

# Carcinoid heart disease as the Presentation of Ovarian Neuroendocrine Tumour in the absence of Liver Metastases

Arambewela MH<sup>1</sup>, Al-Mukhtar A<sup>2</sup>, Macdonald M<sup>3</sup>, Dennis A<sup>4</sup>, O'Toole L<sup>5</sup>, Howarth G<sup>6</sup>, Munir A<sup>1</sup>

Department of Endocrinology<sup>1</sup>, Department of Hepatobiliary Surgery<sup>2</sup>, Department of Gynaecology<sup>3</sup>, Department of Anaesthesiology<sup>4</sup>, and Department of Cardiology<sup>5</sup> Department of Histopathology<sup>6</sup> Sheffield Teaching Hospitals NHS Foundation Trust.

## Abstract

Ovarian neuroendocrine tumors (NET) are rare. We present a 65 year old lady who presented with carcinoid heart disease in the absence of liver metastases who had successful clinical and biochemical resolution following surgical excision of the primary tumor. This case highlights the importance of multidisciplinary team discussion in patients with NET.

## Introduction

Ovarian carcinoid tumors are rare neoplasms accounting for only 0.3%-1.0% of all NETS and <0.1% of all ovarian tumors (1). Occurrence of carcinoid heart disease in ovarian NET is exceedingly rare with only few reported in literature (2).

## Case details

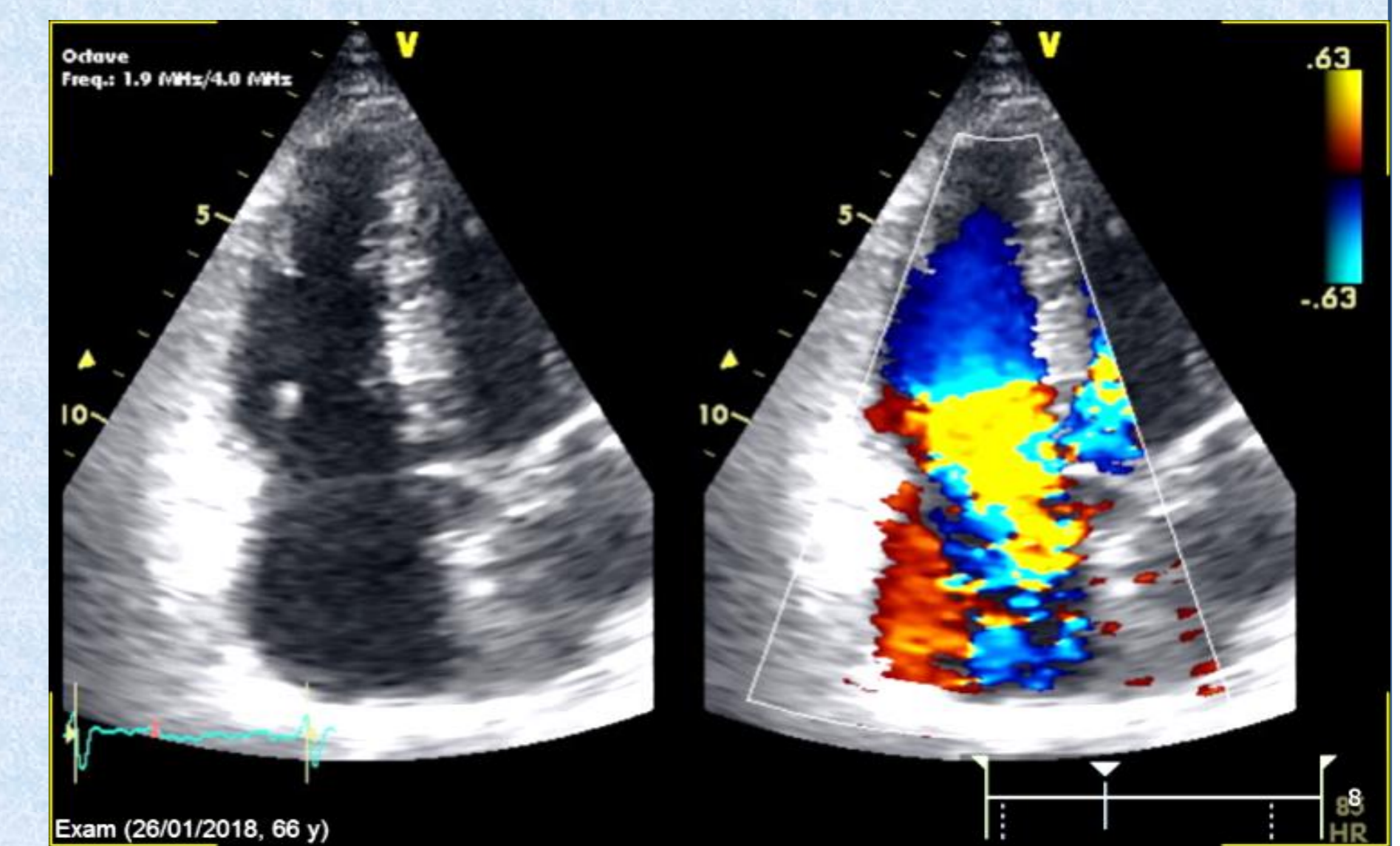
A previously fit 66 year old female, presented with a 6 month history of shortness of breath and ankle oedema. Echocardiogram revealed severe fixed tricuspid regurgitation, pulmonary valve disease, dilated right heart chambers and preserved left ventricular function. Carcinoid heart disease was suspected and she was referred to our neuroendocrine tumour clinic. She gave a seven year history of facial flushing and diarrhoea.

Examination revealed a fixed violaceous facial flush, injected red eyes and evidence of right heart failure. The liver was pulsatile and a large pelvic mass was palpable.

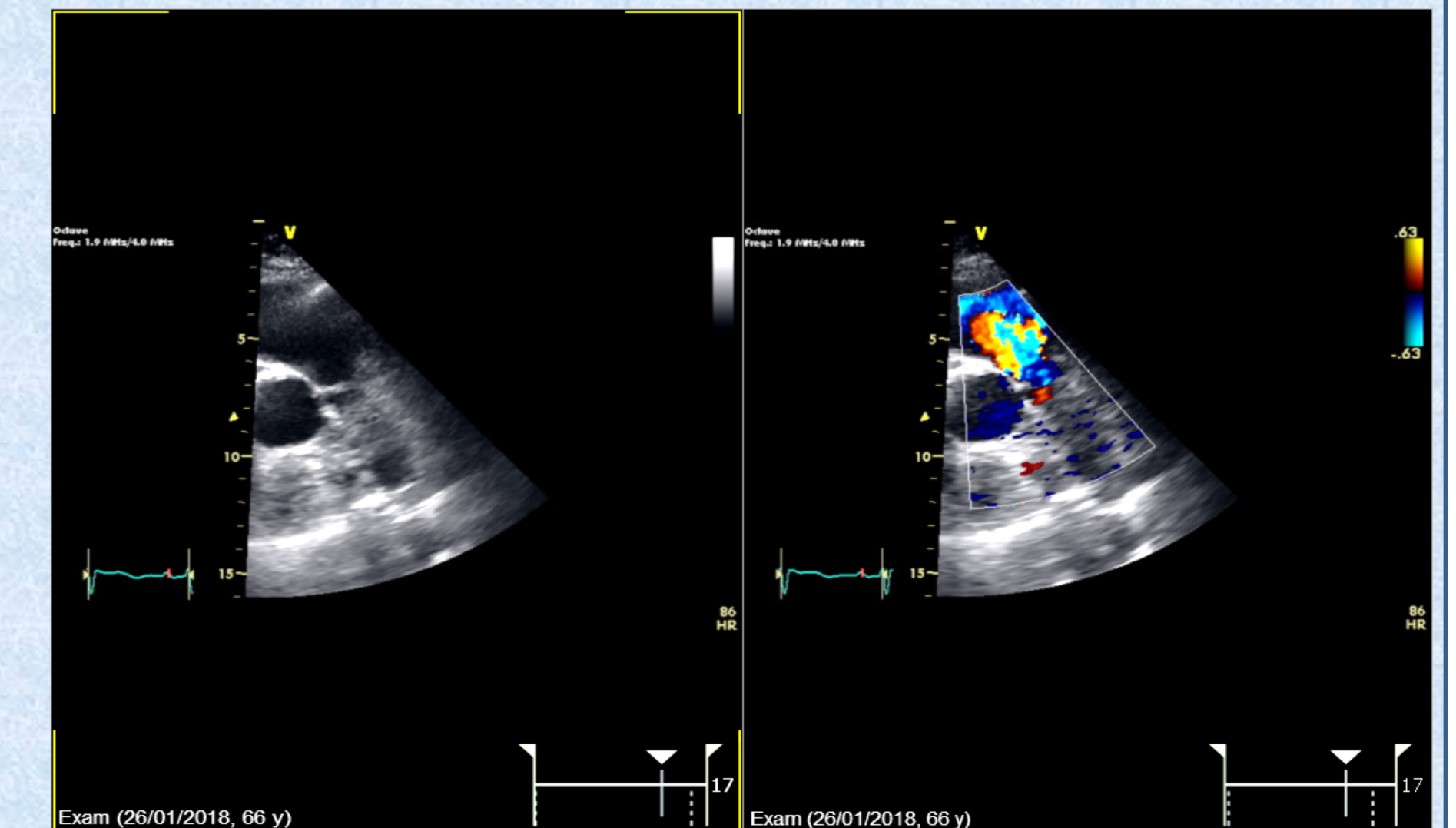
Investigations revealed a chromogranin A of 242 nmol/L (0-6) and 5-hydroxyindolacetic acid (5HIAA) at 857 umol/24 hrs (5-37). CT and MRI showed a large solid/cystic 15 cm mass arising from the right ovary, avid on Octreoscan. No lesions were noted in the bowel or liver.

Her carcinoid syndrome was controlled with octreotide 50 mg thrice daily subcutaneously with cross over to lanreotide 120 mg deep sc injection every 28 days. Her right heart failure was managed with bumetanide 1 mg daily. After careful review in the Neuroendocrine MDT including gynaecology, cardiology, anaesthetic and HPB surgical review; a joint procedure with the neuroendocrine surgical team and gynaecologist was performed. She underwent an uneventful oophorectomy under octreotide cover (starting at 50mcg/hr the day before surgery).

Histology revealed an ovarian insular carcinoid tumour with lympho vascular space invasion, ENETS grade 1 (KI 67 <1 %). Her 24hr urinary 5HIAA, and chromogranin A, 6 weeks after surgery normalized to 17 umol/24 hrs and 1.8 nmol/L. However she declined cardiac valve surgery.



2DEcho Apical four chamber view - dilated right heart & TR



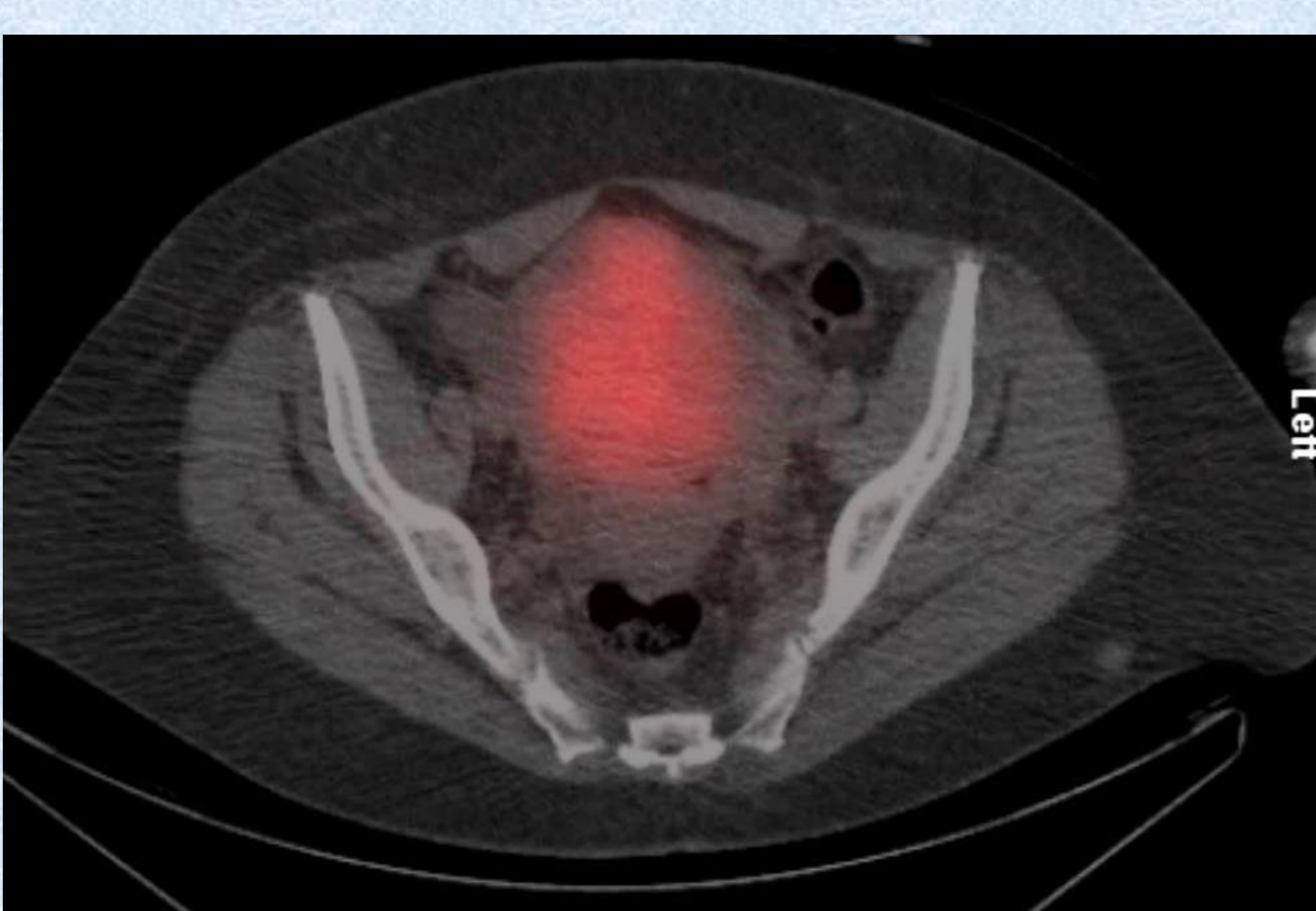
2DEcho Parasternal short axis view showing PR



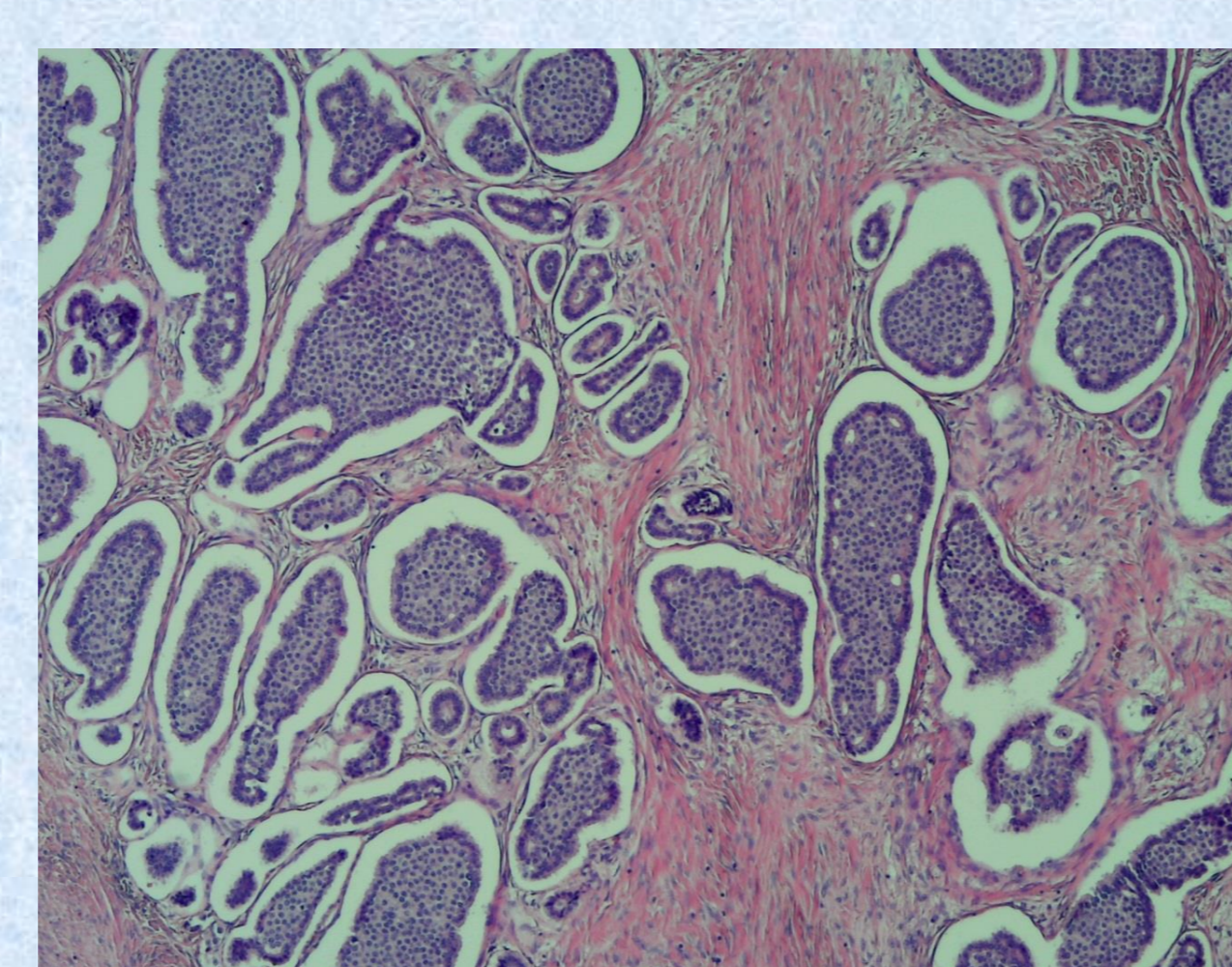
MRI sagittal view of pelvic mass



Octreoscan whole body – increased uptake in pelvic mass



Octreoscan axial view – increased uptake in pelvic mass



H&E staining of ovarian tissue showing evidence of insular carcinoid

## Discussion

Vasoactive substances secreted from the carcinoid tumors give rise to the systemic manifestations of carcinoid syndrome as well as carcinoid heart disease. These complicate less than 10% of the rare ovarian NETs (3). Most NETs have to metastasize to the liver to result in symptoms of carcinoid syndrome; however, primary ovarian carcinoid tumors are unique in this regard because the venous drainage of the ovaries bypasses the portal venous system (4). Ovarian NETs are divided into trabecular, strumal, mucinous and insular types, the latter being the most common and the only one associated with carcinoid syndrome.

## Conclusion

Ovarian carcinoid presenting with carcinoid heart disease and syndrome is very rare and needs to be considered in females who present with right heart valve disease. This case highlights the need for MDT discussion in patients with NET.

## References

1. Damen N. Ovarian carcinoid presenting with right heart failure. *BMJ Case Rep.* 2014;28:pii: bcr2014204518
2. Mordi IR, Bridges A. A rare case of ovarian carcinoid causing heart failure. *Scott Med J.* 2011;56:181
3. Shin HW, Kim H, Yoon HJ, et al. Ovarian tumor-associated carcinoid heart disease presenting as severe tricuspid regurgitation. *J Cardiovasc Ultrasound.* 2011;19:45–49
4. Artaza A, Beiner JA, Gonzalez M, Aranda I, de Teresa EG, Pulpon LA. Carcinoid heart disease: report of a case secondary to a pure carcinoid tumour of the ovary. *Eur Heart J* 1985;6:800-5