

Management of Neuroendocrine Tumor with Carcinoid Heart Disease and Liver Metastases: Multidisciplinary Approach

Background and proposes

Heart is frequently involved in oncological processes, either by direct tumor existence/activity or secondary to antitumor treatments. This case exemplified how cardio-oncologists, being part of multidisciplinary team, reach a consensus management, key to success.

Case description



- 46 years-old female, no medical history.
- Clinical course: 4-5 months with progressive dyspnea, occasional palpitations and hot flushes.
- Physical examination: **Systolic murmur tricuspid area.**

Transthoracic echocardiogram

- Pulmonary and tricuspid valves: **Retracted/thickened veils.**
- **Free tricuspid regurgitation.** Double mild pulmonary valve lesions.
- **Severe right ventricle (RV) dilatation:** volume overload signs.

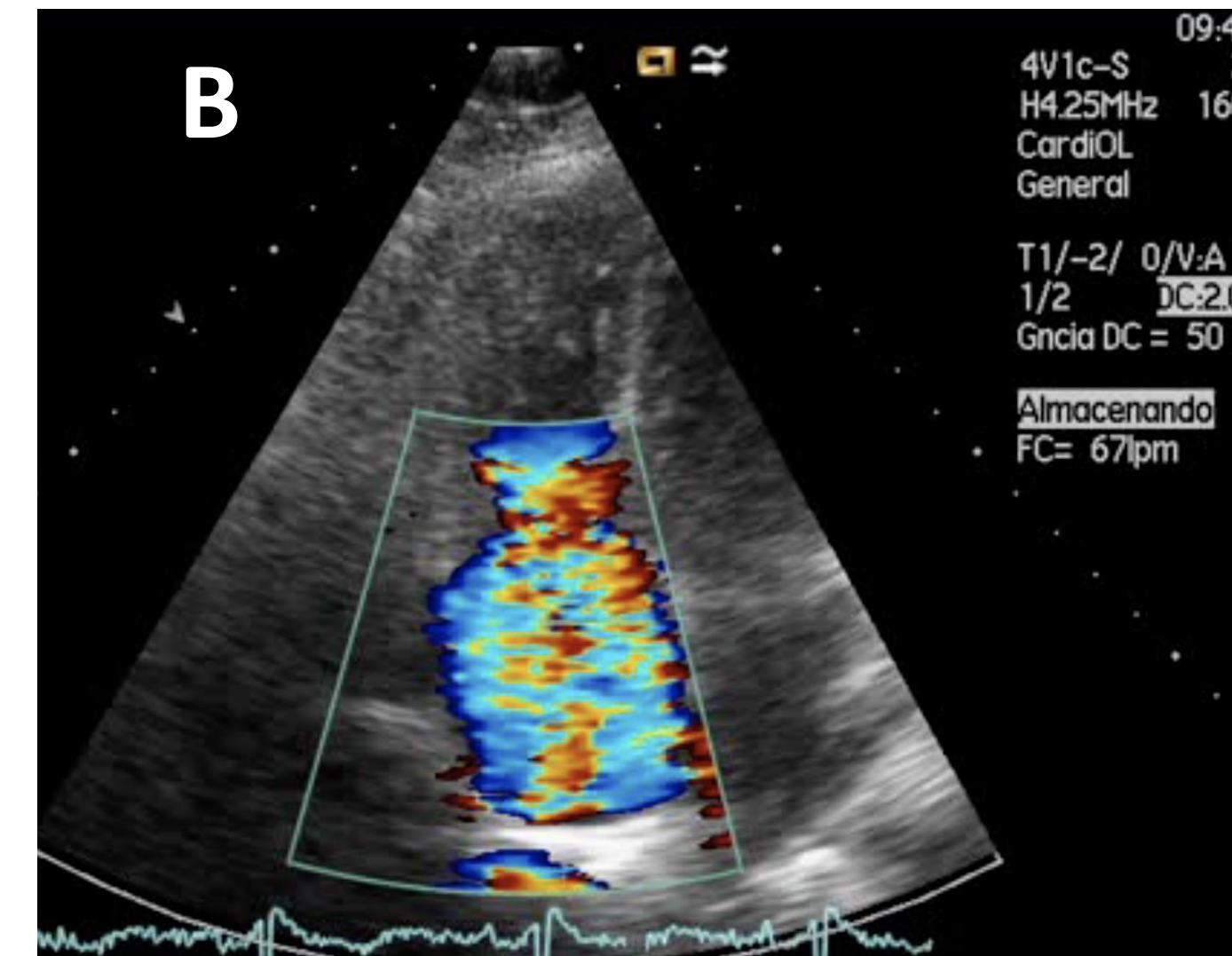
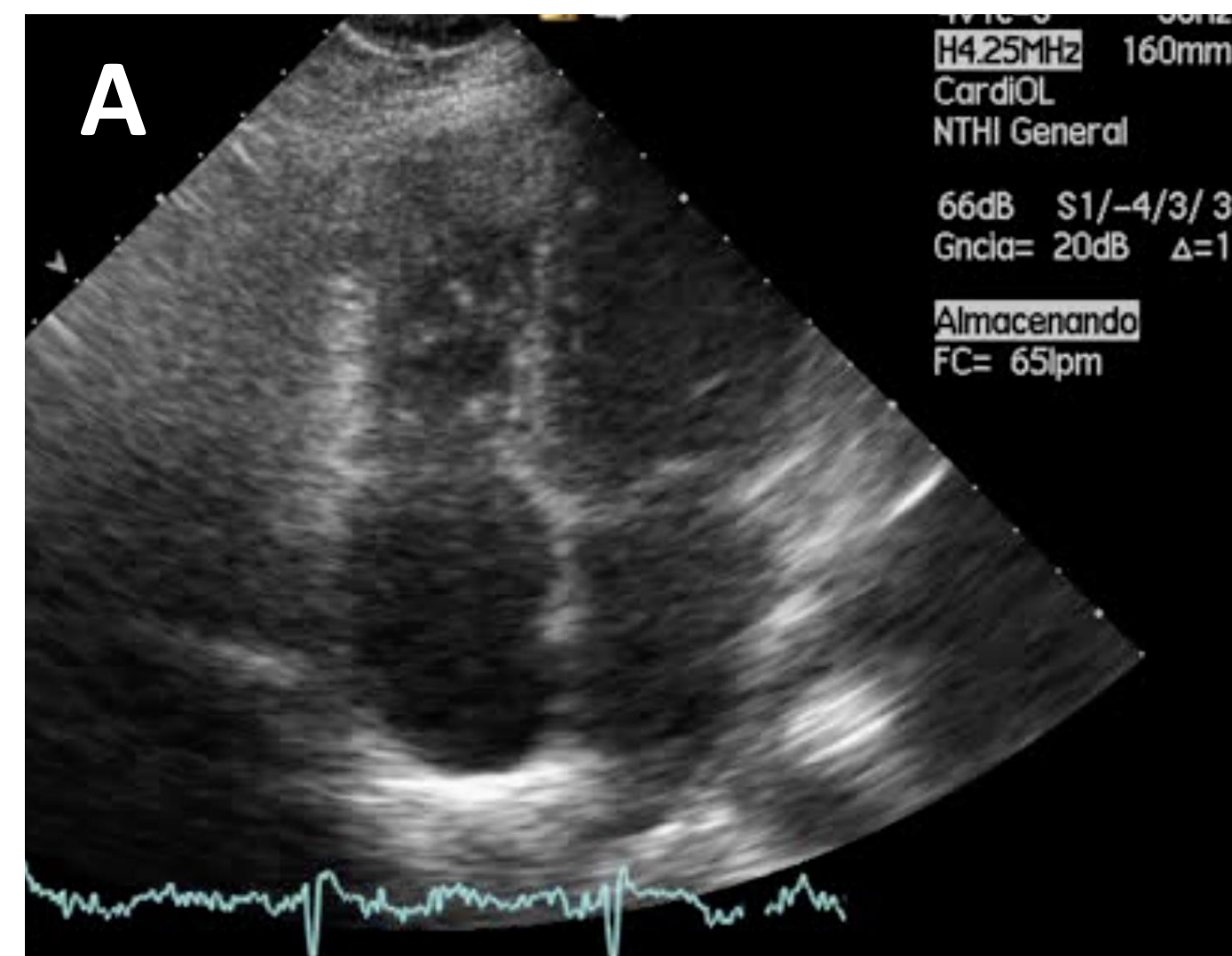


Image 1. A: Echocardiogram apical 2-chambers view (RV-focused,) showed tricuspid affection. **Image B:** Free severe regurgitation in Doppler color flow.

Cardiac magnetic resonance:

- Severe tricuspid regurgitation (**RF 43%, coaptation defect 9mm**).
- Mild dilated right ventricle (**198cc / i 108cc/m2**).
- Ventricular septum rectification.
- LVFE / RVFE (**63%**) preserved.

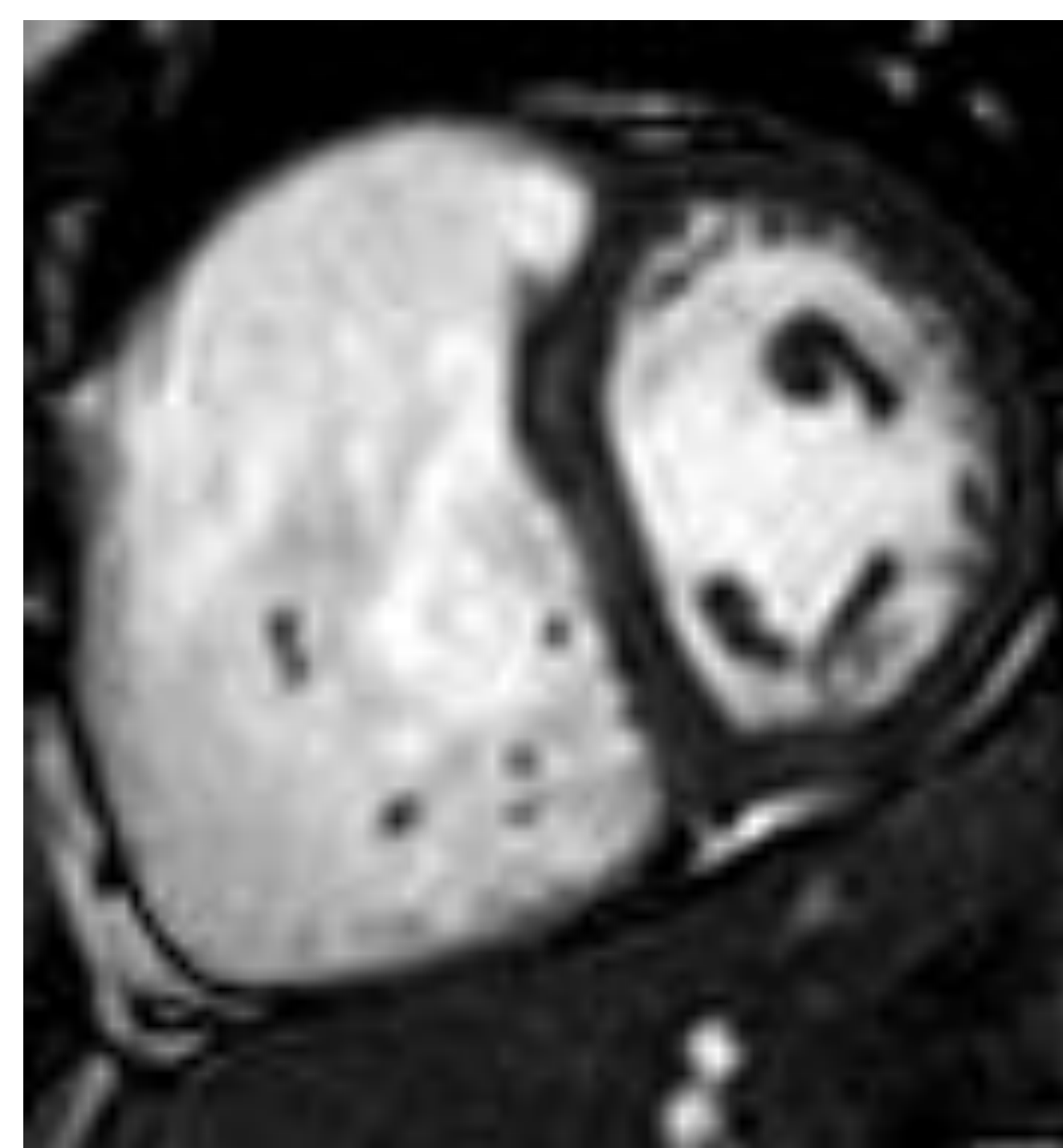


Image 2: Short-axis cine sequence (mid-ventricle), VD overloaded, with interventricular septum rectified.

Suspicion of carcinoid syndrome

Biochemical test:

↑ **Chromogranin-A:** 593 ng/ml [0.0 -100]
↑ **5-hydroxyindoleacetic:** 20.85 mg/24h [2-9].

CT-Body:

Primary tumor in ileum + liver metastases.

PET-CT:

↑ **Metabolic activity.**

OctreoScan:

↑ **Somatostatin's receptors.**

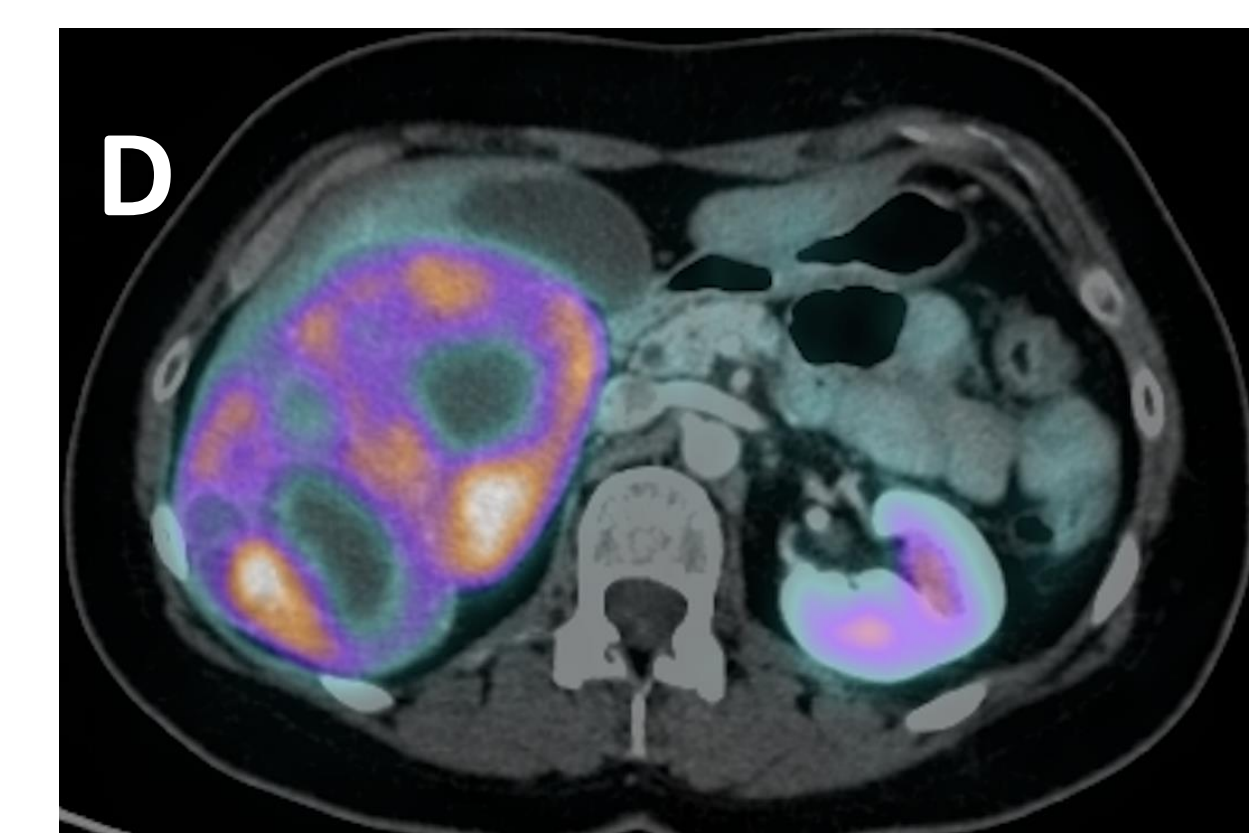
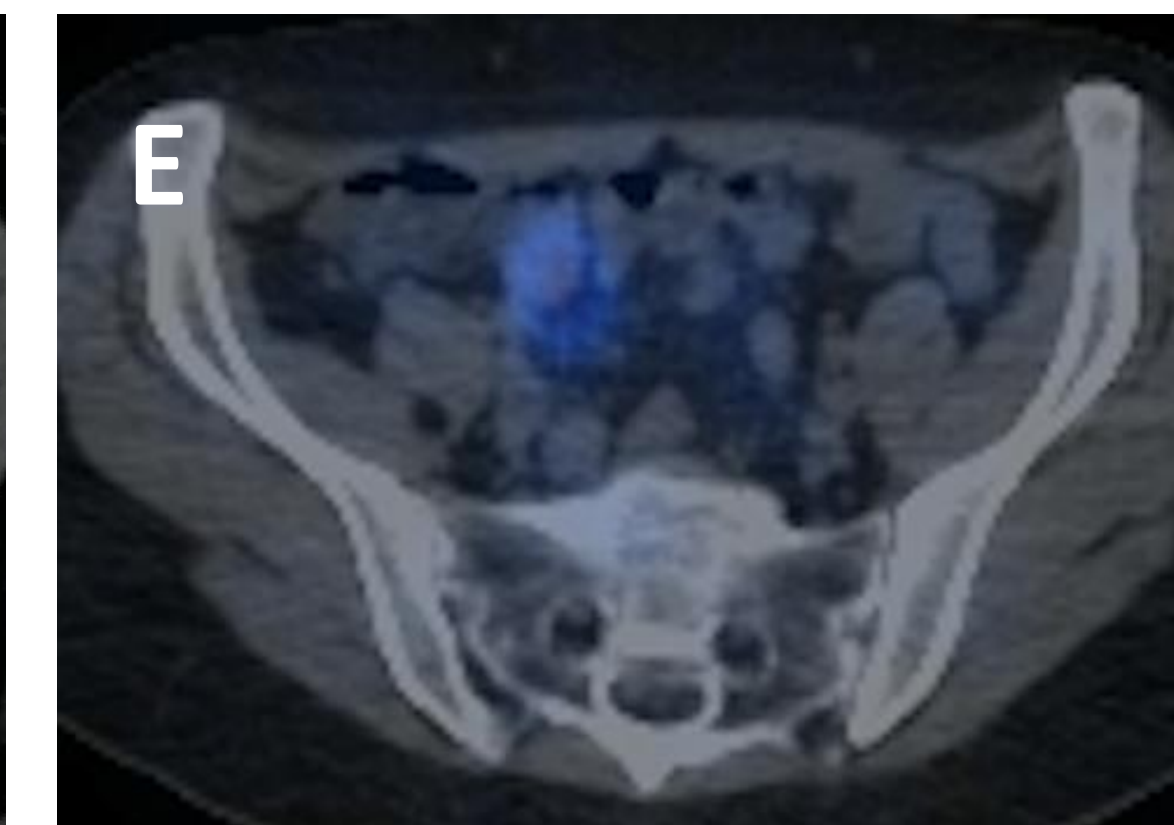
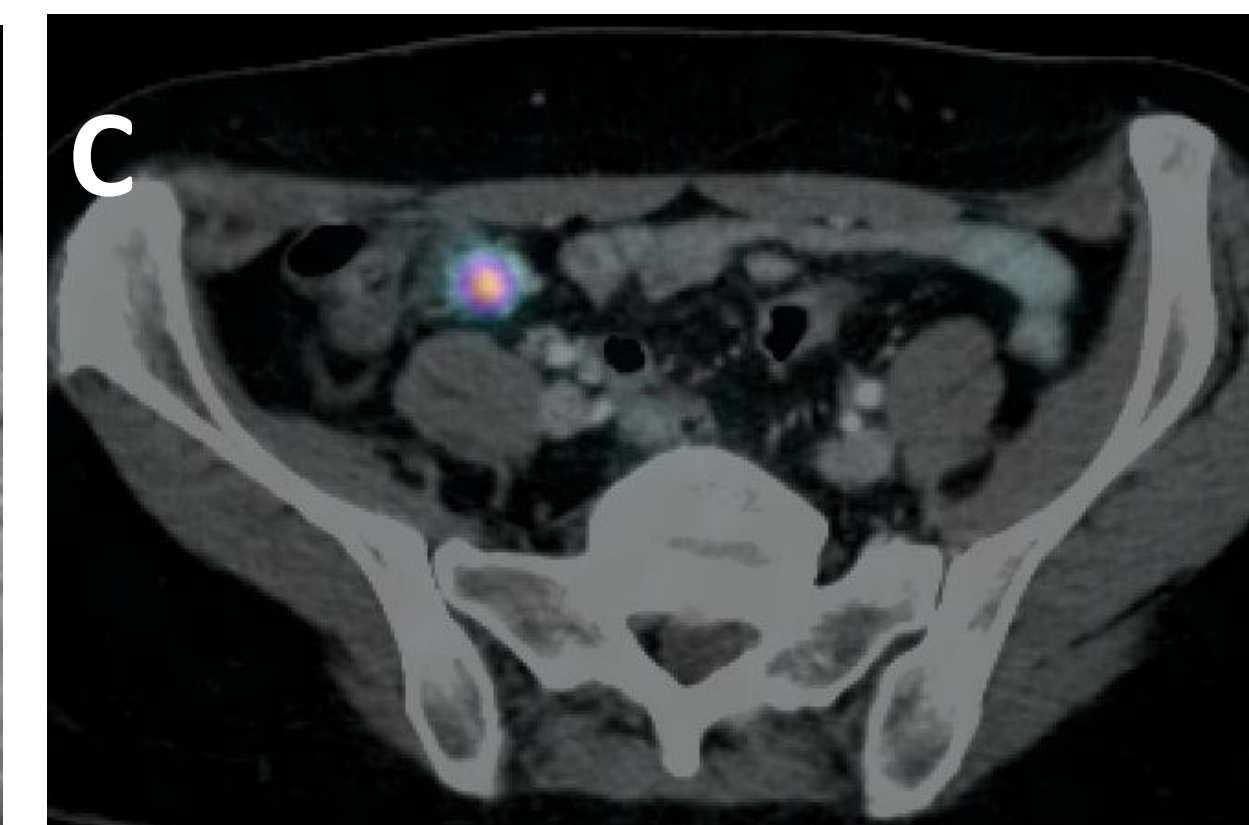
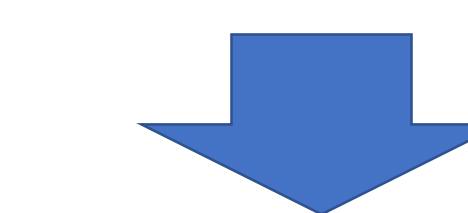


Image 3. A: Coronal CT, primary tumour in distal ileum (red arrow) . **Image B:** Axial CT cut multiple liver metastases. . Axial PET-CT, high activity in primary tumour (C) and liver metastases (D) and OctreoScan somatostatin receptors in primary tumour (E) and liver metastases (F).

Liver biopsy:

Well-differentiated low-grade neuroendocrine tumor (LGNET) (Ki67 1%)



DIAGNOSIS:

CARCINOID SYNDROME SECONDARY TO GASTROINTESTINAL CARCINOID TUMOR.

CARCINOID SINDROME WITH HEART VALVE AFFECTATION

Prognosis:

Determined by **heart involvement + metastases.**

Treatment strategy:

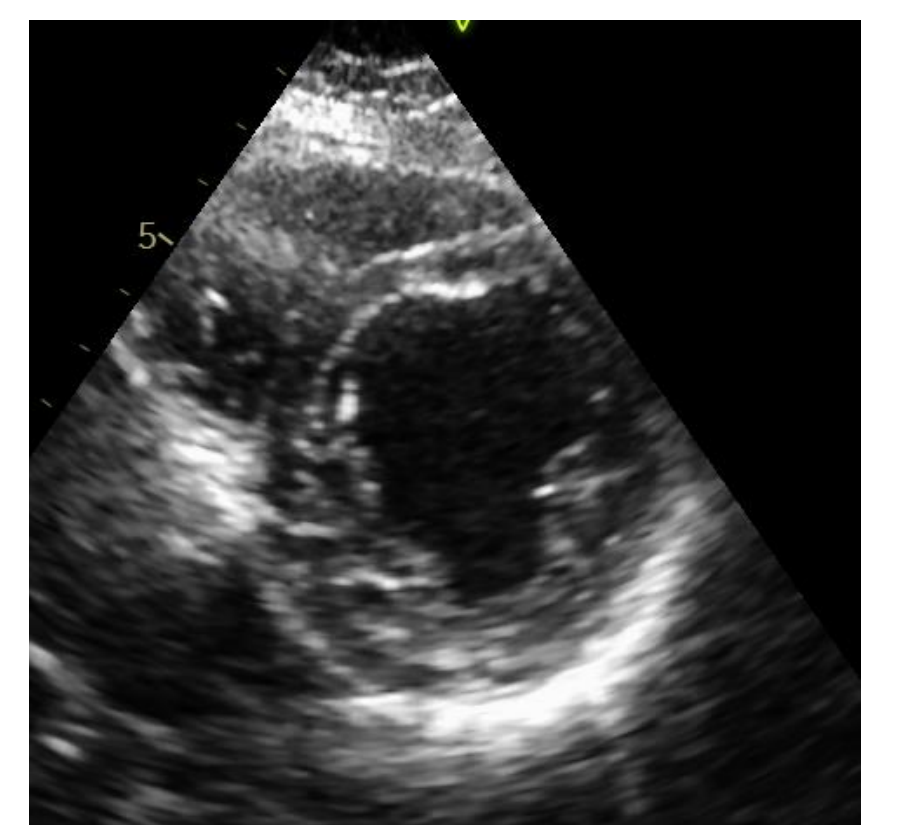
- **Symptoms:** Medical treatment with Somastatine analogues (Lanreotide 150mg/1 month).
- **Curative treatment** (+ carcinoid crisis prophylaxis):
 1. Heart valves: Replacement surgery.
 2. Tumor: surgical exeresis + metastases.

KEY TO SUCCESS:
Multidisciplinary team
(Cardio-oncologist, oncologist, general surgery, endocrinologist)

Follow-up: 6 months

Cardiopathy

- Asymptomatic CV.
- **RV Reversal remodeling.**
- Normofunctional prothesis.



Neuroendocrine tumour

- Follow-up: oncology + general surgery + endocrinology.
- Control CT: tumor stability. Asymptomatic.
- Tumor-board decision: **Pending surgery of right liver metastases (curative targetting).**

Biological tricuspid and pulmonary valvular surgical replacement

3 months

Laparoscopic resection surgery:
TNE + left liver metastases.

3 months

Right liver metastases radioembolization (Y90)

Discussion

LGTNE are rare slow-growing tumors, originate in cells of the diffuse neuroendocrine system, more frequently in gastrointestinal and respiratory systems, **75% with metastases at diagnosis**. CS, secondary to hormonal overproduction, appears in 20-30% of patients: flushing, abdominal pain/diarrhea, bronchoconstriction. More than 50% with CS have cardiac involvement (associated with reduced survival): **tricuspid almost all**, 50% pulmonary and 90% right chambers dilatation. **Time for surgery, prosthesis choice**, anticoagulation, somatostatine analogues treatment and serotonergic crisis prevention during tumor surgery are aspects that require an individualization of each case and be **discussed by multidisciplinary team**.

References: 1. Hayes AR, et al. *Endocrinol Metab Clin North Am.* 2018;47:671–682. 2. Davar J, et al. *J Am Coll Cardiol.* 2017;69:1288–1304. 3. Ito T, et al. *Curr Opin Endocrinol Diabetes Obes.* 2018;25:22–35.

Disclosures

No relevant financial relationship to declared.