months postop.
1982/Wick et al[Ref. 4]	53/M	Chest pain, dysphagia, SVC syndrome	Ant. mediastinum/?/SVC	RT & CT	died with metastasis at 28 months postop.
1992/Morita et al[Ref. 10]	53/M	Asymptoms	Ant. mediastinum/? lung, pericardium	Excision	?
1996/Hsu et al	20/F	Chest pain, dyspnoea	Ant. mediastinum/ 14x12x12 cm/ lung	Excision + RT	alive 10 months postop.

SVC – superior vena cava, RT – radiotherapy, CT – chemotherapy

There is still a limited experience in the management of thymic carcinoma. Complete resection of these tumours is sometimes difficult because of the presence of invasion of the mediastinal structures. However, surgical resection should be attempled whenever possible to decrease the tumour burden. The role of postoperative irradiation in the treatment of sarcomatoid carcinoma of the thymus is unknown because of limited experience in this field. In our previous study of thymic carcinoma, we showed that pathological stage, type of resection, postoperative radiotherapy, and cell type did not indicate a significantly favorable result.[2]

We presented a 20-year-old patient with a giant tumour, biphasic histology and with evident disease after surgery. She was believed to be at high risk of recurrence. Hopefully, complete resection and adjuvant radiotherapy in this patients can lead to a more favorable outcome.

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