



Sub-Acute Bowel Obstruction Preventing Sudden Cardiac Death: A Case of Synchronous but Unrelated Left Atrial Myxoma and Colorectal Adenocarcinoma

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Abstract

Primary cardiac tumours are an extremely rare entity, which may be completely asymptomatic or present with a wide range of clinical symptoms and signs such as congestive heart failure, murmurs, stroke or arrhythmias.

We discuss the case of a 53 year old with synchronous but unrelated left atrial myxoma and colorectal adenocarcinoma. To our knowledge, this is the first report in the literature of a patient with an atrial myxoma incidentally discovered as part of staging following a diagnosis of colorectal cancer.

Introduction

Although left atrial myxomas are the most commonly encountered benign neoplasm by the cardiac surgeon, they remain relatively rare. The commonest neoplasms of the heart are metastatic (autopsy frequency 1.5-21%). Primary cardiac tumours have an autopsy frequency of 0.001%-0.28%. Three-quarters of these are benign and three-quarters of these are myxomas [1]. Myxomas most commonly occur in the left atrium (90%) and are generally attached to the atrial septum in or adjacent to the fossa ovalis. The incidence is greatest in those aged 30-60 years, with a notable female preponderance [2].

Carney's complex, a neuroendocrine-cardiac syndrome is a common cause of familial recurrent cardiac myxomas. Other clinical features include pigmented skin lesions, Schwannomas and multiple recurrent mucocutaneous myxomas. Endocrine neoplasms and/or overactivity may also be present [3].

The presentation of myxomas is variable. Asymptomatic myxomas may be picked up incidentally on routine imaging e.g. echocardiography, computed tomography (CT). Others may present with dyspnoea, palpitations, heart murmurs, syncope, pulmonary hypertension, pulmonary oedema or constitutional symptoms. Notably, it often presents late with myxomatous thrombo-embolisation manifesting as strokes or end-organ dysfunction [2,4].

Diagnosis of cardiac myxomas is usually made using Transthoracic Echocardiography (TTE), which has a sensitivity of 95.2% [2]. However, trans-oesophageal echocardiography although more invasive, remains the gold standard for diagnosing cardiac myxomas with a sensitivity approaching 100% [5]. In a large cohort study with 112 patients, less than 2% of left atrial myxomas were diagnosed using CT or MRI [4].

We believe this is the first reported atrial myxoma in the literature, which was diagnosed incidentally on cross-sectional imaging during staging for colorectal cancer. Various similar rare associations have been reported in the literature including cardiac metastases from colorectal cancer [6,7], synchronous atrial myxomas with a small bowel myxoma [8] and adenosquamous lung carcinoma [9].

Case Presentation

A 53 year old woman presented to the A&E department with symptoms and signs of sub-acute bowel obstruction. Her past medical history significantly included hypertension and a previous appendicectomy. She reported a family history of lung cancer (mother). She also demonstrated no evidence of Carney complex phenotype on clinical assessment.

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Figure 1: Contrast CT shows thickening of ascending colon suspicious for malignancy.



Figure 2: CT Thorax shows 6 cm mass within the right atrium.

A contrast CT Abdomen/Pelvis identified a dilated caecum and small bowel with thickening of the ascending colon suspicious for malignancy (Figure 1). Colonoscopy demonstrated an ulcerated, obstructing stricture of the ascending colon which appeared malignant. She subsequently underwent an urgent right hemicolectomy to relieve the obstruction and tissue was sent for histopathological diagnosis. Histology confirmed Dukes C1 (pT4b N2 - 8/24 positive lymph nodes) moderately differentiated adenocarcinoma extending onto the peritoneal surface.

Following MDT discussion, adjuvant chemotherapy was offered to the patient. A staging PET – CT/CT Thorax was undertaken which revealed a 6 cm abnormal mass involving the left atrium (Figure 2). A trans-oesophageal echocardiography (TOE) was organised to characterise this mass further. This confirmed the likelihood of a left atrial myxoma (Figure 3). The patient reported no significant symptoms associated with this other than occasional shortness of breath on exertion which was steadily worsening over the previous six months. Clinically she had no murmurs or signs of congestive cardiac failure.

In view of the cardiac and embolic risks associated with this large myxoma, a decision was made to proceed with urgent surgical resection. Access was gained via midline sternotomy and the pericardium lifted to obtain optimal exposure of the heart in the mediastinum.

Cardiopulmonary bypass was established via bi-caval cannulation. Cardioplegia was delivered using an aortic root vent. A standard aortic cannulation was performed and the myocardial protection strategy involved cooling to 32 degrees Celsius. Cold blood cardioplegia was delivered at twenty minute intervals.

The left atrium was carefully incised and dissected allowing

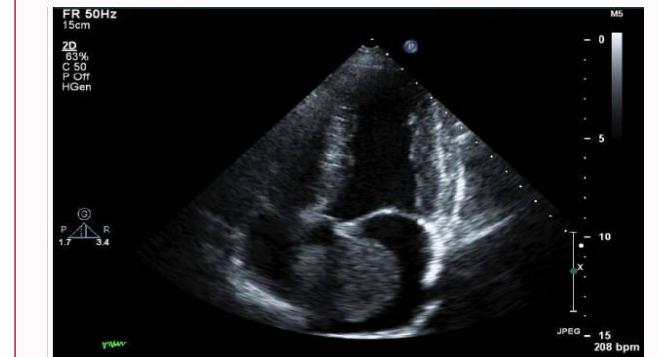
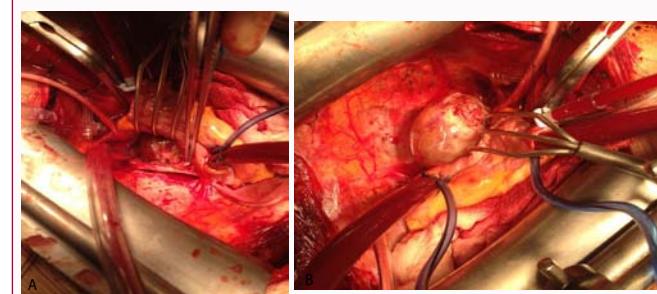


Figure 3: TOE performed to characterise the left atrial mass demonstrates likely atrial myxoma.



Figures 4a and 4b: Intra-operative photographs of left atrial myxoma prior to excision.



Figure 5: Resected atrial myxoma.

the tumour to be identified and exposed. The tumour was carefully removed en-bloc to prevent embolisation leaving an intact left atrial wall and septum. Notably it was tethered to the posterior left atrial wall on a broad stalk, with minimal involvement of the inter-atrial septum (Figures 4A and B).

Following complete excision (Figure 5), primary closure of the left atrium was undertaken and the patient re-warmed. The patient was weaned off cardiopulmonary bypass following standard decannulation. Haemostasis was achieved thereafter and the chest closed using sternal wires.

The patient was transferred to the Intensive Care Unit (ICU) post-operatively in normal sinus rhythm without any inotropic support.

She was extubated after a period of observation and subsequently stepped down to the general ward before being discharged home on day five post operatively without any complications.

Subsequent tumour histopathology confirmed an atrial myxoma. No issues were identified at out-patient follow-up 6 weeks post operatively. Chemotherapy was scheduled as planned for the colorectal adenocarcinoma.

Discussion

Surgical resection remains the mainstay of treatment with cardiac myxomas and operative outcomes are excellent with recurrence rates of about 5% [1]. Recurrence rates are significantly higher in patients with Carney's complex (22%) and this highlights the importance of genetic screening in picking up high risk patients [4].

There is an increasing role for genetic screening in patients diagnosed with cardiac myxomas. Mutations in the PRKAR1 – Alpha gene sequence have been known to account for Carney's complex which results in recurrent atrial myxomas, amongst several other clinical features. These patients also have a predisposition to develop tumours of the ovary, pituitary, adrenal, and thyroid glands [3,11]. The use of other novel genetic techniques – genotyping, SNPs, gene profiling of patients along with immunocytochemistry analysis of tissue may also help ascertain whether certain patients with myxoma are pre-disposed to extra-cardiac tumours.

When evaluating the symptoms related to atrial myxoma in this case, it is important to emphasise that the 6 month history of progressively worsening shortness of breath was not elicited nor thought of as relevant by the patient during the initial clinical assessment. In hindsight, following the incidental diagnosis of an atrial myxoma, this would have been a clinically important symptom.

This case report therefore shows how fortunate our patient was to have cross-sectional imaging as part of her staging work-up for colorectal cancer. For instance, had our patient not presented with sub-acute bowel obstruction secondary to a colorectal adenocarcinoma, she may have presented at a later point in life with potentially life threatening or life altering clinical sequelae associated with the atrial myxoma e.g. stroke (secondary to embolic phenomena), arrhythmias/sudden death or heart failure.

Synchronous cardiac myxomas with other neoplasms have been reported in the literature. [8,9] However there is only one other case report that describes asynchronous cardiac myxoma and colorectal cancer and the patient presented with a combination of gastrointestinal and cardiac symptoms and signs [10]. In contrast, our patient's diagnosis of an atrial myxoma was purely incidental with the cardiac pathology being discovered after extra-cardiac symptoms were investigated and treated, rather than concurrently or as the initial diagnosis.

Other case reports describe colorectal cancer with metastatic disease to the heart and notably metastatic cardiac neoplasms are far commoner than primary cardiac neoplasms [2,6,7].

However with the latter, this case report highlights whether or not patients presenting with cardiac myxomas should be screened for extra-cardiac tumours, as there is little clear evidence for this in the literature, and therefore may well be an area of research for the future.

Finally another interesting issue relates to the follow-up of patients with cardiac myxoma. Currently in the UK, patients are discharged back to their referring cardiologist from the cardiothoracic surgical clinic after being usually seen within 4-6 weeks post-operatively. It may well be prudent that these patients be discussed at a regional MDT with oncology input to identify whether they would benefit from further imaging or investigations as part of extra-cardiac screening.

There remains a lot of interesting, unanswered questions in cardiac myxoma management. This is partly because of the low incidence of this disease entity. Future studies should focus on links with extra-cardiac tumours and associated screening, and understanding underlying genetic mechanisms to guide optimal post-operative follow up and treatment.

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