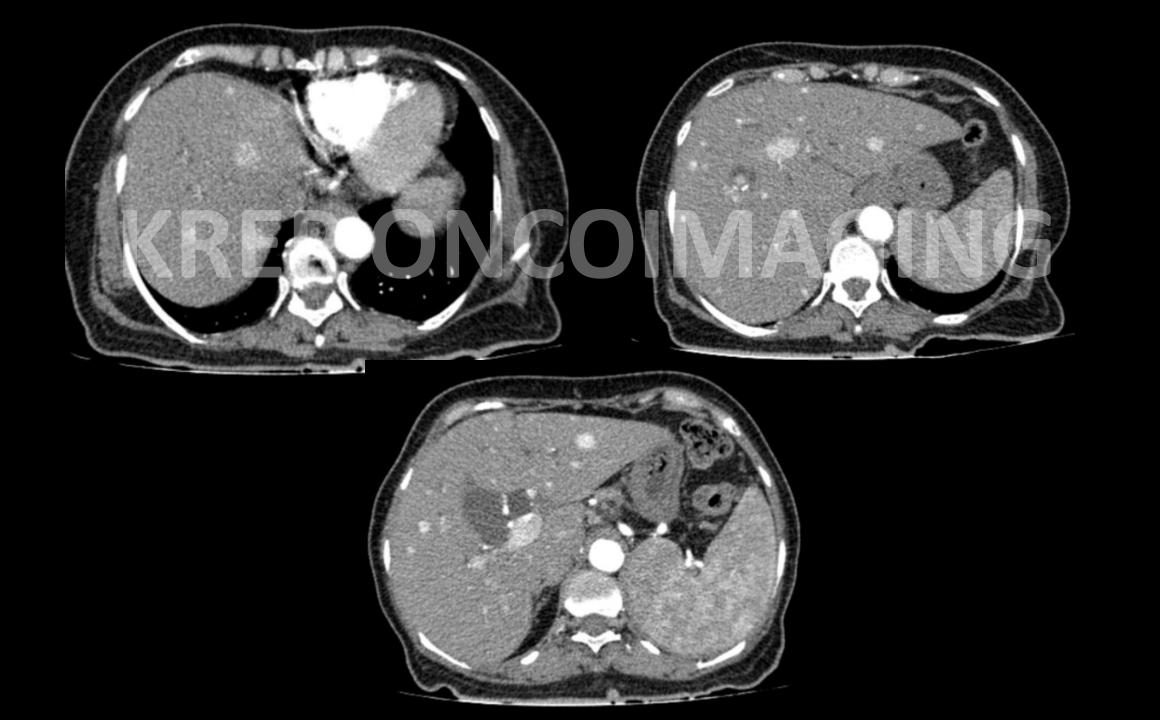


2025

KARNATAKA RADIOLOGY EDUCATION PROGRAM











- Lobulated mass in posterior to head of pancreas measuring 3.5 x 7 x 3 cm (AP x TS x CC) showing intense early arterial phase hyperenhancement with washout on early portal / late arterial and venous phases. There are multiple arterial feeders from celiac axis.
- There is a large tumor thrombus in the main portal vein causing significant occlusion.
- SMA is encased with an arc of contact of about 180 degrees.
- There is moderate communicating IHBRD due to mass effect from the lesion.
- Multiple arterial phase hyperenhancing lesions are noted in both lobes of the liver, suggestive of hypervascular hepatic metastases.
 - Features are suggestive of malignant NET (neuroendocrine tumor) with portal vein tumour thrombus and multiple hepatic metastasis with moderate communicating IHBRD.

1. Pathology & Origin:

- Neuroendocrine tumors arising from paraganglia of the sympathetic or parasympathetic chain outside the adrenal medulla.
- Sympathetic paragangliomas (abdomen/pelvis) are usually functional (catecholamine-secreting);
 parasympathetic (head/neck) are usually non-functional.

2. Common Locations:

- Retroperitoneum along the aorta, organ of Zuckerkandl (aortobifurcation), para-aortic chain, carotid space (carotid body tumor), jugulotympanic region, mediastinum, and bladder wall.
- Extra-adrenal sites account for ~10–15% of all pheochromocytoma/paraganglioma spectrum tumors.

3. Clinical Presentation:

- Functional tumors: episodic hypertension, palpitations, headache, diaphoresis, and tachycardia.
- Non-functional tumors: mass effect symptoms (pain, hoarseness, dysphagia), or incidentally discovered.

4. CT/MRI Morphology (Key Imaging Hallmarks):

- Well-defined, hypervascular soft-tissue mass with strong arterial enhancement.
- MRI:
 - T1: iso- to hypointense
 - T2: markedly hyperintense ("light-bulb bright" in many cases)
 - Post-contrast: avid, heterogeneous enhancement
- Large lesions may show necrosis, hemorrhage, or cystic change.

5. Functional Molecular Imaging:

- 68Ga-DOTATATE PET/CT is preferred (somatostatin receptor imaging) highest sensitivity for multifocal and metastatic disease.
- FDG-PET/CT is useful in SDHB mutation-associated tumors (more aggressive).
- MIBG scintigraphy used when considering MIBG therapy.

6. Genetic Associations (Very Important Clinically):

- Up to 30-40% are hereditary.
- Strongest association: SDHB mutation, which confers high metastatic risk.
- Others: VHL, RET (MEN2), NF1, SDHD, SDHC.
- Genetic testing is recommended in all patients, especially if <45 years or multifocal/metastatic.

7. Patterns of Spread & Malignant Potential:

- No histologic feature reliably distinguishes benign from malignant malignancy is defined by metastasis (lymph nodes, bone, lung, liver).
- SDHB-associated tumors are more likely to metastasize.

8. Oncoradiologist Reporting Priorities:

- Exact anatomic origin and relation to major vessels/nerve plexuses.
- Local invasion vs displacement.
- Multifocality (scan from skull base → pelvis).
- Functional imaging avidity (DOTATATE vs FDG) → helps determine therapy route.
- Suggest biochemical correlation (plasma metanephrines) and genetic counseling.

Metastases

Although the majority of liver metastases are hypodense and enhance less than the surrounding liver, metastases from certain primaries demonstrate an increase in the number of vessels, resulting in a hyperechoic ultrasound appearance, and arterial phase hyperenhancement on CT or MRI which washes out on delayed scan (cf. hemangioma which does not show washout). The primaries typically include:

- renal cell carcinoma (RCC)
- thyroid carcinoma
- neuroendocrine tumor (NETs)
 - gastrointestinal NETs
 - islet cell tumors
 - pheochromocytoma
- leiomyosarcoma
- choriocarcinoma
- melanoma
- breast cancer
- colonic carcinoma 5
- ovarian cystadenocarcinoma 5

Contributors

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