Case 4: Answer

from University of Tsukuba Hosp.

We deeply appreciate Dr. Masato Sugano, Dept. of Pathology, Univ. of Tsukuba Hosp. for his assistance of this presentation.

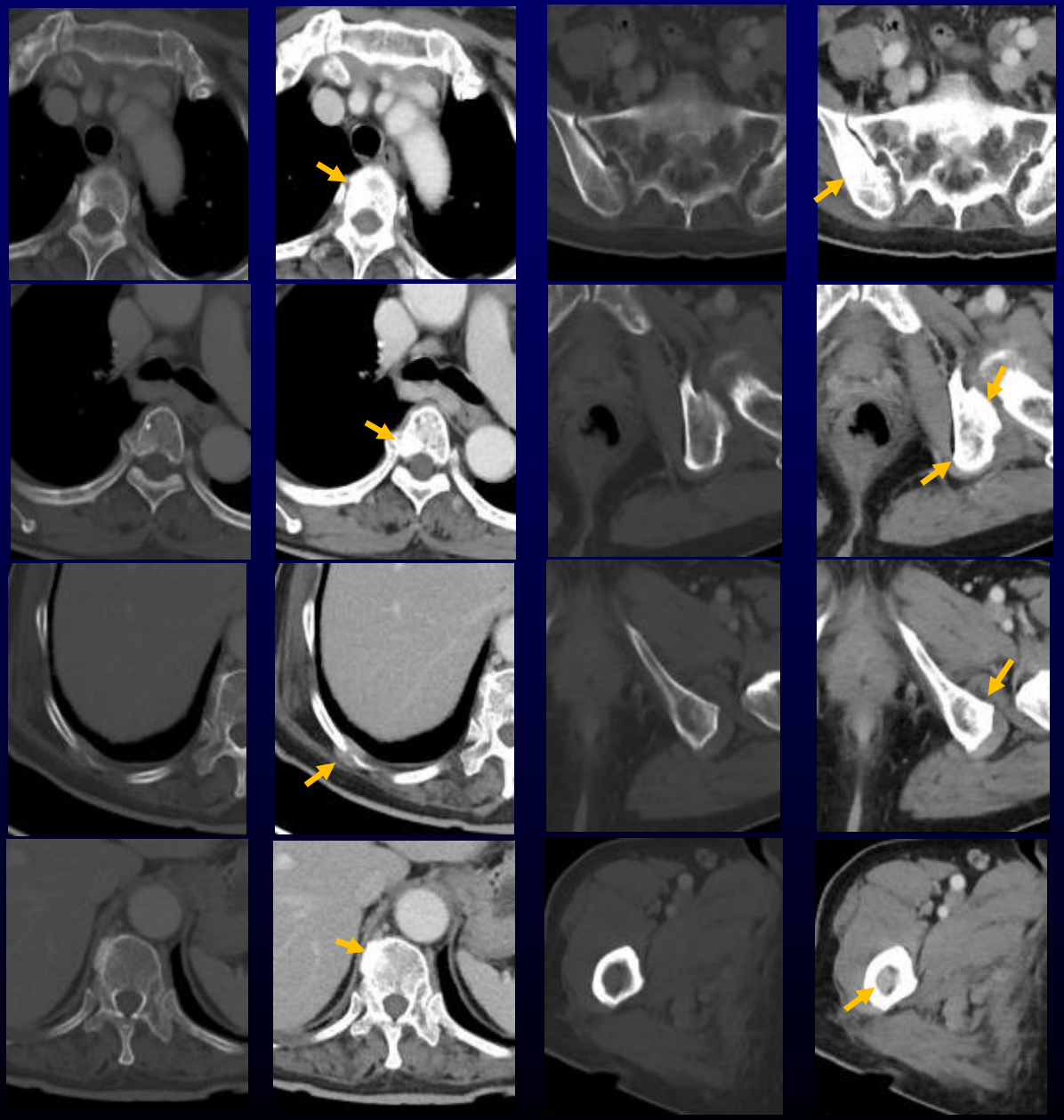
Summary: clinical & imaging findings

- 73F, persistent painless facial edema beginning from the eyelid
  →heart failure?? SVC syndrome?
- Recently right back pain

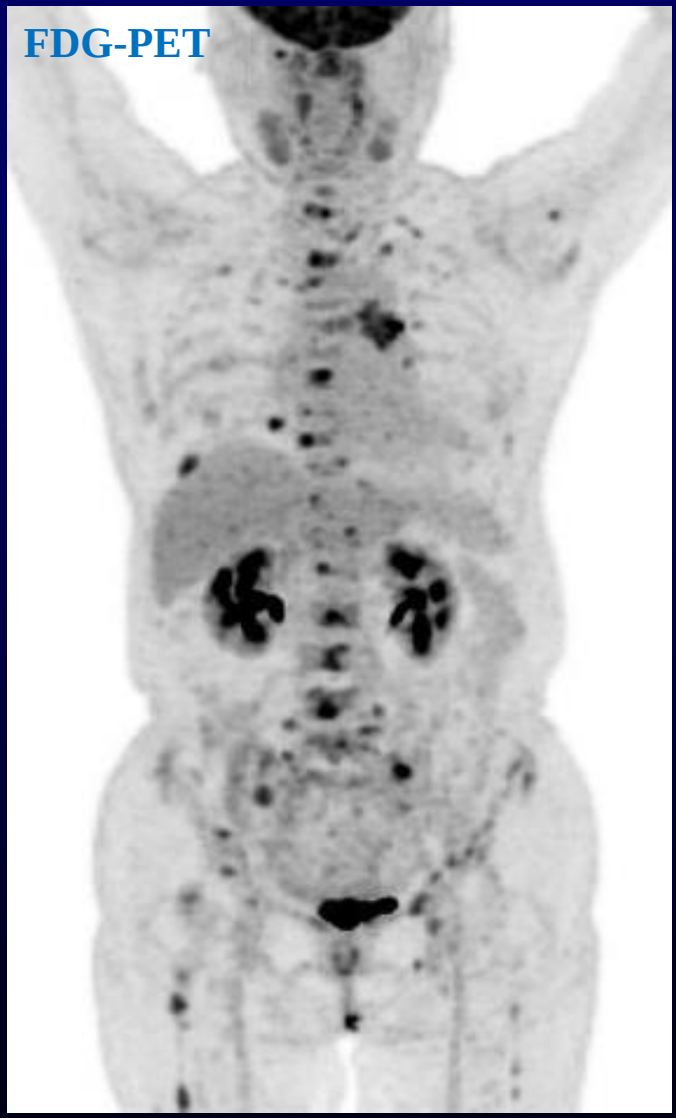
- Anterior mediastinal mass with lobulated margins
- Moderate enhancement, nearby LN swelling
  → malig, mediastinal tumor
- No calc./fat
- RA & RV dilatation, normal PA
- SVC & azygos dilatation, no SVC synd.
  → s/o tricuspid regurgitation
  → cardiac US: mild TR with TV degeneration
- fluid in the descend. colon → diarrhea?

Then what is the cause of her rt. back pain? Bone mets.?

bone window ST window bone window ST window



FDG-PET



Searching bone mets. in the soft tissue window setting of postcontrast CT

Advantages

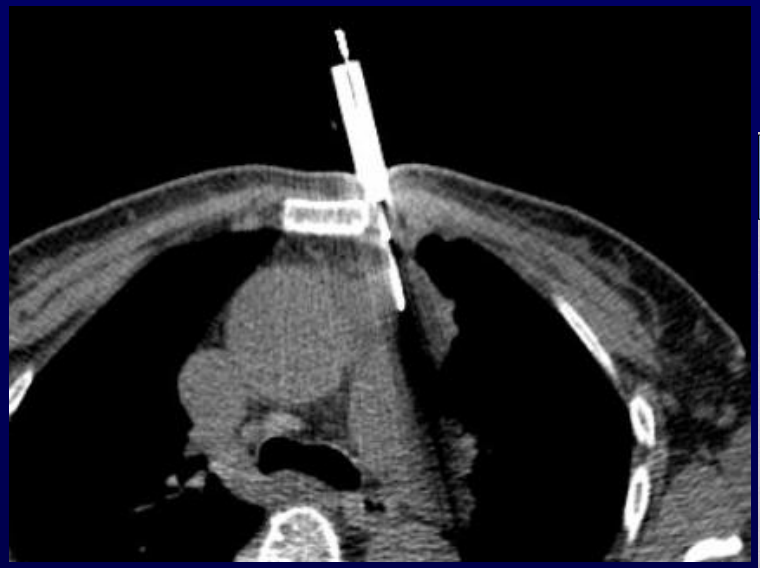
- **can identify tumors themselves, not as the mold of bony changes**
- **can see periosseous soft tissue mass & BM mets.**
- **can evaluate spinal canal extension**
- **can detect skeletal abnormalities by narrower window setting, esp. sclerotic changes**
- **no need to change window settings**

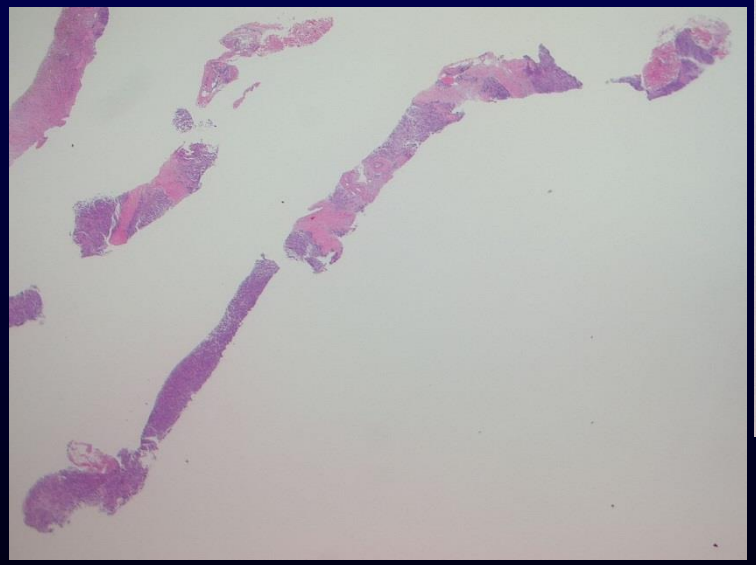
Disadvantages

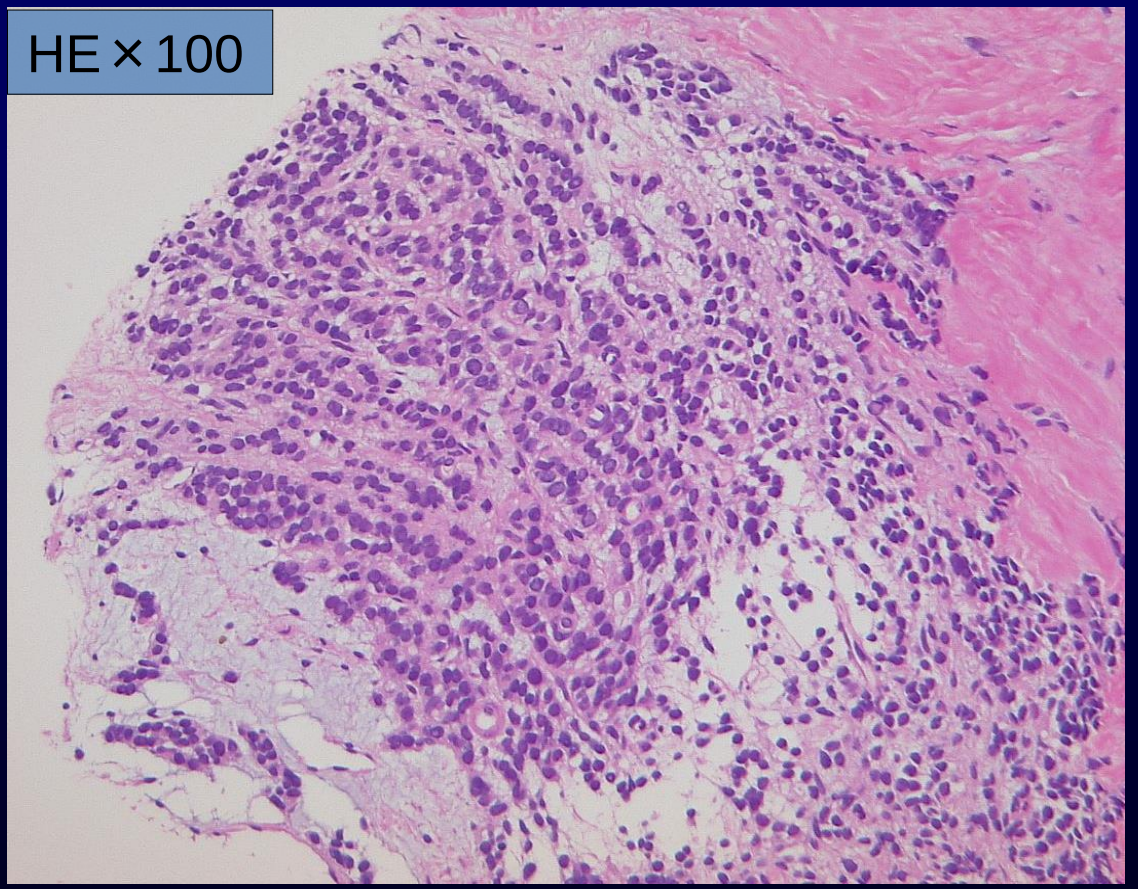
- ***need training to be accustomed to***
- ***not applicable below age of 45***
- ***less sensitive in the upper spines***
- ***cannot characterize bone lesions***

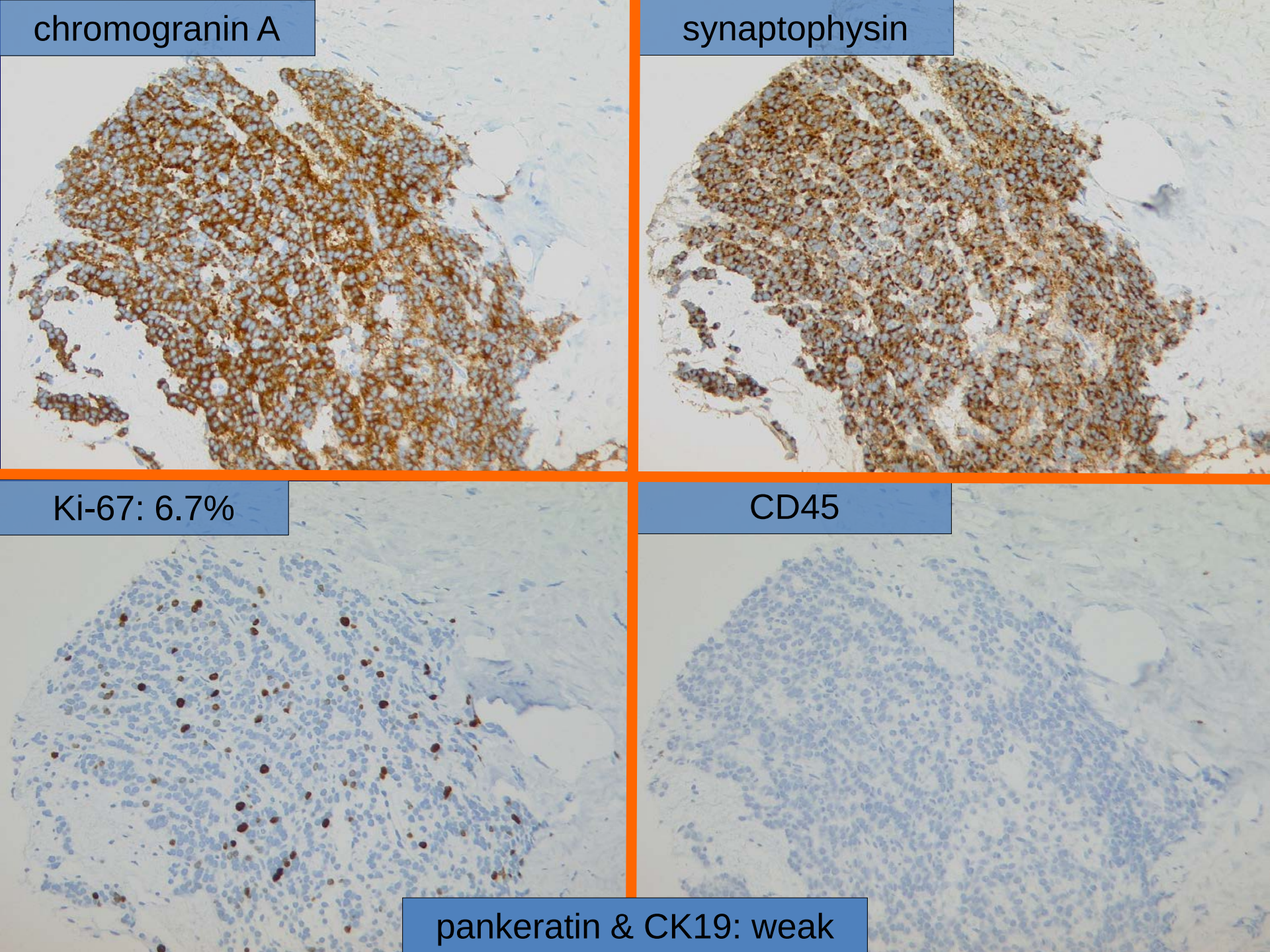
You need to see the bone window setting after detection.

Next, we did CT-guided biopsy of the mediastinal tumor.








chromogranin A
synaptophysin
Ki-67: 6.7%
CD45
pankeratin & CK19: weak

Final Diagnosis

Thymic atypical carcinoid
with
Carcinoid syndrome & bone mets.

Skin biopsy of the face showed marked mucoid deposition.
After close Hx taking, it became evident that she had frequently experienced diarrhea & facial flushing.
5-HT was not elevated in this patient.

Thymic Carcinoid (NET grade 1/2)

- thymic carcinoid: malignant neoplasm, neural crest origin, thought to arise from amine precursor uptake & decarboxylase (APUD) cells.
- average age 45, M:F = 3:1
- symptoms/signs: may relate to mass effect/invasion, SVC syndrome, Cushing syndrome (~ 40%), carcinoid syndrome
- imaging: dominant anterior mediastinal mass ± calc. &/or local invasion, LN & distant met. more common (50-65% pts; lung, liver, brain, bone) & pleural met. less common than thymoma, often hyperenhancing on CT, PET/CT for detection of met. & surveillance: limited by low proliferative activity, Octreoscan (++)
- DDx: thymoma, malig. germ cell neoplasm, Hodgkin lymphoma, metastatic lymphadenopathy
- up to 20% with MEN syndromes, foregut carcinoids with 10% of type 1 MEN & rarely with type 2 MEN/type 1 NF
- 5-year survival rate 65%

Carcinoid syndrome

- hypersecretion of vasoactive substances (mostly serotonin [5-hydroxytryptamine: 5HT] , esp. in midgut tumors, sometimes bradykinins, tachykinins, histamine, substance P, & ACTH)
- flushing, diarrhea & bronchospasm (aka. Thorson-Bioerck syndrome, Cassidy-Scholte syndrome)
- cardiac involvement (50%), US/MR/CT: thickened, retracted, highly reflective TV &/or PV leaflets, enlarged RA & RV
  ← direct release of secretory products into systemic circulation or overwhelmed hepatic metabolism
- 2nd-ranked cause of TS (59%) (always concomitant TR, 97%)
- most commonly midgut origin (ileocecal region/appendix), sometimes foregut origin (lung) .
- fibrous plaques: composed of smooth muscle cells mixed with mucopolysaccharide & collagen ← mediated by serotonin 1B receptor subtype probably inducing fibroblast proliferation

Take Home Points

- Thymic carcinoma, thymic carcinoid, & thymoma are not reliably differentiated on imaging.

- Distant metastases, esp. sclerotic bone metastases, & lymphadenopathy are more common in thymic carcinoid. Carcinoid syndrome may be suggested on imaging.

- Bone metastasis can be surveyed in the soft tissue window setting of postcontrast CT more effectively than in the bone window setting.

References

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