Atypical interstitial granuloma annulare: a possible uncommon systemic manifestation of atrial myxomas

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Abstract

A 73-year-old man presented with lower back pain and bilateral palmar lesions. Urine culture grew *Streptococcus agalactiae* and C-reactive protein was raised. He was treated with antibiotics for urinary tract infection with suspected pyelonephritis. An echocardiogram ordered to look for infective endocarditis was suggestive of a left atrial myxoma. He subsequently developed an acute right common iliac artery embolus, which resolved with conservative management. He underwent a successful open-heart left atrial mass and appendage resection, with histopathology confirming atrial myxoma. Skin biopsies of the lesions ten weeks post onset showed granuloma annulare of the palms.

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Introduction

Cardiac myxomas pose significant diagnostic challenge due to a variety of non-specific symptoms and signs, hence they are often missed with potential lethal complications. We report a case of a 73-year-old man presenting with palmar lesions and incidentally diagnosed left atrial myxoma who was found to have atypical palmar interstitial granuloma annulare on histopathology from skin biopsies. This case is the first reported case of granuloma annulare in a patient diagnosed with left atrial myxoma.

Case presentation

A 73-year-old man presented with two days' history of lower back pain, which was preceded by bilateral palmar lesions (Figure 1) present for ten days. He had no shortness of breath, chest pain or constitutional symptoms. His past medical history included Gleeson 8 prostate cancer (localised) treated with transurethral resection of the prostate (TURP) and cyproterone acetate, type II diabetes mellitus (T2DM) (poorly controlled), ischaemic heart disease, congestive heart failure, chronic obstructive pulmonary disease, hypertension, atrial fibrillation, obstructive sleep apnoea and chronic left hip pain managed with opioids. His admission medications included aspirin, bisoprolol, methadone, cyproterone acetate, desvenlafaxine, digoxin, esomeprazole and metformin. There were no recent changes in medications prior to development of a rash. He reported no family history of malignancy or cardiac tumours. He is a retired truck driver, living at home with his wife.

Figure 1 Bilateral non-scaly, annular, erythematous and pruritic plaques

Examination revealed blood pressure 140/80 mmHg, heart rate 80 beats per minute and temperature 36.5°C. There were no added heart sounds or murmurs. Abdominal examination revealed bilateral flank tenderness. The palmar lesions were mildly tender to palpation (Figure 1). No Osler's nodes, Janeway lesions or splinter haemorrhages were noted.

Urine culture grew *Streptococcus agalactiae* and C-reactive protein was raised at 68. Computed tomography of chest,

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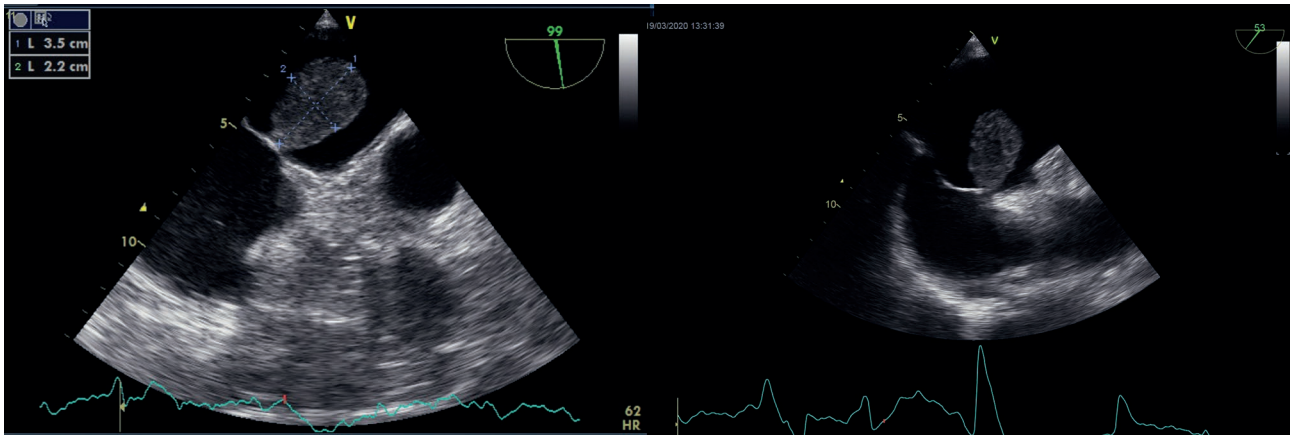
Figure 2 Transoesophageal findings of the patient showing a large, mobile left atrial mass measuring 3.5 × 2.3 cm attached with pedicle to the intra-atrial septum suggestive of left atrial myxoma



abdomen and pelvis with contrast, full blood and white cell counts, erythrocyte sedimentation rate, blood cultures, viral serology and anti-nuclear antibody were all unremarkable.

The patient was started on intravenous ceftriaxone for urinary tract infection with suspected pyelonephritis. This was later de-escalated to oral amoxicillin/clavulanic acid.

A transthoracic echocardiogram was ordered to investigate the palmar lesions for potential infective endocarditis. This unexpectedly showed a large mobile mass within the left atrium. An urgent transoesophageal echocardiogram was suggestive of left atrial myxoma (Figure 2).

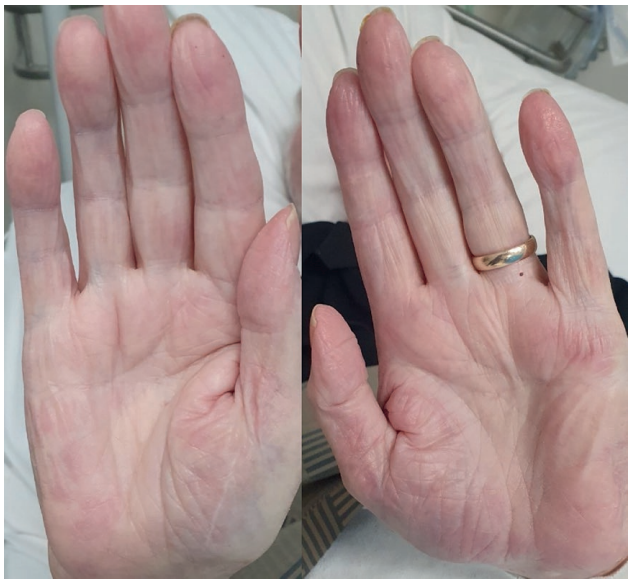
The patient subsequently developed right lower limb paraesthesia, with a computed tomography angiogram showing an acute right common iliac artery embolus. Heparin infusion was started, and he was escalated to a tertiary hospital with vascular intervention options. Fortunately, the right lower limb symptoms resolved with conservative management and he underwent a successful left atrial mass and appendage resection nine days after initial presentation, with histopathology confirming atrial myxoma.

Skin biopsies were taken from his palms seven weeks post myxoma resection when he was coincidentally admitted for acute cholecystitis, *Enterococcus faecium* bacteraemia and mild right great toe cellulitis. These conditions were managed with antibiotics and an inpatient laparoscopic cholecystectomy. The palmar lesions had regressed gradually post myxoma resection and were barely visible at seven weeks post surgery (Figure 3). Histological findings showed interstitial non-necrotising granulomatous dermatitis, suggestive of granuloma annulare (interstitial type) (Figure 4). Special stains for fungi and acid-fast bacilli were negative.

Discussion

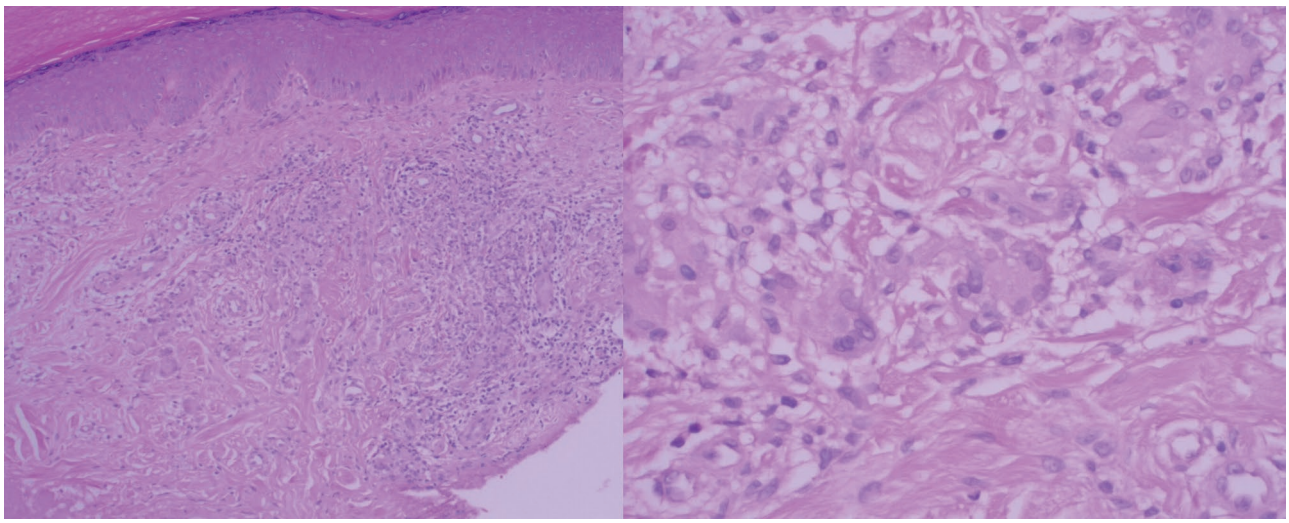
Cardiac myxoma is the commonest primary cardiac neoplasm, accounting for 30–50% of all primary heart tumours.[1] Over 70% of cardiac myxomas originate from the left atrium.[1] It is often referred to as 'the great mimicker' of other diseases as 90% of patients present with systemic symptoms including fever, weight loss, malaise and arthralgia.[2] Multiple cutaneous manifestations of cardiac myxomas have been reported, including telangiectasia, serpiginous lesions, livedo reticularis and tender papules or macules on the extremities.[2] Cardiac myxomas are also seen in Carney complex, an autosomal-dominant familial neoplastic syndrome associated with endocrinopathy and spotty skin pigmentation such as lentigines or naevi.[2]

Figure 3 Barely visible bilateral palmar lesions post myxoma resection



This is the first reported case of granuloma annulare in a patient with left atrial myxoma. Granuloma annulare is an idiopathic, chronic and non-infectious granulomatous skin disease.[3] Atypical variants include granuloma annulare with photosensitive distribution, unusually severe forms and lesions in uncommon sites such as the face, palms (as noted in this case) and ears.[3] Atypical granuloma annulare has

Figure 4 Haematoxylin and eosin stain on the skin biopsies taken from the patient revealed granulomatous inflammation involving the upper-mid dermis with multi-nucleated giant cells and scattered lymphocytes, with surrounding degenerated collagen fibres (at 100× and 200× magnification)



been linked with systemic disease such as diabetes, thyroid disease, malignancy and solid tumours including lung, breast, colon, cervical, testicular and prostate cancer.[3]

The sole previous case report documenting granuloma annulare in a prostate cancer patient had simultaneous presentations of both disease and persistent skin lesion after prostate surgery.[4] In our reported case, it is unlikely that this patient's stable prostate cancer in remission is linked to the new palmar lesions and atrial myxoma. In addition, this patient has poorly controlled diabetes, but this is a long-standing condition and the association of granuloma annulare with diabetes mellitus remains controversial, with more recent studies showing no statistical association between the two diseases.[3]

Cardiac myxomas tend to be detected late when embolic phenomena occur. It was found that up to 50% of left atrial myxomas result in systemic embolisation,[5] which can lead to morbid complications such as intracardiac obstruction, stroke, myocardial infarction, gut ischaemia and limb ischaemia.[1] At present, the challenging diagnosis of cardiac myxoma relies heavily on having a high clinical suspicion due to the lack of specific clinical symptoms and signs. Once the diagnosis of cardiac myxoma is made, treatment with surgical resection is almost curative as only 5% of sporadic cardiac myxomas recur after resection in ten years.[5]

Conclusion

This case highlights the potential diagnostic utility of cutaneous manifestations like granuloma annulare in arriving at an earlier diagnosis of atrial myxoma. Besides screening for systemic diseases, clinicians should consider investigation for cardiac myxoma with echocardiography in patients who present with atypical granuloma annulare.

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