CASE REPORT

Therapeutic pulmonary artery stenting for metastatic bronchial carcinoid

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danielvawdrey@doctors.org.uk**SUMMARY**

We present a case of a middle-aged man with a 3-month history of progressive shortness of breath and peripheral oedema. Ten years prior to this, he had undergone a left pneumonectomy for metastatic bronchial carcinoid. Clinical examination revealed significant right heart failure, supported by transthoracic echocardiography. CT pulmonary angiogram revealed the cause to be marked progression of the bronchial carcinoid causing severe external compression of right pulmonary artery (RPA). In view of the distressing symptoms, a palliative endovascular intervention to the RPA was attempted to relieve obstruction, improve blood flow through the right lung and offload the right ventricle. This was performed under general anaesthesia involving interventional cardiology and radiology specialists together with a specialist anaesthetic team with extensive experience of managing carcinoid patients. The result was a marked improvement in symptoms and right heart function and the patient was discharged 2 days later.

BACKGROUND

Carcinoid tumours are relatively uncommon with an annual incidence of 2/100 000 population.¹ Prognosis is directly related to tumour spread with an approximate 5-year survival rate of 80% for localised disease and 30% for distant metastatic disease.² There are numerous complications that may result from carcinoid metastases that include thrombosis due to platelet activation, carcinoid crisis due to vasoactive peptide release and subsequent profound hypotension, hepatic failure and cardiac involvement.^{3 4}

This case details the management of another life-threatening complication arising from metastatic carcinoid. More specifically, bronchial carcinoid compression of pulmonary arterial outflow and subsequent right heart failure. The case highlights the benefits of information gained from non-invasive imaging, the multidisciplinary team approach to treatment and the importance of expert anaesthetic management in order to avoid carcinoid crisis when physical intervention is required.

CASE PRESENTATION

A middle-aged man presented to the local hospital with a 3-month history of progressive shortness of breath and peripheral oedema. Ten years prior to this, he had undergone a left intrapericardial pneumonectomy for metastatic bronchial carcinoid. During

surgery, the carcinoid tumour was found to be encasing the left pulmonary artery (LPA) and as much as possible of the tumour was removed with the LPA.

On admission, he was markedly breathless at rest and examination revealed marked right heart failure with jugular venous pressure elevated to the level of his earlobes and bilateral pitting oedema to mid-thighs. Auscultation revealed a loud, high-pitched systolic murmur at the left upper sternal edge. There was dullness and decreased air entry on the left side, consistent with the previous pneumonectomy (figure 1), but normal breath sounds throughout the right side.

INVESTIGATIONS

CT pulmonary angiogram revealed no evidence of pulmonary thromboembolic disease however, there was marked progression of the bronchial carcinoid within the mediastinum causing severe external compression of right pulmonary artery (RPA; figure 2). Transthoracic echocardiography showed right heart dilation with systolic septal flattening, severe right ventricular (RV) systolic impairment and severe tricuspid regurgitation (figure 3A,B). The estimated RV systolic pressure (using peak tricuspid regurgitant jet velocity) was 65–70 mm Hg. The morphology of all of the valves was normal with no evidence for cardiac carcinoid involvement.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses for right heart failure in cases of bronchial carcinoid include the following

Figure 1 A plain chest radiograph showing mediastinal shift towards the left side secondary to the previous left pneumonectomy.

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Figure 2 (A) CT pulmonary angiogram (coronal reconstruction) and (B) an axial image showing external compression of the right pulmonary artery (arrows). (C) Three-dimensional surface-rendered CT reconstruction showing the slit-like orifice of the right pulmonary artery due to the external compression (arrow). (D) Transthoracic echo (parasternal short-axis view) showing the pulmonary valve (asterisk), bronchial carcinoid mass (arrowhead) and the main pulmonary artery (arrow).

Figure 3 Transthoracic echo images at end-diastole and peak systole. (A) and (B; pre): Prior to RPA stent showing severe right heart dilation with systolic septal flattening. (C) and (D; post): Postprocedure images showing resolution of systolic septal flattening.

1. Cardiac carcinoid involvement with valvular compromise;
2. Pulmonary embolism;
3. External compression of pulmonary artery.

In this case, non-invasive imaging made it possible to diagnose right heart failure secondary to severe external compression of the RPA by bronchial carcinoid.

TREATMENT

In view of the distressing symptoms, it was decided to attempt a palliative procedure with endovascular intervention to the RPA to relieve obstruction, improve the blood flow to the right lung and offload the RV. This was performed under general anaesthesia involving interventional cardiology and radiology specialists together with a specialist anaesthetic team with extensive experience of managing carcinoid patients. Continuous intravenous Octreotide was used with bolus doses required to treat hypotension immediately following induction and in doing so, avoid carcinoid crisis. Prior to intervention, pressures were as follows: peak RV systolic pressure 73 mm Hg (end-diastolic 11 mm Hg), proximal pulmonary artery 73/6 mm Hg, distal RPA 5/2 mm Hg (mean 4 mm Hg). A stiff guidewire was passed across the narrowed proximal portion of the RPA and a self-expanding 18×60 mm Sinus XL stent was deployed with successful re-expansion of the RPA (figure 4). There was immediate relief of obstruction and peak RV systolic pressure fell to 35 mm Hg.

OUTCOME AND FOLLOW-UP

Immediately following extubation, the patient noted a dramatic improvement in symptoms. This correlated well with repeat transthoracic echocardiography which revealed a significant improvement in right-heart dilation and function (figure 3C,D). The patient was discharged from hospital 2 days later and lived for a further 7 months postprocedure.

DISCUSSION

Carcinoid tumours arise from neuroendocrine cells and are most commonly found in the distal ileum, elsewhere in the gastrointestinal tract and the bronchi. They release multiple vasoactive peptides, including serotonin, which is inactivated by the liver and lungs. Carcinoid syndrome results from systemic circulatory distribution of these vasoactive peptides (from hepatic metastases or primary bronchial carcinoid).⁵ Symptoms include flushing, palpitations, diarrhoea and wheeze. Annual incidence of carcinoid tumours is 2/100 000 population of whom, approximately 10% develop carcinoid syndrome.¹ Prognosis is directly related to tumour spread with an approximate 5-year survival rate of 80% for localised disease and 30% for distant metastatic disease.²

Biological, radiological and histological investigations may be required to help confirm a diagnosis. This includes 24 h serotonin metabolite 5-hydroxyindoleacetic acid urine collection, CT, radiolabelled octreotide scintigraphy and biopsy, for example, through an endoscopy or a bronchoscopy.⁶

Medical management of carcinoid is directed towards relief of symptoms and possible inhibition of tumour growth, using somatostatin analogues (such as octreotide) which reduce serotonin release.⁷ Surgery however may be curative and is therefore the treatment of choice. Debulking of metastatic disease also confers significant symptomatic benefit.⁶ In cases whereby surgical resection of hepatic carcinoid is not possible, hepatic artery embolisation can improve symptoms and slow tumour progression.⁸ Chemotherapy strategies for palliation of metastatic carcinoid are currently being evaluated in clinical trials.

Figure 4 Stent deployment within right pulmonary artery (RPA). (A) Angiography of the RPA prior to stenting. (B) Self-expanding stent deployed within the RPA. (C) RPA angiogram after stenting.

Complications resulting from metastatic carcinoid include thrombosis due to platelet activation, hepatic failure, cardiac involvement and carcinoid crisis.^{3 4} Complication involving external compression of pulmonary artery and subsequent right heart failure is less well documented. Cardiac involvement due to a serotonin-mediated fibrous plaque deposition over the endocardial surface develops in approximately 50% of patients with carcinoid syndrome. It is far more common for the right heart to be affected (with tricuspid valve dysfunction and severe tricuspid regurgitation) as serotonin is metabolised in the lungs before reaching the left atrium. Improvement in survival rates has been noted in recent years which may be due to valve surgery for cardiac carcinoid.⁹ Non-invasive imaging made it possible to determine that external compression of RPA was the cause of the right heart failure in this case rather than cardiac carcinoid involvement.

Carcinoid crisis can occur due to the release of high levels of vasoactive peptide (including serotonin) into the systemic circulation causing bronchospasm, tachycardia and vasodilation with subsequent profound hypotension. This may be triggered by anaesthetic induction or physical disruption of carcinoid tumour, for example, by invasive therapeutic procedures.⁶ The procedure described in this case was supported by a specialist anaesthetic team with extensive experience of managing carcinoid patients. Continuous intravenous octreotide was administered in conjunction

with bolus doses in order to antagonise effects of serotonin and therefore prevent carcinoid crisis. The importance of a multidisciplinary team approach and expert anaesthetic management is highlighted by the favourable outcome in this case.

Learning points

- ▶ Metastatic bronchial carcinoid is associated with numerous complications, including external compression of pulmonary artery and resultant right heart failure.
- ▶ Palliative endovascular intervention may be considered in such cases where relief of obstruction to vital blood flow is required.
- ▶ When such intervention is deemed appropriate a multidisciplinary team approach should be followed, including specialist anaesthetic input with experience in managing carcinoid patients in order to avoid carcinoid crisis.

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REFERENCES

- 1 Quaadvlieg PF, Visser O, Lamers CB, *et al.* Epidemiology and survival in patients with carcinoid disease in the Netherlands. An epidemiological study with 2391 patients. *Ann Oncol* 2001;12:1295–300.
- 2 Modlin IM, Lye KD, Kidd M. A 5-decade analysis of 13,715 carcinoid tumors. *Cancer* 2003;97:934–59.
- 3 Gustafsson BI, Kidd M, Chan A, *et al.* Bronchopulmonary neuroendocrine tumors. *Cancer* 2008;113:5–21.
- 4 Poynter LR, Tewari N, Khawaja HT. Metastatic carcinoid tumour mimicking cholecystitis, and a rare case of intussusception. *BMJ Case Rep.* Published online 8 March 2011.
- 5 Bendelow J, Apps E, Jones LE, *et al.* Carcinoid syndrome. *Eur J Surg Oncol* 2008;34:289–96.
- 6 Ramage JK, Ahmed A, Ardill J, *et al.* Guidelines for the management of gastroenteropancreatic neuroendocrine (including carcinoid) tumours (NETs). *Gut* 2012;61:6–32.
- 7 Rinke A, Muller H, Schade-Brittinger C, *et al.* Placebo-controlled, double blind, prospective, randomized study on the effect of octreotide LAR in the control of tumor growth in patients with metastatic neuroendocrine midgut tumors: a report from the PROMID Study Group. *J Clin Oncol* 2009;27:4656–63.
- 8 Bloomston M, Al-Saif O, Klemanski D, *et al.* Hepatic artery chemoembolization in 122 patients with metastatic carcinoid tumor: lessons learned. *J Gastrointest Surg* 2007;11:264–71.
- 9 Møller JE, Pellikka PA, Bernheim AM, *et al.* Prognosis of carcinoid heart disease: analysis of 200 cases over two decades. *Circulation* 2005;112:3320–7.

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