CASE REPORT

Giant thoracic mass: an unusual presentation of primary pulmonary Hodgkin's lymphoma

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SUMMARY

Primary pulmonary Hodgkin's lymphoma (PPHL) is rare. PPHL without peripheral lymphadenopathy or hepatosplenomegaly is exceptionally uncommon. We present a 61-year-old woman believed to have a solitary intrapulmonary fibrous tumour on a CT and a CT-guided biopsy, until surgical excision. Histopathology and immunohistochemistry of the excised mass confirmed PPHL. PPHL is a very rare differential diagnosis of large solitary intrapulmonary masses. A CT-guided biopsy is recommended, as it can be diagnostic, reserving excision for cases where the diagnosis remains in doubt.

BACKGROUND

Pulmonary involvement is reported to occur in 15–40% of patients with Hodgkin's disease.¹ However, the presentation of primary pulmonary Hodgkin's lymphoma (PPHL) is very rare, with just over 70 cases recorded internationally between 1927 and 2006.¹

While it is rare, it should nevertheless be considered as a differential diagnosis of lung masses, since a CT-guided biopsy can avoid surgical resection. By describing this case we demonstrate how, despite modern diagnostic techniques, it is still not always possible to make a preoperative diagnosis.

CASE PRESENTATION

A 61-year-old semiretired female therapist presented with a 2-month history of dry cough and breathlessness. B symptoms were not appreciated at diagnosis but on retrospect she had noticed pain in her arm on drinking alcohol. She smoked only five

cigarettes per year. On examination she looked well with no palpable lymphadenopathy. Her breath sounds were equal and normal bilaterally. She had worked in a smoky environment previously. The patient had one relative who had Hodgkin's lymphoma.

INVESTIGATIONS

A chest X-ray demonstrated a large, well-circumscribed mass in the left hemi-thorax (figure 1). A chest CT confirmed a large mass within or invading the left upper lobe (figures 2–4). There was no direct mediastinal invasion but there was thought to be associated mediastinal adenopathy. A CT-guided biopsy was performed which showed fibrous tissue. Germ cell markers were normal.

DIFFERENTIAL DIAGNOSIS

The well-circumscribed nature of the tumour suggested a benign aetiology, which was in keeping with the negative CT-guided biopsy results. A working diagnosis of solitary fibrous tumour or benign intrapulmonary disease was made.

TREATMENT

Given its size it was thought to require excision. This was approached through a clamshell thoracotomy to provide control of proximal vascular structures. A fibrous mass, originating in the lower lobe but rigidly fixed to the pericardium, the upper lobe and the proximal pulmonary artery, was seen. It was removed en bloc by wedge resection, together with the pericardium. The left phrenic nerve was directly involved, and was excised as part of the

Figure 1 Initial chest X-ray demonstrating the large mass in the left hemi-thorax.

Figure 2 Preoperative CT chest demonstrating the large mass which occupied almost one-third of the hemi-thorax, and partially compressed the pulmonary artery.

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Figure 3 Anterior volumetric rendered CT image showing the mass occupying one-third of the left hemi-thorax.

specimen. The pericardium was reconstructed with a large Marlex/Goretex patch. Hilar and ipsilateral mediastinal nodes were sampled.

OUTCOME AND FOLLOW-UP

The tumour was 135×120×95 mm. Histopathology demonstrated a soft tissue mass which was characterised by large areas of sclerosis, within which were nodules of lymphoid cells. The nodules were composed of mononuclear blasts surrounded by a mixed population of mature lymphocytes, eosinophils, plasma

Figure 4 Anterolateral volumetric rendered CT image of the giant thoracic mass.

Figure 5 Postoperative histology high magnification slide (×400) showing strong CD30 expression (using immunohistochemistry) in the Reed-Sternberg cells.

cells and occasional neutrophils. The blasts had multilobated nuclei with prominent nucleoli. Several Reed-Sternberg cells are seen (figure 5). The lymphoid blasts expressed CD30, MUM1, PAX5 and EMA. There was no immunoreactivity for CD20 or CD3 or CD23 and EBV. Background infiltrate expressed CD3, CD20 and CD138 on appropriate cell types. Morphology or immunophenotype excluded possibility of a diffuse large cell B cell lymphoma or a high grade T cell lymphoma. Sclerosing mediastinal B cell lymphoma was excluded. Hilar and mediastinal nodes were not involved, and the tumour was confined to the lung. The appearances and immunophenotyping were in keeping with nodular sclerosing classical Hodgkin's lymphoma.

She made a good postoperative recovery. A postoperative CT showed no evidence of residual disease. Subsequently, she received six cycles of Adriamycin, Bleomycin, Vinblastine and Dacarbazine chemotherapy. A Positron emission tomography-CT (PET-CT) demonstrated that the disease was in metabolic remission after 2 months (ie, after 2 cycles) of chemotherapy. Further CT chests at 10 and 15 months postoperatively confirmed no evidence of active lymphoma. She remains well without evidence of recurrence 16 months later.

DISCUSSION

Pulmonary manifestations of Hodgkin's lymphoma are almost always associated with mediastinal or hilar node involvement.² In contrast, primary pulmonary Hodgkin's lymphoma is described as a separate entity from nodal Hodgkin's lymphoma, and from Hodgkin's which affects the lung secondarily.³

The diagnosis of PPHL requires three criteria to be fulfilled: (1) the disease should be restricted to the lung in which the neoplastic process arises; (2) histological results consistent with Hodgkin's lymphoma should be present; and (3) other possible pathological causes should be excluded.^{1 4}

The majority of patients with PPHL present with 'B-symptoms' (weight loss, night sweats and fevers). The absence of B symptoms, as in this case, has been reported in the literature on fewer than 10 occasions.³

The imaging appearances of PPHL are non-specific. On plain radiographs and a CT these may present as solitary or multiple pulmonary soft tissue nodules, infiltrates or mass lesions in one or both lungs³; cavitation and air bronchograms are well-documented. There is no zonal or lobar predilection of these lesions, but PPHL presenting as a large mass is rare. When very large, as in this case, the anatomical tissue of origin can be

obscured even on CT imaging. The radiological differential diagnosis includes both benign and malignant conditions.

The diagnosis can often be made on histological examination of a CT-guided biopsy. The typical histological appearance is of multiple nodules with irregular margins.

The case we present fulfils the diagnostic criteria for PPHL. The occurrence of a single solid, giant mass, however, is highly unusual. The lack of systemic symptoms is even more unusual.

In this case, a CT-guided biopsy did not make the diagnosis preoperatively, and the patient went on to have major chest surgery. An intraoperative biopsy should be considered when major lung resection (lobectomy or pneumonectomy) will be required for removal, which was not the case with this patient. One previous case report has alluded to the difficulty in obtaining an accurate diagnosis in PPHL with CT-guided biopsy, with the patient being diagnosed with non-small-cell lung cancer.⁵ The case we present demonstrates that a negative preoperative biopsy does not exclude the diagnosis of PPHL.

Learning points

- ▶ Primary pulmonary Hodgkin's lymphoma (PPHL) is a rare diagnosis and can be difficult to make.
- ▶ PPHL should be considered in the differential diagnosis of lung masses.
- ▶ CT guided biopsy—although not in every case—can avoid surgical resection.
- ▶ Intraoperative biopsy should be considered when a major lung resection—lobectomy or pneumonectomy—will be required for removal.
- ▶ PPHL is a condition that can be treated by avoiding surgical resection. Surgery should be reserved for those in whom the diagnosis remains in doubt.

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REFERENCES

- 1 Lluch-Garcia R, Briones-Gomez A, Monzo-Castellano E, *et al*. Primary pulmonary Hodgkin's lymphoma. *Can Respir J* 2010;17:e106–8.
- 2 Diederich S, Link TM, Zühlendorf H, *et al*. Pulmonary manifestations of Hodgkin's disease: radiographic and CT findings. *Eur Radiol* 2001;11:2295–305.
- 3 Radin AI. Primary pulmonary Hodgkin's disease. *Cancer* 1990;65:550–63.
- 4 Kern WH, Crepeau AG, Jones JC. Primary Hodgkin's disease of the lung. Report of 4 cases and review of the literature. *Cancer* 1961;14:1151–65.
- 5 Kumar R, Sidhu H, Mistry R, *et al*. Primary pulmonary Hodgkin's lymphoma: a rare pitfall in transthoracic fine needle aspiration cytology. *Diagn Cytopathol* 2008;36:666–9.

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