Diogenes syndrome causing life-threatening complications of Paget's disease

Introduction

This article describes a case of an 83-year-old woman who was admitted to an old age psychiatric unit in a state of neglect and with pulmonary oedema. She had marked skeletal abnormalities and a new harsh ejection systolic murmur as a result of Paget's disease and critical aortic stenosis. This is the first case of the senile squalor syndrome associated with severe Paget's disease described in the literature. Fifty years since its first recognition, a universal service model to prevent the medical and psychiatric complications of senile squalor remains to be achieved.

Discussion

Diogenes syndrome is a condition, predominantly of the elderly, characterized by social isolation, severe self-neglect and hoarding behaviour (Cipriani et al, 2012). The latter can manifest as syllogomania, the compulsive hoarding of rubbish. Clark et al (1975) coined the term 'Diogenes syndrome' for a condition first described by Macmillan and Shaw (1966).

Although often considered a harmless variant of an eccentric isolation, this case shows how Diogenes syndrome can contribute to life-threatening medical complications. Almost half of patients with the syndrome are dead within 5 years (Hanon et al, 2004). According to Dyer et al (2007) 50% of patients have hypertension, 30% dementia, 25% diabetes, 20% arthritis, 18% cerebrovascular disease and 10% ischaemic heart disease but as far as the authors are aware Paget's disease complicating Diogenes syndrome has not been described.

This patient's impairments in mental state and eccentricity had driven a reluctance to engage with health services. She had multiple nutritional deficiencies and untreated Paget's disease. Paget's disease can cause an increase in cardiac output heart failure as a result of arteriovenous malformations in the expanded bone marrow. Paget's disease can cause high output heart failure, but this patient presented with critical aortic stenosis, perhaps caused by mechanical stress as well as active cellular and inflammatory processes (Strickberger et al, 1987).

A variety of psychiatric presentations may underlie the senile squalor syndrome. These include dementia, alcohol abuse, obsessive compulsive disorder, and psychotic or affective disorders; but a third to half of patients have no mental illness (Cooney and Hamid, 1995; Reyes-Ortiz, 2001). Whether the syndrome represents a frontal dysexecutive syndrome, the end stage of personality disorder or a prodromal dementia has been much debated (Vostanis and Dean, 1992; Grignon et al, 1999). This patient's cardiac failure may in turn have contributed to her impaired cognitive function as suggested by the small vessel disease on magnetic resonance imaging.

Perhaps most soberingly, 50 years after Macmillan set up his landmark service, a model of combined psychiatric and medical health care remains elusive. BJHM

Dr Musa Sami *is CT3 Psychiatry,* **Dr James Tucker** *is General Practitioners Vocational Trainee,* **Dr Karin Norris** *is Associate Specialist in Old Age Psychiatry and* **Dr Giles Townsend** *is Consultant Psychiatrist in the Department of Old Age Psychiatry, West Park Hospital, Surrey and Borders NHS Partnership Foundation Trust, Epsom, Surrey KT19 8PB, and* **Dr Olusola Odemuyiwa** *is Consultant Cardiologist in the Department of Cardiology, Epsom General Hospital, Epsom, Surrey*

Correspondence to: Dr M Sami (musasami@doctors.org.uk)

Case Report

An 83-year-old woman with a benign history of Paget's disease was noticed to be ever more reclusive after a fall. Her reputation as something of a recluse and eccentric had been shaped by her isolated upbringing in Nazi-occupied Guernsey, but until her husband died 40 years ago she appeared to have led a colourful existence as a regular patron of grand concerts.

Over the year before her admission to hospital she had grown withdrawn and allowed no one into her flat. Concerned neighbours left food on her doorstep. In the end social services and the mental health team carried out an assessment through her letterbox and detained her under section 2 of the UK Mental Health Act 1983.

On admission she was a small, hunched, frail woman with severe scoliosis, kyphosis and bowing of her legs. She weighed 46 kg and appeared malnourished. Her fingernails were grossly overgrown and her clothes, which were noted to be 'like cardboard', had to be 'prised off' her.

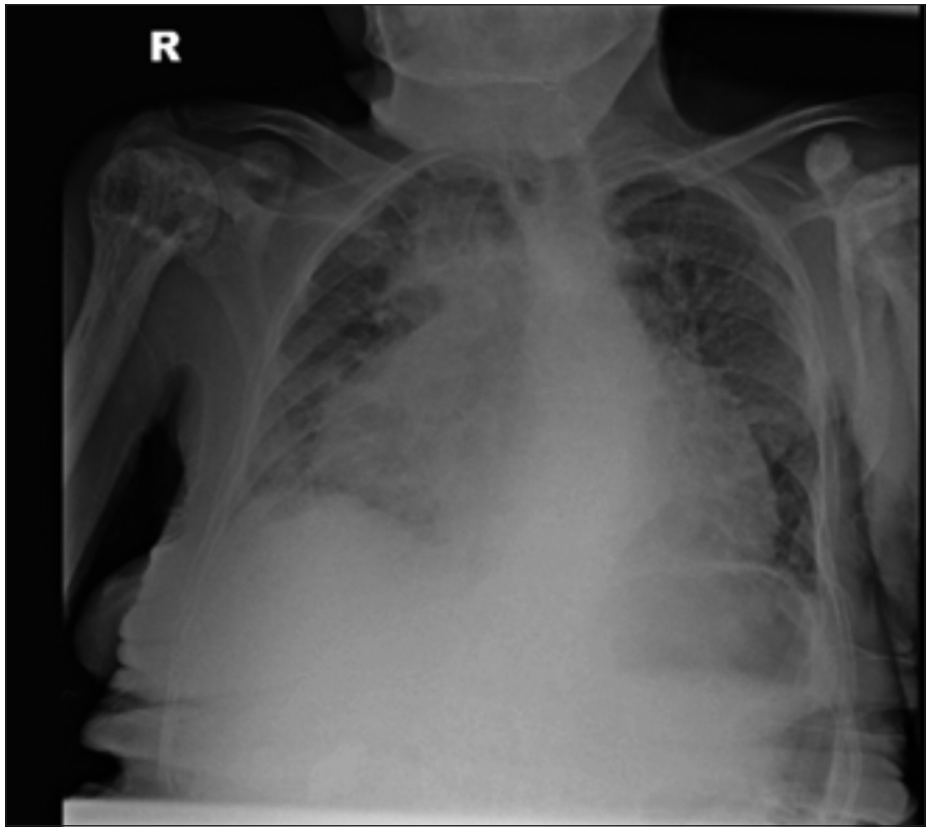
The patient was breathless on minimal exertion, with a loud ejection systolic murmur and widespread crackles in her chest. She had a right third cranial nerve palsy. Laboratory results showed a microcytic anaemia with a haemoglobin of 8.7 g/dl, alkaline phosphatase markedly raised at 1417 IU/litre, and low vitamin D levels. Her thyroid-stimulating hormone level was 37 mU/litre (normal <5 mU/litre) but thyroxine levels were normal. Electrocardiogram showed right bundle–branch block but no acute changes. A chest X-ray showed marked cardiomegaly and pulmonary oedema (*Figure 1*) and the echocardiogram confirmed critical calcified aortic stenosis with a valve area of 0.4 cm^2 (normal valve area >2 cm^2). The left ventricular ejection fraction was also depressed at 40%. Initial treatment consisted of nutritional supplements, thiamine, salbutamol and frusemide. She also had levothyroxine and vitamin D but a trial of bisphosphonate therapy caused dyspepsia.

Cognitive testing revealed a mini mental state examination (MMSE) score of 24/30 and Addenbrooke's Cognitive Examination-Revised (ACE-R) score of 78/100, losing marks on the orientation and fluency items. Immediate recall and retention was preserved. Magnetic resonance imaging revealed evidence of small vessel disease and bilateral hippocampal atrophy. As a result she was diagnosed as having atypical early mixed dementia contributed to by multiple medical comorbidities. Cardiac surgery was not deemed appropriate.

The patient was grateful for the care but always wished to return home. After multidisciplinary assessment identified her lack of capacity she was discharged to a nursing home from where she attends follow-up for her medical and psychiatric conditions.

Cipriani G, Lucetti C, Vedovello M, Nuti A (2012) Diogenes syndrome in patients suffering from dementia. *Dialogues Clin Neurosci* **14**(4): 455–60
Clark AN, Mankikar GO, Gray I (1975) Diogenes syndrome: a clinical study of gross neglect in old age. *Lancet* **i**: 366–8
Cooney C, Hamid W (1995) Review: Diogenes syndrome. *Age Ageing* **24**: 451–3

Figure 1. Chest X-ray on admission showing widespread cardiac failure and cardiomegaly. A suspicious right apical mass was shown to be venous congestion. Bony features of Paget's disease are also present.



Dyer CB, Goodwin JS, Pickens-Pace S, Burnett J, Kelly PA (2007) Self-neglect among the elderly: a model based on more than 500 patients seen by a geriatric medicine team. *Am J Public Health* **97**(9): 1671–6
Grignon S, Bassiri D, Bartoli JL, Calvet P (1999) Association of Diogenes syndrome with a compulsive disorder. *Can J Psychiatry* **44**: 91–2
Hanon C, Pinquier C, Gaddour N, Saïd S, Mathis D, Pellerin J (2004) Diogenes syndrome: a transnosographic approach. *Encéphale* **30**: 315–22
Macmillan D, Shaw P (1966) Senile breakdown in standards of personal and environmental cleanliness. *BMJ* **ii**: 1032–7
Reyes-Ortiz CA (2001) Diogenes syndrome: the self-neglect elderly. *Compr Ther* **27**: 117–21
Strickberger SA, Schulman SP, Grover MH (1987) Association of Paget's disease of bone with calcific aortic valve disease. *Am J Med* **82**: 953–6
Vostanis P, Dean C (1992) Self-neglect in adult life. *Br J Psychiatry* **161**: 265–7

LEARNING POINTS

- A life-threatening complication of severe Paget's disease was only successfully managed after a patient with features of Diogenes syndrome was detained under the Mental Health Act.
- Detailed physical work-up and screening for an underlying dementia or medical comorbidity and joint medical–psychiatric liaison is vital for optimal care of patients with Diogenes syndrome.
- Careful capacity assessment and re-housing may be required.