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Metastasising thymoma

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It is now widely accepted that the term thymoma should be restricted to neoplasms arising from intrathymic epithelial cells. Most thymomas consist of epithelial cells mixed with nonneoplastic lymphocytes. In about 4% of cases tumours consist overwhelmingly of epithelial cells, 1 and when these are of the spindle type electron microscopy may be needed to establish their epithelial nature by showing desmosomes and tonofilament bundles. 2 This report describes the first case of spindlecell thymoma presenting as a splenic tumour. Review of recent published reports revealed three cases of splenic metastasis from thymomas. In our case the diagnosis was supported by ultrastructural studies.

Case report

A 47-year-old man was investigated in November 1970 for a mass in the left hypochondrium associated with anaemia (haemoglobin concentration 9.5 g/dl), which was found to be of iron-deficiency type and was attributed mainly to haemorrhoids. He was given iron and in November 1971 his haemoglobin concentration reached 12 g/dl. He had persistent hypochondrial pain, however, and in February 1972 splenectomy was undertaken at another hospital. The only recorded gross description of the specimen is "A necrotic cystic mass of splenic origin." Histological examination showed a tumour described in the report as a probable haemangiopericytoma, although there was doubt about its exact nature. After splenectomy, the anaemia improved, and the patient was discharged. In March 1979 he reappeared with a nodular mass in the laparotomy scar; this was excised and was found to be histologically similar to the splenic lesion. Concurrently the patient had a persistent anaemia. In November 1980 he was reinvestigated. A chest radiograph showed a rounded shadow in the right cardiophrenic angle; this had been present for several years and was thought to be due to partial eventration of the diaphragm. It appeared, however, to have enlarged slightly during the preceding two years. Computed tomography confirmed that this shadow was of soft tissue density; ultrasound showed that the mass was partly solid and partly cystic. His haemoglobin concentration was 9.5 g/d. Serum electrolyte concentrations were normal and liver function tests and plasma electrophoresis gave normal results. On 6 January 1981 thoracotomy showed a tumour in the anterior mediastinum projecting into the right pleural cavity. The tumour was encapsulated and easily removed. There were, however, separate nodules in the costodiaphragmatic sulcus and palpation also indicated further nodules on the inferior aspect of the diaphragm.

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The tumour was an encapsulated, lobulated mass of slightly firm, greyish-pink tissue $13\times 8\times 4.5$ cm and weighing 180 g. Sections showed sharply defined fibrous septa and occasional small foci of haemorrhage. No cysts or areas of calcification were present. Histologically, the tumour possessed a thick fibrous capsule from which bands extended to produce lobules composed mainly of spindle cells with a few round cells (fig 1). Mitotic figures were almost totally absent. At the periphery of the tumour there were foci of residual thymic tissue. Reticulin impregnation suggested that it was an epithelial neoplasm. The appearances were consistent with a thymoma of predominantly spindle-cell type with scanty lymphocytes. Review of sections from the spleen showed a picture identical to that in the mediastinal lesion.

Blocks from formalin-fixed material were processed appropriately and examined in a Philips 201C transmission electron microscope. Ultrastructurally, most of the tumour cells possessed elongated processes forming in many places complex cellular interdigitations. Many desmosomes were present (fig 2). Tonofilaments, some of which were inserted into well-developed desmosomes, were seen in occasional cells. No myofilaments or Luse bodies were seen. Despite poor fixation, the appearances were considered of diagnostic value and supported the diagnosis of thymoma.

Discussion

The thymus is an epithelial organ which, during development, is infiltrated by lymphocytes and other mesenchymal cells. Thymoma has been defined as a neoplasm of thymic epithelial cells of either spindle or round type. This definition excludes other thymic neoplasms such as malignant lymphomas, germcell tumours and carcinoid tumours.²

About 75% of thymomas are located in the anterior mediastinum. About 15% occupy both the anterior and the superior mediastinal compartments and 6% are situated within the superior mediastinum. The remaining 4% may develop in the neck or in the middle or inferior mediastinum close to the diaphragm. Very rarely is thymoma located in the posterior mediastinum.¹

Most thymomas consist of a variable mixture of epithelial cells and lymphocytes. Pure epithelial cell thymoma is rare and accounts for only 4% of cases. The prevalence of thymoma of predominantly spindle-cell type in two large series varied from 12% to 23% ¹³ Spindle-cell thymomas tend to be slow growing and rarely associated with myasthenia gravis, but they may be associated with red blood cell aplasia and hypogammaglobulinaemia. ²

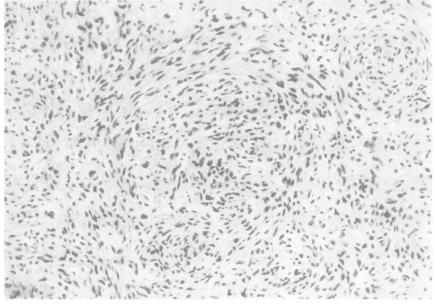


Fig 1 Spindle-cell thymoma (haematoxylin and eosin, $\times 105$).

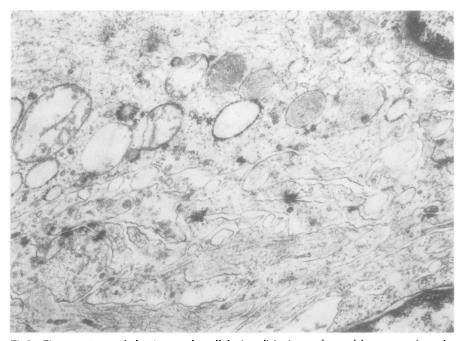


Fig 2 Electron micrograph showing complex cellular interdigitations and several desmosomes (uranyl acetate and lead citrate, \times 22 000).

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Most thymomas behave benignly. The reported incidence of invasive thymoma varies from 7% to 30% and the overall five-year survival varies from 23% to 60%. ²⁴ Metastasising thymoma is uncommon. In a total of 322 cases reviewed the frequency of metastasis was 4%; in most of these cases only lymph nodes were affected. ¹ Haematogenous dissemination of thymoma is rare. Review of recent published reports, however, revealed three thymoma cases in which the spleen was affected. ⁵⁻⁷

None of the three cases reported previously revealed any evidence of myasthenia gravis, red blood cell aplasia, or hypogammaglobulinaemia. In our case there was no evidence of myasthenia gravis or hypogammaglobulinaemia but the patient had a long history of anaemia, which was possibly due to disseminated malignancy. Interestingly, in all four cases with splenic metastasis the tumours were of spindle-cell or spindle-cell and round-cell type.

Although thymomas with a scanty lymphocytic component are thought to be the most aggressive lesions, it is extremely difficult to predict on the basis of histological or electron microscopy examination which will behave aggressively.²³⁶⁻¹⁰

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