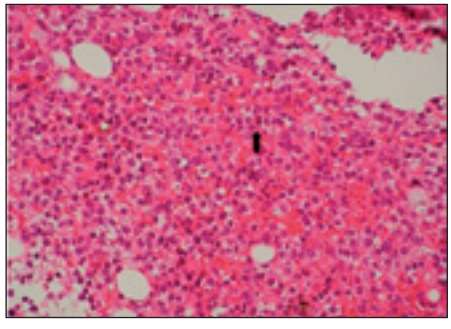
Right atrial myxomas and hairy cell leukaemia: coincidence or causal relationship?

Introduction

Atrial myxomas are the most common primary cardiac tumour (40–50%) (Reynen, 1995). Approximately 90% are solitary and pedunculated and 75–85% occur in the left atrium.

Most cases are sporadic and are most commonly attached to the interatrial septum. When pedunculated, atrial myxomas may prolapse across and obstruct the mitral or tricuspid valves (Ramsdale et al, 1986).

Figure 1. Bone trephine shows extensive, lymphoid cell infiltrate. The cells have scanty cytoplasm and small nuclei, which often appear to be surrounded by a clear halo (arrow). Immunocytochemistry (not shown) was compatible with hairy cell leukaemia.



Discussion

Hairy cell leukaemia is an uncommon, chronic B-cell lymphoid leukaemia and accounts for 2% of all leukaemias (Bouroncle et al, 1958). Cladribine can lead to a complete remission in over 90% of patients (Piro et al, 1990). Patients with hairy cell leukaemia are thought to have an increased risk of secondary tumours of different haematopoetic and non-haematopoetic types (Kampmeier et al, 1994; Kurzrock et al, 1997).

Figure 2. Echocardiography showed a large ovoid mass (twin red arrows) within the right atrium and a smaller pedunculated mass attached low down on the interatrial septum (green arrow). MYX=myxoma.

In a study of 117 patients with hairy cell leukaemia, 36 (31%) patients had an additional malignancy, with a total of 44 separate neoplasms (Au et al, 1998). Some presented before, some after and some (as in this case) concurrently with the diagnosis of hairy cell leukaemia. The majority of tumours were adenocarcinomas (gastrointestinal tract, prostate and breast), squamous cell carcinomas (lung),

Figure 3. Opening the right atrium reveals a large mass attached by a short stalk to the free wall of the right atrium.

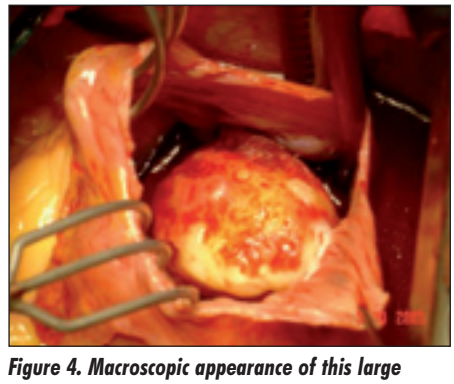


Figure 4. Macroscopic appearance of this large myxoma.

Case Report

A 69-year-old man presented with a 6-week history of progressive breathlessness, fatigue and epistaxis. There was no significant past medical history apart from atrial fibrillation diagnosed 2 years earlier when echocardiography was normal. On examination, he was anaemic and had a new mid-systolic murmur. A full blood count revealed pancytopenia (haemoglobin=10.9 g/dl; white cell count=1.7×10^9/litre; platelets=52×10^9/litre) and bone marrow histology confirmed a diagnosis of hairy cell leukaemia (*Figure 1*). A 'staging' computed tomography scan of the thorax and abdomen showed no lymphadenopathy but splenomegaly and a mass in the right atrium. Transthoracic echocardiography confirmed the presence of a large right atrial mass, attached to the atrial free wall and a small pedunculated mass attached low on the interatrial septum (*Figure 2*). Both right and left ventricular contractility were impaired and there was mild tricuspid regurgitation.

He was treated with intravenous cladribine (2-chlorodeoxyadenosine) to a total dose of 0.9 mg/kg. His blood count recovered to normal levels over the next 3 months and the splenomegaly resolved. There was, however, no change in the size of the right atrial mass which was then removed surgically.

At open heart surgery, both the small pedunculated tumour and the larger, ovoid, more sessile, gelatinous right atrial mass (60 mm diameter) were excised (*Figures 3* and *4*). Histology showed fusiform cells in a myxoid stroma typical of myxoma (*Figure 5*), with the larger mass possessing laminated thrombus and degenerative changes.

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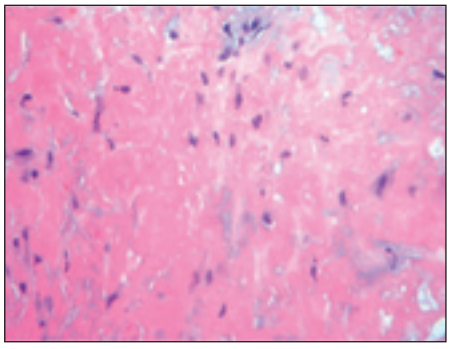

Figure 5. Histology shows much myxoid stroma with stellate/fusiform-shaped cells.

melanoma and non-melanoma skin cancers and sarcomas.

This case appears to be the first atrial myxoma occurring in association with hairy cell leukaemia. It is important clinically to ensure meticulous follow up of patients with hairy cell leukaemia for early detection and treatment of other malignancies. Such follow up should include cardiac physical examination and echocardiography.

Conclusions

Hairy cell leukaemia is associated with an increased frequency of second malignancies. This is the first reported association with atrial myxoma. Vigilance for malignant tumours is essential during follow-up of patients with hairy cell leukaemia. **BJHM**

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No births tonight

What words can I find to describe Channel Five's *Birth Night...Live!* (8 Oct). How about banal, superficial, saccharin, and boring. Previewers had worried about the ethics of showing a live natural birth, and how even an uncomplicated birth might frighten young mothers-to-be, never mind the risk of something going horribly wrong. As it turned out, only the continuity went wrong, and more than once at that.

But 2 hours live in the obstetric unit of the Queen's Medical Centre, Nottingham, failed to produce a natural birth at all. There was one routine section – done at 8.30 pm on a Sunday night – but that was it. They missed Esme Grace, born by water birth before the programme started, and all the other births were caesareans filmed elsewhere.

Two hours was not really 2 hours. There were the welcome advert breaks, and interspersed with Gabby Logan breathlessly saying how exciting it all was, and how a birth was literally minutes away, there was a fair amount of time taken up with describing some common obstetric problems: twin-to-twin transfusion treated by intrauterine laser, monochorionic quads, and congenital diaphragmatic hernia treated by intrauterine endoscopic placement of a tracheal balloon. One can forgive the poor mum for describing the hernia as 'diaphrammatic', but Logan happily parroted this.

One of Logan's fellow presenters was Penny Lancaster. She is the fiancée of Rod Stewart, and introduced an air of *Hello!*-on-the-television to the proceedings. Mostly she featured in birth rooms with mums awaiting their next contraction, but nothing much ever happened so it was 'Back to Gabby' who told us once again how exciting it all was. There were also six incredibles, four amazings, six wonderfuls, four fabulouses, five fantastics and – the overall winner – seven miracles.

Gabby to a mother who'd been told that if she didn't have intrauterine surgery her twins would die: 'How did you make the decision?' Gabby to another mother, earlier seen crying because the obstetrician had told her that her baby was likely to die: 'Was there a time when you thought Daniel was not going to make it?' As Daniel turned out to be still on a ventilator, and had developed pneumonia, this was not just a stupid but also an insensitive and unethical question.

Logan's other co-presenter was Andrew Castle. If I'd been the anaesthetist in theatre I would have stuffed his microphone down his throat. He was nauseating. Among his pearls of wisdom while commentating on the elective section was: 'What strikes everybody, I think, who is not used to this procedure is the sheer lack of drama and utter professionalism of everybody in here.' What was he expecting? Hysteria and incompetence?

In between the exciting visits to birthing rooms in which little was happening and to Andrew in the operating theatre, we saw photos of recently born babies sent in by viewers. Channel 5 and Gabby Logan seemed not to realize that every baby is indeed a miracle to its parents, but logically babies are no more miraculous than anything else in biology, and other people's babies are, to most other people, not remotely interesting. A chance to inform – about pain relief, about the relative merits of natural and caesarean births, and many other things – was thrown away in favour of sensationalism, which turned out not to be sensational at all. **BJHM**

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