Coexisting retroperitoneal and mediastinal fibrosis

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Summary

A rare case of coexisting retroperitoneal and mediastinal fibrosis is reported. Increasing awareness of this association may lead to earlier recognition of significant symptoms and more effective therapy.

KEY WORDS: fibrosis, retroperitoneum, mediastinum.

Introduction

The clinical features of retroperitoneal fibrosis (Lepor and Walsh, 1979) and mediastinal fibrosis (Light, 1978) are protean. The aetiology of the majority of cases is unknown, but coexistence of these two rare conditions and their association with other fibrotic disorders suggest that they may be different manifestations of a single disease (Comings *et al.*, 1967). In some reported cases of combined retroperitoneal and mediastinal fibrosis, complications of mediastinal disease appeared many years after the diagnosis of retroperitoneal fibrosis, or vice versa. However, in most cases, including the patient reported here, additional fibrotic lesions in the thorax or the abdomen were chance findings at necropsy. Nevertheless, awareness of the association may lead to earlier recognition of complications at the other site and possibly more effective treatment.

Case report

The patient underwent a left pyelolithotomy in 1961, aged 54 years. In 1969, he presented with bilateral loin pain and anorexia. An intravenous urogram showed bilateral hydronephrosis, confirmed by retrograde ureteropyelography. The latter also showed constriction of both ureters at the L4/5 level, proximal dilatation, and medial deviation of the lower right ureter consistent with retroperitoneal fibrosis. At operation, a fibrous plaque extending from the duodenum to the pelvic brim involving both

ureters was found. Bilateral ureterolysis was performed. Histology confirmed retroperitoneal fibrosis. There was no prior exposure to any drug that could be incriminated. He made an excellent postoperative recovery, and follow-up intravenous urograms were satisfactory.

In 1977, the patient developed angina, treated with nitrates. Four years later, he became dyspnoeic and lost weight. In April 1981, subendocardial myocardial infarction, functional mitral incompetence and left ventricular failure were diagnosed. His chest radiograph revealed pulmonary oedema and a left hilar mass which tomography suggested was due to vascular structures.

In November 1981, he was readmitted with dyspnoea, left ventricular failure and a moderate-sized left pleural effusion.

Investigations showed a mild normochromic, normocytic anaemia. Pleural fluid aspirated from the left chest had a protein content of 29 g/litre. It contained a few groups of dark, round, reactive mesothelial cells. However, a pleural biopsy was interpreted to show oat cell carcinoma with associated lymphocytic inflammatory reaction. Fibreoptic bronchoscopy was normal.

Despite his poor condition, it was decided that chemotherapy might be beneficial and he was given adriamycin/vincristine followed by methotrexate/cyclophosphamide. Over the next 3 weeks, he deteriorated rapidly and died.

At necropsy, no tumour was found in any intrathoracic structure. The aorta was surrounded by dense fibrous tissue up to 0.5 cm thick extending from the ascending portion to the arch. The origins of the pulmonary arteries were similarly encased but the pulmonary veins were spared. The pelvic and left-sided retroperitoneal tissues also showed dense fibrous thickening which surrounded the left ureter and iliac vessels. The left kidney was hydronephrotic and its ureter was dilated proximally. Histologically,

the affected mediastinal and retroperitoneal tissues showed dense hyaline fibrosis. The left kidney also showed some interstitial fibrosis, and the right kidney had changes of chronic pyelonephritis.

Discussion

Mediastinal fibrosis was discovered at necropsy in this patient 13 years after the initial diagnosis of retroperitoneal fibrosis. Although oat cell carcinoma is notoriously difficult to diagnose on pleural biopsy, chemotherapy was administered because it is potentially beneficial. No tumour was found at post-mortem examination, and the patient probably died of ischaemic heart disease and intractable cardiac failure.

Coexisting retroperitoneal and mediastinal fibrosis is rare. Since the initial report by Tubbs (1946), probably fewer than 40 cases have been recorded in the medical literature, although the true incidence is probably higher (Morgan, Loughridge and Calne, 1966). The fibrotic lesions are usually anatomically separate, but fibrosis extending continuously from the retroperitoneum to the mediastinum has been described in at least 2 cases (Cameron *et al.*, 1961; Light, 1978). Most patients had symptoms referable either to the abdomen or the thorax only, but a substantial proportion had clinical features of disease in both sites. The commonest presenting feature of retroperitoneal fibrosis is pain associated with obstructive uropathy (Lepor and Walsh, 1979); while that of mediastinal fibrosis is superior vena caval obstruction (Light, 1978). However, many other structures such as the inferior vena cava, iliac vessels, pulmonary vessels, tracheobronchial tree, oesophagus and coronary artery may be directly involved, giving rise to protean manifestations.

In addition, association with auto-immune disorders and other inflammatory fibrotic lesions in anatomically remote sites have been described. Examples of the latter include Riedel's thyroiditis, pseudotumour of the orbit and sclerosing cholangitis (Comings *et al.*, 1967). The term 'multifocal fibrosclerosis' has been used to describe various combinations of these conditions.

There are many reported causes of retroperitoneal fibrosis (Manna *et al.*, 1981) amongst which tuberculosis and histoplasmosis can also cause mediastinal fibrosis. Nevertheless, in most patients, the aetiology is unknown. In these idiopathic cases, treatment with steroids in the early stages has been reported to be effective, whereas in later stages, surgery to relieve obstruction may be the only alternative (Longmire, Goodwin and Buckberg, 1967).

Several possible mechanisms may be responsible

for dyspnoea in mediastinal fibrosis. These include tracheal or bronchial compression, fibrotic narrowing of coronary arteries leading to ischaemic heart disease and cardiac failure, obstruction of main pulmonary arteries, obstruction of main pulmonary veins, thromboembolic occlusion of small intrapulmonary arteries and pulmonary interstitial fibrosis (Arnett *et al.*, 1977).

Although this patient's left coronary artery was narrowed, and his pulmonary arteries encased, the degree of obstruction attributable to mediastinal fibrosis was not sufficient to account for his dyspnoea and intractable heart failure. He probably died from unrelated ischaemic heart disease. Nevertheless, since some of the above mechanisms are potentially treatable, further investigation of such patients may be warranted. Finally, in any patient with either retroperitoneal or mediastinal fibrosis, one should be alert to complications arising from other sites which may also be affected by the fibrotic process.

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