Thymic lymphoma—an unusual presentation

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Eiser, N. M. and Samarrai, A. A. R. (1975). Thorax, 30, 588–591. Thymic lymphoma—an unusual presentation. A patient with a thymic lymphoma invading both chest wall and lung is presented. Extensive systemic-pulmonary arterial communications had developed through this lymphoma. Hodgkin's disease should be included as a rare cause of systemic to pulmonary communications.

Hodgkin's disease of the thymus is a well recognized entity and it is known to spread locally to lung parenchyma, mediastinal structures, and the sternum (Steiner, 1943; Marshall and Wood, 1957; Fechner, 1969; Keller and Castleman, 1974). Intrathoracic Hodgkin's disease involving the chest wall has also been reported (Goldman, 1971).

Our patient had nodular sclerosing Hodgkin's disease of the thymus gland invading both the anterior chest wall and the upper lobe of the left lung. Extensive communications between branches of the left internal mammary artery and the vessels of the left upper lobe had developed through this lymphoma. The lesion was brought to light following trauma.

CASE HISTORY

A 22-year-old Persian nurse was well until six weeks before admission when she sustained an accidental blow to the sternum. Three hours later she had an haemoptysis and two weeks later a further three haemoptyses. A tender swelling developed at the left parasternal edge. In Iran, nine months previously, a routine chest radiograph and full blood count had been reported normal, although the ESR was 100 mm/hr and the Mantoux test was positive. Three months later her brother died of Hodgkin's disease.

Examination on admission revealed a healthy but pale young woman with facial hirsutism. There was a small, non-pulsatile, tender, soft tissue swelling at the left sternal edge in the second intercostal space. Neither lymph nodes, liver nor spleen were palpable. The peripheral pulses were normal and the blood pressure, in both arms, was 90/50 mmHg. A well localized continuous mur-

mur was heard just below the swelling on the anterior chest wall.

INVESTIGATIONS The haemoglobin was 9.9 g/dl, MCV 74 fl, MCH 24.6 pg, and ESR 91 mm/hr. The total and differential white cell counts were normal but the platelet count was 620,000/ μ l. The electrocardiogram was normal, but the chest radiograph (Fig. 1) showed a lobulated, cavitated mass, 6 cm in diameter, anteriorly in the left upper zone. Bronchoscopy was normal apart from some reddening of the mucosa and distortion of the bronchial walls at the bifurcation of the anterior and apicoposterior branches of the left upper lobe bronchus.

A selective left internal mammary angiogram demonstrated an abnormally dilated artery, along whose length a number of dilated feeder vessels supplied a large pathological circulation communicating with the upper lobe branch of the left pulmonary artery (Figs 2 and 3).

It was thought that this communication was a pre-existing vascular malformation brought to light by trauma. To verify the nature of this lesion and to institute appropriate treatment, a left thoracotomy was performed on 5 July 1974. A tumour was found involving the upper lobe of the left lung and the anterior mediastinum, with the left internal mammary artery disappearing into it. The mediastinal tumour was resected together with the left upper lobe and all enlarged hilar nodes. The patient made an uneventful recovery.

Histology The mediastinal part of the tumour was thymus, largely replaced by collections of lymphocytes, plasma cells, lacunar cells, giant cells, and occasional Sternberg-Reed cells, separated by

FIG. 1. PA and left lateral chest radiographs showing a lobulated cavitated mass anteriorly in the left upper lobe.

FIG. 2. Selective left internal mammary angiogram demonstrating an abnormally dilated artery communicating with a pathological circulation.

dense fibrous tissue. Blood vessels were involved in the process. The main pulmonary vessels disappeared into lobulated yellow tissue with ill-defined edges and central necrosis which replaced the left upper lobe and which showed the same microscopic features as the thymic tumour. Reactive hyperplasia only was found in the hilar lymph nodes. The histology, therefore, was of nodular sclerosing Hodgkin's disease involving the thymus and the left upper lobe.

Further investigations were performed for staging purposes. Abdominal lymphangiography was normal. Laparotomy and splenectomy were performed. The spleen weighed 110 g and was free of Hodgkin's disease; biopsies of para-aortic lymph nodes and the liver were also negative. A ^{67}Ga scan showed abnormal uptake suggestive of residual lymphoma in two areas in the anterior mediastinum just below the level of the manubrium sterni, and a destructive lesion on the posterior surface of the upper end of the sternum was seen on lateral tomography.

The patient's final stage was determined as II_{EA} (Carbone *et al.*, 1971). She was given post-

operative radical irradiation, using an upper mantle technique, and additional chemotherapy with nitrogen mustard, vincristine, procarbazine and prednisone.

COMMENT

It has been proposed (Lowenhaupt and Brown, 1951; Lattes, 1962) that the lesion of the thymus previously described as 'granulomatous thymoma' or 'Hodgkin's thymoma' is a primary thymic epithelial neoplasm, which loses its thymic epithelial features and is indistinguishable from Hodgkin's disease when it spreads to lymph glands. This seems improbable since the lesion, unlike other thymomas, responds better to irradiation than to surgery and has never been associated with myasthenia gravis. Thompson (1955) believed that thymic epithelial neoplasm was the primary lesion in Hodgkin's disease, but necropsy studies of 86 cases of Hodgkin's disease (Marshall and Wood, 1957) revealed that only 26% had gross thymic involvement. It appears now that 'granulomatous thymoma' is Hodgkin's disease whose histology is modified by a unique response of the thymic epithelium (Fechner, 1969; Keller and Castleman, 1974).

FIG. 3. Angiogram revealing high pressure filling of the upper lobe branch of the left pulmonary artery.

Hodgkin's disease is not often limited to the anterior mediastinum at initial diagnosis, but Keller and Castleman (1974) reported 44 patients where this was the case. The thymus alone was affected in 10 patients whose five-year survival of 80% was comparable with that of patients with stage I nodular sclerosing Hodgkin's disease at any site. However, three of the five cases whose thymic disease had spread into adjacent lung were dead within five years. Our patient had local spread to lung, sternum and chest wall. Musshoff et al. (1968) report a remission rate of 92% in patients with local non-lymphatic spread but only 62% where non-lymphatic spread is disseminated.

Most workers now agree that postoperative irradiation should be given in thymic Hodgkin's disease since there is a high recurrence rate after surgery alone (Katz and Lattes, 1969). Extended field radical irradiation has been shown to give a better survival rate in patients with nodular sclerosing Hodgkin's disease (Kaplan and Rosenberg, 1966; Keller et al., 1968; Keller and Castleman, 1974). In his review of six patients with intrathoracic Hodgkin's disease, presenting with chest wall masses, Goldman (1971) concluded that in such cases, particularly when local bone is eroded, full rather than palliative irradiation should be given. Our patient was treated both with radical irradiation, in view of the spread from the thymus to the sternum and chest wall, and with chemotherapy because of the adjacent lung infiltration.

Initially our patient was thought to have a traumatic internal mammary arteriovenous communication. Subsequent angiography demonstrated an extensive pathological circulation connecting the left internal mammary artery to the left pulmonary artery, more suggestive of a pre-existing vascular malformation. However, at thoracotomy the pathological circulation was found to be secondary to a tumour. This proved to be nodular sclerosing Hodgkin's disease of the thymus whose spread to lung and chest wall had involved the left pulmonary artery and left internal mammary artery.

A similar case had recently been reported (Dunn and Wexler, 1974) but these authors state that the origin of the Hodgkin's disease was lung parenchyma not thymus.

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