Unusual presentation of more common disease/injury

A malignant retroperitoneal mass – A rare presentation of recurrent thymoma

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A 60-year-old man presented with a suspected retroperitoneal mass, after primarily resected thymoma (type B1/B2, Masaoka stage 1). A germ cell tumour was excluded and a diagnostic biopsy was performed. The mass appeared to be a local recurrence of the primary thymoma, for example, a droplet metastasis, progressed to type B3.

BACKGROUND

Thymoma can occur at any age, but has the highest incidence in the fifth and sixth decade. When symptomatic, patients usually present with symptoms attributable to the mediastinal mass such as dyspnoea, cough, chest pain, recurrent infections, or with paraneoplastic symptoms such as those attributable to myasthenia gravis.

Recurrences of thymoma generally are locally, that is, in the mediastinum, the pleurae or lung, due to droplet metastases.^{1 2} Lymphogenous and haematogenous metastases are rare.³

In this paper, we present a patient with a solitary retroperitoneal mass. Finally, this appeared to be a recurrence of the primary thymoma. As far as we know this location of a local recurrence is unusual.

CASE PRESENTATION

A 60-year-old man presented himself at the medical emergency department with acute onset of chest pain. His medical history consisted of a primarily resected thymoma (type B1/B2, Masaoka stage I) 10 years ago, with a local recurrence in the anterior mediastinum 6 years ago treated with resection and adjuvant radiotherapy (60 Grey), no relation with an inducing moment was identifiable. No abnormalities were found on physical examination

INVESTIGATIONS

Laboratory data were within normal range, except for an elevated D-dimer 0, 44 mg/l (0, 0–0, 25 mg/l). Pulmonary embolism was excluded by a spiral chest CT scan, however, a right-sided retroperitoneal mass (9×4×13 cm) was observed (figure 1), that did not appear to have a relation with the lung.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis at that time included sarcoma, lymphoma, germ cell tumor or metastasis of an unknown tumour. A recurrence or haematogenous metastasis of the thymoma seemed unlikely, unless the thymoma had

dedifferentiated.⁴ A fluorodeoxyglucose positron emission tomography scan (figure 2) revealed, besides the retroperitoneal mass, additional hotspots in the thoracoscopic scar, hilar region of the right lung and in the mediastinum (Naruke 4 R). Additional laboratorial tests showed a normal β -hCG (<0,01 IU/l) and α 1-foeto-proteine (5, 2 ug/l (<10,0)), excluding a germ cell tumour. A diagnostic biopsy procedure of the retroperitoneal mass was performed. The histopathology revealed a thymoma type B3 (figure 3) consisting of a combination of atypical, polymorph epithelial cells and small lymphocytes, with a phenotype of immature T cells (TdT positive cells).

By critically reviewing the CT and positron emission tomography-scans finally the retroperitoneal mass appeared to have a very subtle connection with the right pleural cavity (figure 4). Therefore, the mass could be classified as a droplet metastasis of the primary thymoma and should be regarded as a local recurrence.

OUTCOME AND FOLLOW-UP

The tumour was completely resected. There was no indication for adjuvant chemotherapy or radiotherapy.¹

DISCUSSION

In the present paper, we presented the case of a local recurrence of a thymoma at an unusual location.

Thymic epithelial tumours consist of thymoma (encapsulated or invasive) and thymic carcinoma. Tumour stage is the strongest prognostic factor for thymoma. The most widely used is the Masaoka system, in which complete excision, invasive growth into capsule and/or adjacent structures are important features revealing stage I–IV. Traditionally, thymomas are classified on histomorphological grounds in which nowadays also a ‘functional’ classification plays a role. This WHO classification shows a strong correlation of histological subtyping (A, AB, B, B1–3, carcinoma) with staging. Treatment by surgery and additional radiotherapy in cases of incomplete resection, is known to obtain a 5 years survival of 98% in stadium I and

Figure 1 CT scan showing a retroperitoneal mass.

Figure 2 Fluorodeoxyglucose positron emission tomography scan showing a retroperitoneal mass, hotspots in the thoracoscopic scar and hilar region of the right lung.

II.⁵ Meta analysis showed that the recurrence rate for stage I thymoma is 4%.⁶ In recurrences the predominant histological subtype of the primary tumour is B3 or C type.⁴

B3 thymoma, account for approximately 10 to 14% of all thymoma. Morphologically, this tumour type is predominantly composed of epithelial cells that have a round or polygonal shape and that exhibit no or mild atypia. The

epithelial cells are admixed with a minor component of non-neoplastic lymphocytes.

Prognosis according to the WHO classification is based on the histological classification at the first presentation without histological follow-up, although there seems to be some evidence that histological progression occurs in thymoma, the effect on prognosis remains unknown.⁷

Figure 3 (A) Type B1 thymoma. (B) Recurrence with type B3 thymoma (H&E staining).

Figure 4 CT scan showing the retroperitoneal mass with the subtle connection with the right pleural cavity.

Primary therapy involves surgery, even if resection is incomplete. The 5 year survival of a type B3 thymoma in the Netherlands is 53,1% (32, 8–69, 8).⁸ This might increase in the next few years since new therapies are being developed. For example in type B3 and C thymoma imatinib, a tyrosine kinase inhibitor, demonstrated some effect in

two out of seven patients, with the suggestion to select on expression of c-KIT in these thymoma.⁹

Recurrences of thymoma occur locally. Lymphogenous and haematogenous metastases are uncommon.³ A transdiaphragmatic retroperitoneal metastasis has been described only a few times before.^{10 11}

Multiple potential mechanisms for transdiaphragmatic spread of thymoma exist. Since a very subtle connection with the right pleural cavity was found we suspect direct muscle invasion of diaphragm and spread in the retroperitoneal space as the main cause of this recurrent thymoma.

Other possible routes involve tumour spread through the diaphragmatic crura or pre-existing gaps in the anteromedial diaphragmatic origin to the retroperitoneal space.^{10–12}

In the patient, the retroperitoneal mass was suspected not to be a recurrence of the prior thymoma because of its supposed localisation and the histological classification of the primary mediastinal thymoma (B1/B2). However, the biopsy showed a B3 thymoma, compatible with dedifferentiation.

We concluded that the retroperitoneal mass was an unusual local recurrence.

Learning points

- ▶ Local recurrence of thymoma can occur below the diaphragm.
- ▶ Histological dedifferentiation occurs in thymoma.
- ▶ All patients with a suspected retroperitoneal mass should be evaluated carefully.

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Competing interests None.

Patient consent Obtained.

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