Bi-atrial myxomas presenting as recurrent pulmonary emboli in a girl

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Summary

A 14-year-old girl whose sole presenting features were symptoms of pulmonary embolism, was found to have bi-atrial myxomas. The diagnosis was made pre-operatively, and the patient had a successful outcome following elective surgery. The absence of other features such as systemic embolism, atrioventricular valvular obstruction, fever and constitutional symptoms makes this a most unusual case. Right atrial myxoma should be considered in any patient presenting with pulmonary embolism for which there is no obvious source.

KEY WORDS: myxoma, pulmonary embolism, lentigines.

Introduction

Myxomas account for 50% of primary cardiac tumours, and characteristically present with one or more of the following: symptoms of atrio-ventricular valvular obstruction, systemic embolism, non-specific constitutional symptoms, such as fever and weight loss, together with anaemia, an elevated erythrocyte sedimentation rate (ESR) and raised serum gamma-globulin (Sutton *et al.*, 1980). About 75% of myxomas are found in the left atrium, 20% in the right atrium and 5% in the ventricles (Goodwin, 1963). Although there have been twenty-two reported cases of bi-atrial myxomas (Sutton *et al.*, 1980; Imperio *et al.*, 1980; Fitterer, Spicer and Nelson, 1976; Herpin *et al.*, 1980; Carvalho *et al.*, 1980), the majority of these were not diagnosed until autopsy, or unexpectedly at exploratory surgery.

We report an unusual case of bi-atrial myxomas in a patient who presented solely with recurrent pulmonary emboli, and had successful surgery following pre-operative diagnosis.

Case report

A 14-year-old girl was admitted to St Vincent's Hospital in July 1981, with a history of recurrent bilateral pleuritic chest pain on three separate occasions over the preceding 6 months. She also complained of loss of energy for the same period of time. Her past history was unremarkable.

General examination was normal, but multiple unusual dark brown freckles were noted around the nose and malar areas. Cardiovascular findings were variable but at different times the following were noted: a third heart sound, a grade 2/6 'to and fro' systolic murmur, best heard at the left sternal border, and an evanescent friction rub.

The erythrocyte sedimentation rate (Westergren) was elevated to 21 mm in the first hour, but fell to normal pre-operatively. Haemoglobin concentration was normal. Liver function tests showed a slight elevation of all hepatic enzymes. An electrocardiogram was normal and a chest X-ray revealed a small right-sided pleural effusion. A ventilation-perfusion scan showed perfusion defects in both lung fields, and normal ventilation (Fig. 1)—a pattern suggestive of multiple pulmonary emboli. Serological markers for collagen-vascular disease were absent, but immune electrophoresis revealed an elevated IgG level of 22.1 g/litre (normal range 8—18 g/litre), with normal IgM and IgA levels. Doppler and isotope venograms showed normal venous drainage from both lower limbs, and ultrasound examination of the pelvis was normal. One-dimensional echocardiography revealed a sonic mass in the left atrium, and a similar lesion in the right atrium (Fig. 2). Angiocardiography showed a left atrial filling defect, and in addition, a mass lesion in the right atrium. These

FIG. 1. Ventilation (Krypton 81-m) perfusion lung scan, showing multiple perfusion defects (arrows) to both lungs and normal ventilation suggestive of multiple pulmonary emboli.

findings were confirmed by two-dimensional echocardiography.

At surgery, a right cardiectomy revealed a large soft myxoma, filling the right atrium and extending into the inferior vena cava and tricuspid valves. The interatrial septum was opened widely, disclosing a further myxoma in the left atrium. Both tumours took origin from the septum, close to the foramen ovale. The common base of both myxomas was widely excised. The septum was sutured transversely, and a dacron graft was not required. Subsequent histology revealed a typical atrial myxoma. The patient made an uneventful postoperative recovery.

Discussion

Obstruction to blood flow at the atrio-ventricular valve, peripheral embolism and systemic upset account for the majority of the clinical features of atrial myxomas (Sutton *et al.*, 1980). Although occasional patients with myxomas are asymptomatic (Sutton *et al.*, 1980); Morgan *et al.*, 1977), this is unusual. Up to 75% of cases (Buckley and Hutchins, 1979) occur in females, and the majority are diagnosed in the fifth or sixth decades of life. A family in which one parent and three out of seven siblings had

atrial myxomas has also been described (Siltanen *et al.*, 1976).

Diagnosis before surgery or death is unusual because of the non-specific nature of the symptoms and the unobtrusive and subtle cardiac signs. Such patients frequently present to the general physician or the neurologist, rather than the cardiologist, and the diagnosis is unlikely to be made, unless the condition is actively considered.

There are a number of unusual features to the case we have described. In the first place, bi-atrial myxomas causing recurrent symptomatic pulmonary emboli in the absence of peripheral systemic embolism is unusual. In fact, recognized embolism is much more frequent in left (60%) than in right-sided tumours (8–10%) (Meller *et al.*, 1977). Secondly, the lack of systemic symptoms, such as malaise, weight loss, pyrexia and arthralgia, which occur in up to 90% of cases (Goodwin, 1963), is remarkable, and this is reflected in the relatively minor abnormalities in the ESR and serum gammaglobulins. Thirdly, there were no symptoms of atrio-ventricular valvular obstruction. Despite the fact that the right atrial tumour appeared to fill the entire atrial cavity, and was seen angiographically to move freely within the right atrium, the patient never had syncope, and did not exhibit signs of cardiac failure. The friction rub noted

FIG. 2. M-mode echocardiogram showing right atrial myxoma. R.V.=Right ventricle. R.M.=Right atrial myxoma behind the anterior leaflet of the tricuspid valve.

in this case is an uncommon auscultatory finding. It has been suggested that this is caused by movement of the myxoma against the endocardium (Frankenfeld, Waters and Steiner, 1960). There was no ECG nor echocardiographic evidence of pericarditis. Finally, the prominent facial freckling is worthy of comment. This has previously been described in two patients with bi-atrial myxomas (Frankenfeld *et al.*, 1960; Dashkoff *et al.*, 1978), and in addition, lentiginosis (Rees, Ross and Keen, 1973), and progressive skin pigmentation (Kendall and Symonds, 1952) have been described in patients with left and right atrial myxomas respectively. It is possible that such dermatological signs occur more frequently, but are not reported.

In 1951, Prichard stated that 'the diagnosis of cardiac tumours is either impossible, or a matter of chance', and that such tumours 'present a dismal diagnostic prospect'. This, however, is now no longer the case, and the reasons are well illustrated by the

case we have described. The introduction of echocardiography and angiocardiology is largely responsible for the increasing numbers of myxomas which are now diagnosed during life and pre-operatively (Donahoo *et al.*, 1979; Attar *et al.*, 1980). Angiocardiography, while valuable, can occasionally be hazardous (Pindyck *et al.*, 1972), and give false positive and false negative results (Martinez, Giles and Burch, 1974). For these reasons, echocardiography, and in particular two-dimensional echocardiography is now the preliminary investigation of choice for atrial myxomas (Donahoo *et al.*, 1979). The importance of accurate pre-operative diagnosis is reflected in the increasingly favourable surgical outcome, following the introduction of these imaging techniques (Dashkoff *et al.*, 1978).

Once the diagnosis has been established, surgery should be performed as soon as possible (Attar *et al.*, 1980), with complete removal of the myxomas, including wide excision of their base (Kabbani and Cooley, 1973). Although myxomas may develop in any part of the atria, they most commonly arise, as in our patient, in the region of the foramen ovale, and if bi-atrial, are usually attached to the opposite sides of the inter-atrial septum (Imperio *et al.*, 1980).

At operation, it is important to look for secondary valvular damage caused by the tumour, especially with right atrial myxomas (Attar *et al.*, 1980). There was no evidence of tricuspid or mitral dysfunction in our patient, and she had a satisfactory outcome to surgery.

There have been several reports (Attar *et al.*, 1980; Richardson *et al.*, 1979) of recurrence of atrial myxomas, following surgery, although the true incidence is difficult to ascertain, due to the lack of long-term post-operative follow-up studies. The reported rates vary from 4% to 14% (Sutton *et al.*, 1980; Richardson *et al.*, 1979), and have been described as early as 6 months, and as late as 90 months postoperatively (Richardson *et al.*, 1979). Our patient has remained well since surgery was performed 16 months ago, and there is no evidence of recurrence so far. We plan to review her regularly with serial echocardiography.

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(Accepted 10 March 1983)