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A Rare Case of Appendicealcarcinoid: Importance of Pre Operative Diagnosis

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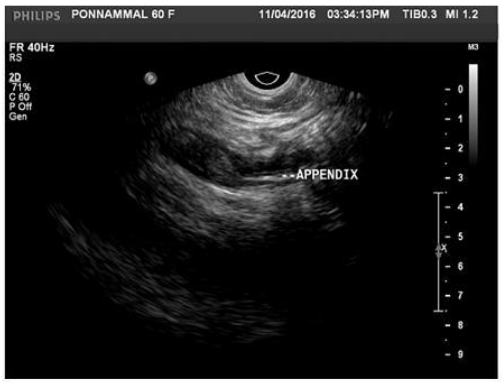
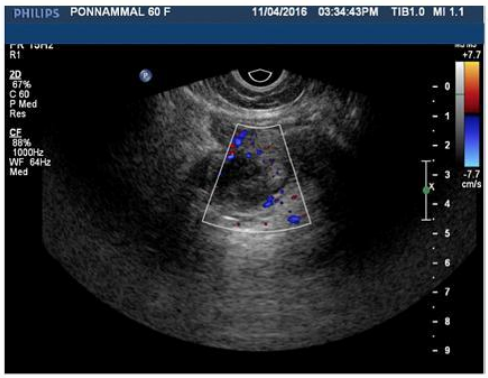
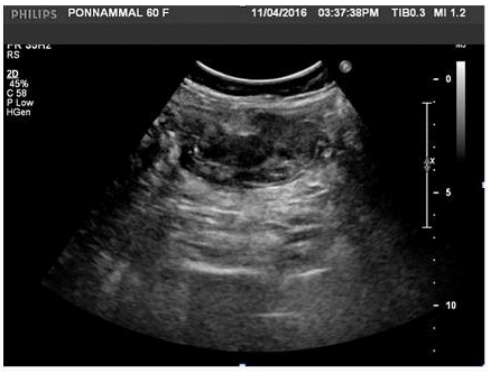
ABSTRACT
Appendicular carcinoid is a rare but most common neuro endocrine tumour of appendix, which arises from endocrine amine precursor uptake and decarboxylation (APUD) cells. It is often asymptomatic. Preoperative suspicion and diagnosis of appendicular carcinoid is very important. USG, CT and Octreoscanare very useful tools in its diagnosis. Optimal management of appendicular carcinoid could be achieved through accurate pre operative diagnosis. We present this case with classical signs of appendicular carcinoid seen in USG and CT.

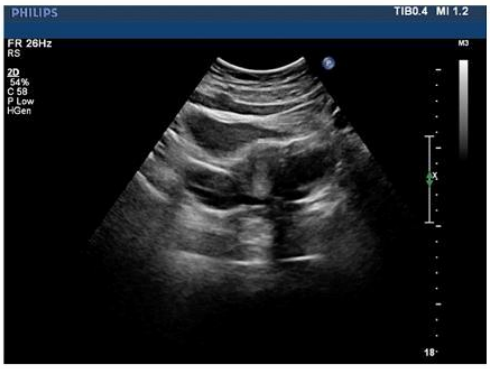
CASE REPORT

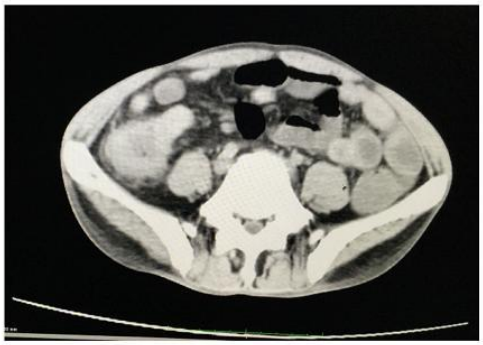
A 60 years old female presented to OPD with complaints of intermittent right lower quadrant pain of 6 months duration. On examination, she had mild tenderness in RIF. Plain X ray showed normal caecal air shadows. No abnormal radiopaque shadows were seen. On ultrasound, a small, well defined mixed echoic mass of size 2.2 x 1.2cm was seen in tip of Appendix. Colour Doppler showed minimal vascularity within the core of the mass lesion. Plain CT showed a small well-defined hypodensemass lesion of size 1.9x1.4 cm with CT value of 35 HU seen in tip of Appendix. Tiny specks of calcifications were noted within the mass. Marked surrounding fat stranding with mild retraction of distal ileal loops was seen. Based on imaging findings – a well defined hypodense mass in tip of Appendix with CT value of 35 HU and marked desmoplastic reaction and minimal vascularity noted in the core of the mass lesion in color Doppler, was suggestive of an appendicular tumour. This case is being presented for its rarity.

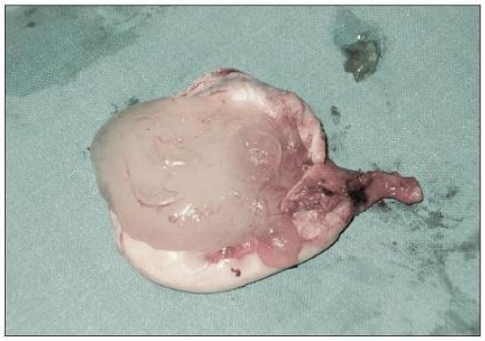


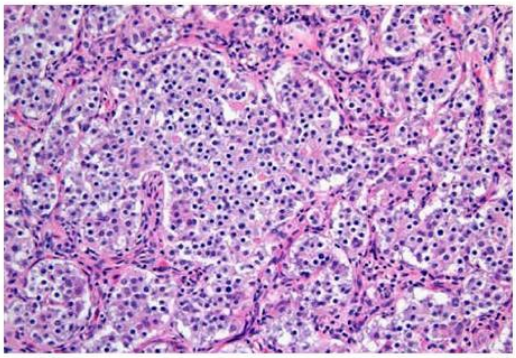












DISCUSSION

Definition

A carcinoid tumor starts in the hormone-producing cells that are normally present in small amounts in almost every organ in the body. A carcinoid tumor usually starts in either the GI tract or lungs, but it also may occur in the pancreas, testicles, or ovaries. An appendix carcinoid tumor most often occurs at the tip of the appendix. Approximately 50% of all appendix tumors are

carcinoid tumors. This type of cancer usually causes no symptoms until it has spread to other organs and often goes unnoticed until it is found during an examination or procedure performed for another reason.[1]

Incidence

Carcinoid tumours are found in 0.3–0.9% of patients undergoing appendectomy.[1,2] with mild male predominance.

Clinical findings

Most are asymptomatic in early development.

Most common presentation is episodic abdominal pain.

Laboratory findings

May products are secreted by carcinoids –

1. Serotonin
2. Histamine
3. Kallikrein
4. Prostaglandin
5. 5 – HIAA (Normal: less than 10 mg in 24 hours urine).

Serotonin mediatedesmoplastic change leading to fibrosis. Metastasis to mesenteric lymph nodes also produces endocrine substances. Fibrosis is characteristically "spoke wheel" towards the adjacent bowel loops causing them to pull closer.

CARCINOID SYNDROME

It is usually associated with liver metastasis.

HISTOLOGY

Two types of carcinoids are described.

1. classic type
2. goblet cell type (More aggressive)

Imaging of Appendicular carcinoid:[3,4]

Usually performed once biochemical diagnosis is confirmed typically by elevated 24 hour excretion of 5 – HIAA.

1. **X-ray and Barium findings:** Nonspecific.
2. **USG**

A smooth rounded asymmetric intra luminal mass lesion, mostly located in the tip of appendix.

3. **Abdominal CT with iv and oral contrast**

Visualization of primary tumour as a soft tissue mass in the region of tip of appendix, may show micro calcifications.

Lymph nodal enlargement – mesenteric, para aortic or retro peritoneal.

Radiating linear strands around the soft tissue mass due to fibrosis (Spoke wheel pattern).

Bowel wall thickening and retraction of adjacent loops.

Detection of liver metastasis, often multiple, may be hyper-vascular.

4. **MRI**

Useful for detection of metastasis.

Low in T1 and high in T2.

Enhances peripherally in arterial phase and hypointense during portal venous phase.

5. **OCTREOSCAN**

Octreotide is a stomatostatin analogue.

Carcinoid tumour cells almost always contain somatostatin receptors and may show increased uptake on scan.

Octreoscan is more sensitive than MIBG scan.

COMPLICATIONS:[1]

1. Carcinoid syndrome- occurs with liver metastasis.
2. Carcinoid crisis: (Severe flushing, changes in BP, Bronchoconstriction, Arrythmias, Confusion /stupor)
3. Carcinoid heart disease:

a) Right sided sub endothelial fibrosis
b) Tricuspid stenosis / Regurgitation
c) Pulmonic stenosis
d) Left side of the heart is protected by mono amine oxidase in lungs.

4. Intussusception
5. Small bowel obstruction
6. Small bowel schemia
7. Has an association with scleroderma.

TREATMENT:[1,2]

Surgical resection with mesenteric lymph node excision.

Appendiceal carcinoids have a more benign course than other GIT carcinoids, rarely metastasizing with a 5 year survival rate of more than 90%.

Appendiceal carcinoid tumours with a size smaller than 2 cm after radical resection need no further treatment because of minimal metastatic behavior.
Goblet-cell carcinoids have a worse outcome than the other types of carcinoid tumours and frequently present with metastatic disease.
Management of crisis is done by Octreotide.

CONCLUSION

Even though Appendiceal carcinoid is a rare entity, accurate Pre operative diagnosis with laboratory and imaging findings is very essential. It may prevent the development of Carcinoid syndrome. Optimal management of Appendiceal carcinoid could be achieved through accurate pre operative diagnosis.

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