

# Pleural-based giant solitary fibrous tumour with associated hypoglycaemia: unusual presentation with pulmonary hypertension in a patient with Doege–Potter syndrome

**Authors:** Qasim Gohir,<sup>A</sup> Shilajit Ghosh,<sup>B</sup> Olivia Boshier,<sup>C</sup> Emma Crawford,<sup>D</sup> Koottalai Srinivasan<sup>E</sup> and Harmesh Moudgil<sup>F</sup>

## ABSTRACT

Refractory hypoglycaemia in a patient with a solitary fibrous tumour (SFT) is very rare and was first reported in 1930 independently by Doege and Potter, leading to it being named ‘Doege–Potter syndrome’. Here, we report the unusual case of a 77-year-old woman with a giant solitary fibrous pleural tumour who presented with complicating pulmonary hypertension and associated heart failure with hypoglycaemia, and subsequently underwent curative resection of the pleural mass with clinical improvement.

**KEYWORDS:** Doege–Potter syndrome, solitary fibrous tumour, hypoglycaemia

**DOI:** 10.7861/clinmed.2023-0274

## Case presentation

A 77-year-old woman presented acutely to secondary care with a 5-week history of worsening breathlessness, especially lying

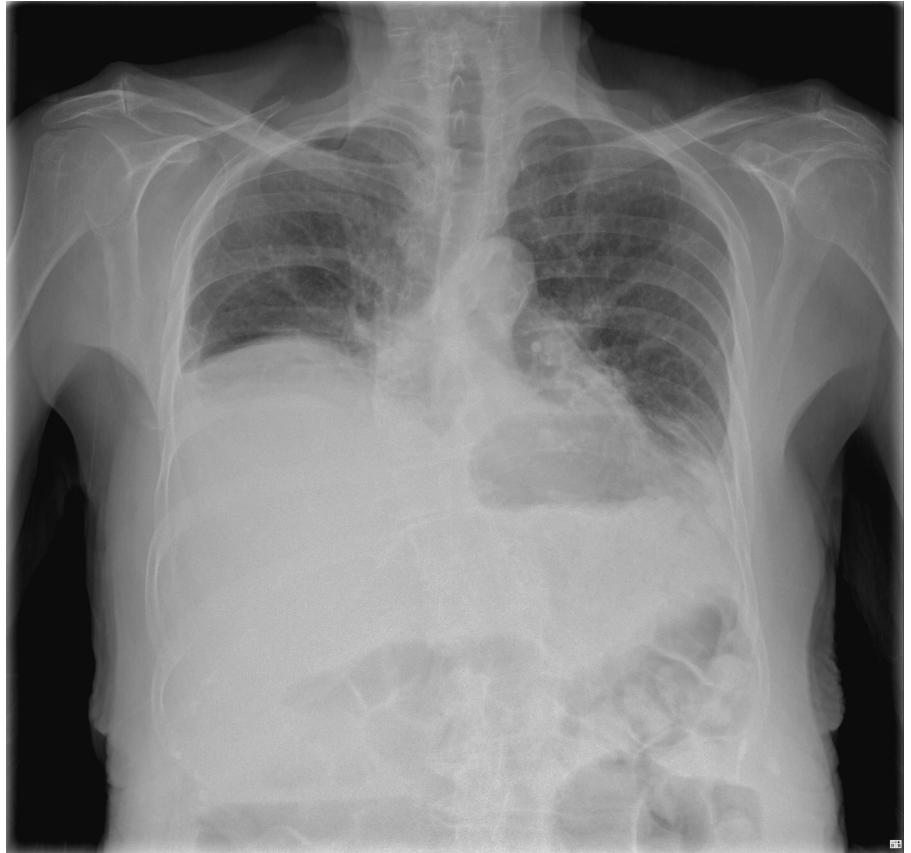
down, and bilateral lower limb swelling. She denied chest pain, cough, haemoptysis, palpitations, dizziness, fever, weight loss, and urinary or bowel symptoms. Her background history included hypertension, a large hiatus hernia and a whiplash injury 10 years previously (no comparative radiology); regular medications included bendroflumethiazide, atorvastatin, ramipril and omeprazole. She had never smoked and was usually independent with a performance status of zero.

On examination, she was alert and orientated but dyspnoeic at rest. She was afebrile. Her blood pressure was 140/76 mmHg, her pulse was 96 beats per minute with atrial fibrillation, her respiratory rate was 17 breaths per minute and her oxygen saturation was 96% breathing air. There was no cervical or axillary lymphadenopathy. Breath sounds were reduced with dullness to percussion in the right hemithorax and fine bi-basal crackles. Her abdomen was soft and non-tender and there was bilateral lower limb oedema. Routine blood tests were within normality with the exception to mild lymphopenia at  $0.8 \times 10^9/L$  ( $1.5\text{--}4.5 \times 10^9/L$ ) and bilirubin at the upper level of normal at  $22 \mu\text{mol/L}$  ( $0\text{--}22 \mu\text{mol/L}$ ) with normal gamma glutamyl transferase at 24 U/L ( $0\text{--}45 \text{U/L}$ ). A chest radiograph showed a raised right hemidiaphragm with possible underlying mass (Fig 1).

Initial treatment was with a beta-blocker and intravenous diuretic for congestive heart failure with new-onset atrial fibrillation. An echocardiogram showed moderate tricuspid regurgitation and raised pulmonary/right ventricular pressure/strain with an intermediate probability of pulmonary hypertension. A bedside ultrasound scan of chest confirmed a raised right hemidiaphragm with passive right lung collapse and pleural effusion too small to aspirate.

A contrast CT scan of thorax (Fig 2) confirmed a large 15-cm right-sided pleural-based mass. A subsequent lung cancer multidisciplinary team (MDT) meeting commented on the mass having calcification and abnormal feeding vessels arising from the coeliac axis, with concerns over shunting and neovascularisation. The MDT suggested a likely benign mass but recommended further imaging and a guided biopsy. A positron emission tomography (PET/computed tomography (CT) scan of chest confirmed a mild FDG uptake with SUV 2.8 to the large right hemithorax pleural heterogeneous mass and with an uptake of SUV 3.8 to the peripheral right pleura at the site of a small

**Authors:** <sup>A</sup>respiratory specialist trainee, Princess Royal Hospital, Shrewsbury and Telford Hospitals NHS Trust, Telford, UK; <sup>B</sup>consultant in cardiothoracic surgery, Royal Stoke University Hospital, University Hospitals North Midlands, Stoke-on-Trent, UK; <sup>C</sup>consultant physician, Princess Royal Hospital, Shrewsbury and Telford Hospitals NHS Trust, Telford, UK; <sup>D</sup>consultant physician, Princess Royal Hospital, Shrewsbury and Telford Hospitals NHS Trust, Telford, UK, and honorary lecturer, University of Keele Medical School, Stoke-on-Trent, UK; <sup>E</sup>consultant physician and hospital dean, Princess Royal Hospital, Shrewsbury and Telford Hospitals NHS Trust, Telford, UK, and honorary senior lecturer, University of Keele Medical School, Stoke-on-Trent, UK; <sup>F</sup>consultant physician, Princess Royal Hospital, Shrewsbury and Telford Hospitals NHS Trust, Telford, UK and University of Keele Medical School, Stoke-on-Trent, UK, and honorary senior lecturer, University of Keele Medical School, Stoke-on-Trent, UK



**Fig 1.** Chest radiograph showing a raised right hemidiaphragm with possible underlying mass.

effusion. Subsequent ultrasound-guided biopsy confirmed a spindle cell, likely fibrous, tumour.

Although initially managed as an outpatient pending tests/discussions, the patient was admitted twice with worsening fluid overload, during which she was noted to have multiple hypoglycaemic episodes with blood glucose often  $<2.0$  mmol/L. There was no previous history of diabetes or prescription of oral hypoglycaemic drugs or insulin.

Tests investigating the hypoglycaemic episodes, including serum C-peptide, insulin and insulin-like growth factor 2 (IGF2) levels were requested but declined because they were not available. The working diagnosis was a giant solitary fibrous tumour (SFT) with hypoglycaemia diagnosing Doege–Potter syndrome (DPS) in addition to cardiac decompensation resulting from atrial fibrillation and compression of the inferior vena cava by the large fibrous tumour.

The cardiothoracic surgeons completely resected the tumour and the patient made a good recovery with no further hypoglycaemic events. The patient's case was discussed at the regional sarcoma MDT; they advised a high chance of future malignancy and advised local surgical and CT surveillance postoperatively as protocol over the following 5 years.

## Discussion

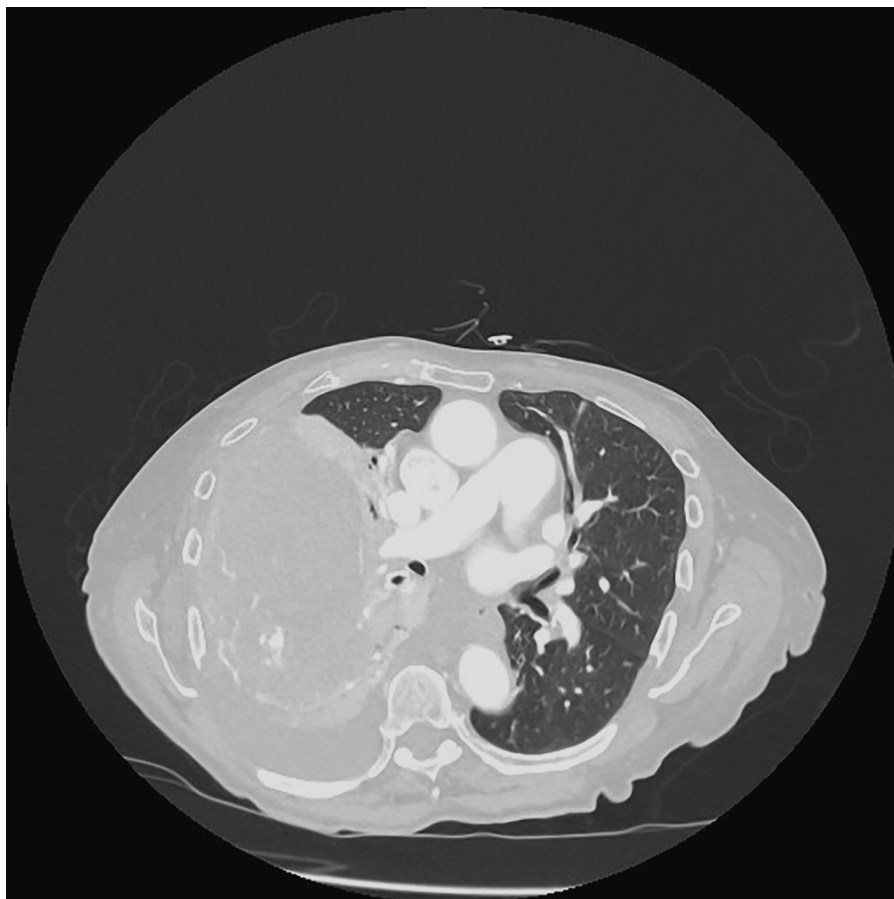
SFTs usually present during the sixth decade, with 78–88% being benign,<sup>1,2</sup> although malignant transformation is not uncommon. Prognosis is excellent with 8% recurrence after first resection and

cure after additional surgery.<sup>2</sup> Although they can arise from any soft tissue, most arise from mesothelial cells and rarely from the pleural fibroblast,<sup>3</sup> with 80% of pleural fibromas originating in the visceral pleura.<sup>4</sup> Recurrence is higher in patients with malignant SFT, particularly when associated with a pleural effusion, with most patients dying within 2 years.<sup>2</sup> It is not common for SFTs to produce hormones, with tumours  $<10$  cm often asymptomatic.

Pleural SFTs represent  $<5\%$  of all pleural tumours and their evolution is unpredictable. Other sites for SFT include the liver, pelvis, peritoneum, meningeal, adrenal, intrapulmonary, urinary bladder and pericardium.<sup>5</sup> Hypoglycaemia associated with pleural SFT is considered a non-islet cell tumour hypoglycaemia (NICTH) and is also found with haemangiopericytomas and myxofibrosarcomas.<sup>5</sup>

DPS is a rare paraneoplastic syndrome caused by pleural SFT, with an estimated incidence of 0.2–0.5 per million per year. It presents as a hypoinsulinaemic hypoglycaemia from the ectopic secretion of a prohormone of IGF2 and is characterised by constant hypoglycaemia, suppressed serum insulin, serum C peptide and growth hormone, and low serum IGF1 against levels of IGF2 that are either normal or elevated.<sup>6</sup> The hypoglycaemia can also result from high glucose usage and insulin receptor proliferation. The most important aspect is the significant change in the IGF2/IGF1 ratio ( $>10$ ) confirming NICTH diagnosis.<sup>5</sup> Other potential causes of hypoglycaemia in patients without diabetes need exclusion, including critical illness, alcohol, cortisol deficiency or malnourishment.

Patients with DPS can be temporarily treated for symptoms with continuous glucose 10% infusion to restore normal glycaemic



**Fig 2.** Computed tomography scan of thorax with contrast showing a large 15-cm right-sided pleural-based mass.

levels.<sup>7,8</sup> In addition, the use of glucocorticoids is effective in glucose-refractory cases because they stimulate gluconeogenesis and suppress the production of IGF2. However, surgical resection is curative and, thus, the gold standard treatment. Alternative approaches to other target areas include radioembolisation with labelled yttrium 90 (Y90) or oncology with temozolomide/bevacizumab.<sup>5</sup>

The improvement from hypoglycaemia from debulking/curative surgery provides supportive evidence for DPS in this case. It is unique not only because of refractory hypoglycaemia with the giant size and site of the pleural tumour causing pressure on mediastinal structures, including the inferior vena cava, but also with the development of pulmonary hypertension with shunting and development of collateral feeding vessels through the large mass. Whether the whiplash injury several years earlier contributed to the raised right hemidiaphragm would only be speculative (no radiology). ■

## References

- 1 de Perrot M, Fischer S, Bründler MA, Sekine Y, Keshavjee S. Solitary fibrous tumors of the pleura. *Ann Thorac Surg* 2002;74:285–93.
- 2 Robinson LA. Solitary fibrous tumor of the pleura. *Cancer Control* 2006;13:264–9.
- 3 Bunton RW, Borrie J. Pleural fibromas: a clinical review and report of six patients. *Ann Thorac Surg* 1982;33:609–13.
- 4 Briselli M, Mark EJ, Dickersin GR. Solitary fibrous tumors of the pleura: eight new cases and review of 360 cases in the literature. *Cancer* 1981;47:2678–89.
- 5 Savu C, Melinte A, Posea R *et al*. Pleural solitary fibrous tumors—a retrospective study on 45 patients. *Medicina* 2020;56:185.
- 6 Han G, Zhang Z, Shen X *et al*. Doege-Potter syndrome: a review of the literature including a new case report. *Medicine* 2017;96:e7417.
- 7 Bodnar TW, Acevedo MJ, Pietropaolo M. Management of Non-islet-cell tumor hypoglycemia: a clinical review. *J Clin Endocrinol Metab* 2014;99:713–22.
- 8 Mohammadi K, Abi Khalil C, Olivier S *et al*. Paraneoplastic hypoglycemia in a patient with a malignant solitary fibrous tumor. *Endocrinol Diabetes Metab Case Rep* 2014;2014:140026.

**Address for correspondence: Dr Qasim Gohir, The Princess Royal Hospital, Apley Castle, Apley, Telford, TF1 6TF, UK.  
Email: qasim.gohir@nhs.net  
Twitter: @QasimGohir**