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Case report

Surgical treatment of carcinoid syndrome metastatic disease: Partial hepatectomy with veno-venous bypass and subsequent tricuspid valve replacement. Case report☆

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ABSTRACT

Introduction: Carcinoid syndrome is a relatively rare condition that may affect the liver and the right heart. Some of these cases may require surgical treatment with a multidisciplinary approach.
Case report: We report the case of a patient with progressive dyspnoea, arterial hypotension and facial flushing, diagnosed with liver and cardiac involvement from carcinoid syndrome. The patient was taken to surgery in two different occasions, first for metastatic liver resection, and then to subsequent cardiac surgery for tricuspid valve replacement.
Conclusions: We report here a case review, including the clinical course, the intra-operative management, and the information available in the literature regarding which procedure to perform initially in these types of cases which are not very common in clinical practice.

Tratamiento quirúrgico de la enfermedad metastásica del síndrome carcinoide: hepatectomía parcial usando un bypass veno-venoso y posterior cambio valvular tricúspide. Reporte de caso

Palabras clave:
Hepatectomía
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RESUMEN

Introducción: El síndrome carcinoide es una condición relativamente rara que puede afectar el hígado y el corazón derecho. En algunos pacientes se requiere tratamiento quirúrgico y un manejo multidisciplinario.

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Informes de caso
Síndrome carcinoide maligno

Presentación del caso: Presentamos una paciente con diagnóstico de síndrome carcinoide con compromiso hepático y cardíaco quien inició con disnea progresiva hasta el reposo, hipotensión arterial y rubor facial. La paciente fue llevada a cirugía en dos tiempos: la resección de las metástasis hepáticas inicialmente y, en un segundo tiempo, cirugía cardíaca para el cambio de la válvula tricúspide. Ambas intervenciones se realizaron con éxito y sin complicaciones.

Conclusión: Presentamos aquí una revisión del caso, la evolución y el manejo intraoperatorio y la información disponible en la literatura para decidir cual procedimiento se debe realizar en primera instancia, ya que estos casos no son frecuentes en la práctica clínica.

Introduction

A carcinoid tumour is a tumour of malignant behaviour arising from neuroendocrine system cells, with the ability to produce bioactive substances such as vasoactive peptides and amines. The incidence of carcinoid tumours is approximately 1 case for every 100,000 people, and close to 50% go on to develop carcinoid syndrome.[1,2] However, a more recent report found a much higher incidence of 3.65 cases for every 100,000.[3]

The most common sites for carcinoid tumours are the gastrointestinal tract (67.5%) and the lungs (25.3%). In the gastrointestinal tract, the sites most commonly affected are the small bowel, the rectum and the stomach.[4]

The vasoactive hormones secreted by the tumour give rise to the so-called "carcinoid syndrome", characterised by facial flushing, arterial hypotension, diarrhoea, pruritus and bronchospasm. These vasoactive substances include serotonin, 5-hydroxytryptamine, 5-hydroxytryptophan, kallicrein, hystamine, prostaglandins, adrenocorticotopic hormone, insulin, catecholamines, parathormone, gonadotropins and substance P.[5]

Once the intestinal wall is invaded, carcinoid tumour cells may extend beyond it and spread through the lymphatic and vascular systems. Tumour cells reach the liver through the portal circulation, reducing the liver's ability to metabolise the products derived from these tumours. When tumour cells are implanted in the liver, their humoral products may reach the right heart through the inferior vena cava (IVC).[5] Pulmonary stenosis and tricuspid regurgitation are usually the predominant cardiac manifestations of this syndrome.

The current pharmacological management of carcinoid syndrome includes somatostatin analogues such as octreotide and lanreotide and, on occasions, high doses of octreotide are required for adequate symptom control.[6]

Cardiac surgery is indicated when the patient develops heart failure, specifically right ventricular failure and/or when there is severe valve dysfunction.[7] Cardiac surgery offers definitive treatment for these patients and, over the past two decades, it has resulted in a significant reduction in mortality associated with this disease.[8,9]

Few cases are described in the medical literature regarding the surgical treatment of liver metastases and heart disease in carcinoid syndrome.[10,11] This case report describes a two-stage surgical treatment of carcinoid tumour metastatic disease: liver resection in the first stage, and tricuspid valve replacement in the second stage.

Clinical case

Informed consent was obtained from the patient for the discussion and publication of this case.

A 45-year-old female patient presented with progressive dyspnoea even at rest, associated with sudden episodes of hot flashes, diarrhoea, cough and facial flushing that started in the previous five months. There were no significant findings in her clinical history, and her surgical history included appendectomy and cholecystectomy performed more than ten years before, with no anaesthetic or surgical complications.

The patient was admitted to the hospital for work-up considering that the initial physical examination revealed the presence of dyspnoea and, on auscultation, a grade 4/4 holosystolic murmur.

In the initial trans-thoracic echocardiogram (TTE), left ventricular ejection fraction (LVEF) was 60%, right ventricular function was normal, but the right ventricle was significantly dilated with severe tricuspid regurgitation associated with moderate pulmonary insufficiency and mild pulmonary stenosis. Cardiac catheterisation revealed normal coronary arteries. Pulmonary artery pressures were normal and only a moderate elevation of the inferior vena cava and right atrial (25 mmHg) pressures was found, due to tricuspid regurgitation and end-diastolic right ventricular pressure increase (35/18 mmHg), probably due to pulmonic valve insufficiency.

Abdominal computed tomography imaging showed a liver mass 14 cm in diameter involving segments II and IVa, as well as other smaller liver lesions; predominant lesions were in hepatic segments I, II, VII, VIII and X. There was no description of the presence of portal hypertension and no description of IVC status.

A tumour mass was also found 1.7 cm from the ileocecal valve. It was presumably the primary tumour, which disappeared with the use of somatostatin analogues and did not require surgical excision. These findings, together with urinary levels of 5-hydroxyindoleacetic acid (a by-product of 5-hydroxytryptamine metabolism) higher than 500 ng in 24 h confirmed the diagnosis of carcinoid syndrome.

Following diagnosis, the patient initiated treatment with octreotide (Sandostatin Novartis, Basel, Switzerland) 30 mg