

**Multimodality Radiological Assessment in Case of Rare Histopathologically Proven Retroperitoneal Desmoid Fibromatosis with Gastric and Pancreatic Invasion**

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**How to citation this article:** Dr Meghana M R, “Multimodality Radiological Assessment in Case of Rare Histopathologically Proven Retroperitoneal Desmoid Fibromatosis with Gastric and Pancreatic Invasion”, IJMACR – April – 2026, Volume – 9, Issue – 2, P. No. 08 – 11.

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**Type of Publication:** Case report

**Conflicts of Interest:** Nil

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**Abstract****Background**

Desmoid tumor is also known as aggressive fibromatosis are rare and comprise only 3% of all soft tissue tumors. They are benign fibromatous tumors characterized histopathologically by abnormal fibroblastic / myo-fibroblastic proliferation. although histologically benign, desmoid tumors are often locally invasive and associated with high local recurrence rate after resection.

**Case Report**

A 19yr old female patient complaints of prolonged abdominal pain, discomfort and nausea, however no history of vomiting. Menstrual history regular and normal. Bowel and bladder habits normal. No history of previous surgery. USG was done to find a large solid cystic mass thought to be arising from pancreatic tail and distal body with solid component showing significant internal vascularity.

CECT was done a large well encapsulated heterodense retroperitoneal lesion measuring approximately 15 x 12 x 10 cm( CC x TR x AP) , on post contrast showing minimal heterogeneous enhancement of solid component and non enhancing cystic spaces of fluid attenuation seen arising from normal enhancing body and tail of pancreas.

**Antero-Superiorly:** lesion is seen indenting greater curvature of stomach, gastro-duodenal junction and 1st part of duodenum resulting in complete obliteration of lesser sac.

**Right laterally:** it is seen displacing 2nd part of duodenum, celiac trunk and superior mesenteric artery causing kinking of splenic artery at its origin from celiac trunk.

**Posterior and left laterally:** Lesion is seen abutting abdominal aorta, splenic vessels, anterior surface of left kidney. Infrarenal portion of the lesion is seen extending

till left paraspinal muscles and posterolateral abdominal wall.

Lesion is causing displacement of bowel loops anterolaterally.

Solid pseudopapillary neoplasm of pancreas was given as probable diagnosis, on follow up of the patient post-surgery histopathological results came out as desmoid fibromatosis with invasion of gastric sub mucosa and distal pancreas.

### Conclusion

A rare presentation of locally invasive retroperitoneal desmoid fibromatosis in a young female with no previous history of surgery/oestrogen therapy, operated with distal pancreatectomy and subtotal gastrectomy done.

Subxiphoid view showing head of pancreas, neck, portal vein and body being seen as large iso-hypoechoic mass showing internal multiple anechoic areas. Mass measures 14.5 x 10 cm in maximum dimension.



Image 1



Image 2

### Case Presentation

A 19y old female patient complaints of prolonged abdominal pain, discomfort and nausea, however no history of vomiting. Menstrual history regular and normal. No history of previous surgery.

USG abdomen and pelvis of the patient was done to find a large retroperitoneal solid cystic mass thought to be arising from pancreatic tail and distal body with solid component showing significant internal vascularity. On spectral doppler arterial pulsatile waveform is seen.

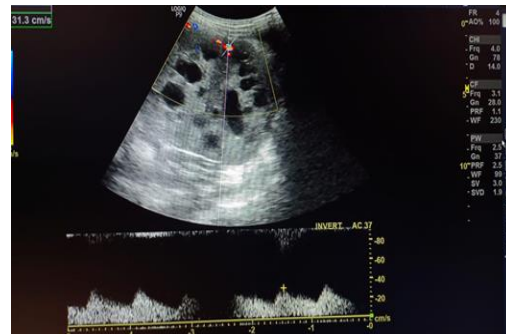


Image 3

On colour doppler we can colour uptake within the lesion giving pulsatile arterial waveform.

Further investigation was carried out in same hospital, CECT was the preferred modality to assess the mass. Considering close proximity to pancreatic body and tail. Pancreatic parenchymal phase, portal-venous phase and delayed was obtained.

### NCCT

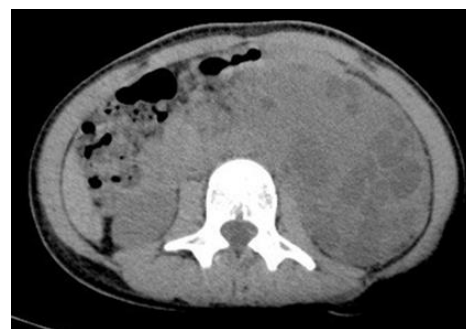


Figure 1: Axial sections of NCCT abdomen at level of right kidney showing heterodense mass in left lumbar quadrant

CECT

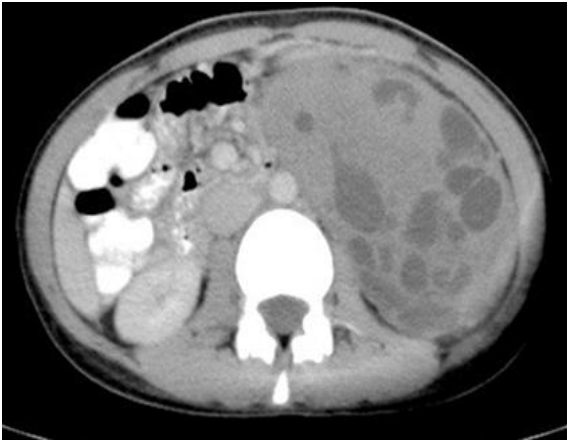


Figure 2: Axial sections of CECT abdomen in pancreatic parenchymal phase showing heterogeneously enhancing mass with non-enhancing hypodense area



CECT

Pelvis showing fluid within pelvic cavity around uterus and ovaries

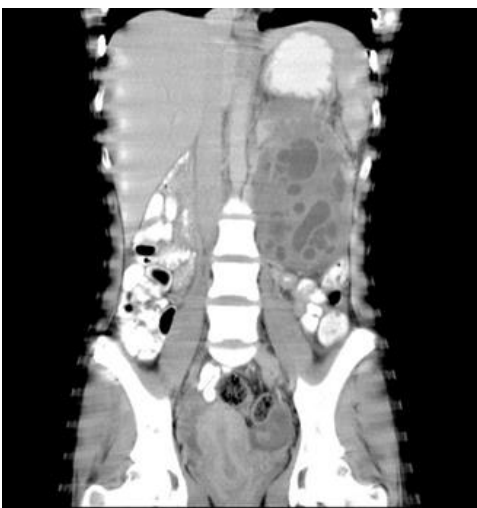


Figure 3:

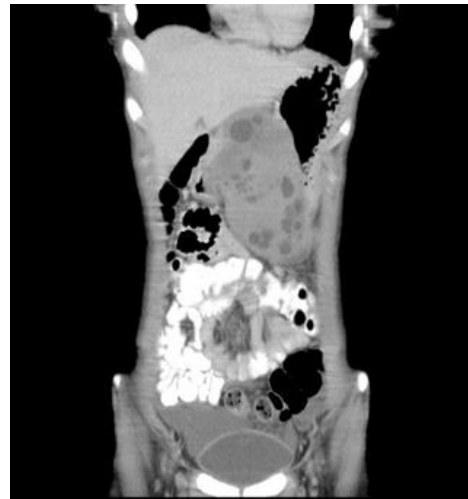


Figure 4:

A CECT was done a large well encapsulated heterodense retroperitoneal lesion measuring approximately 15 x 12 x 10 cm( CC x TR x AP) , on post contrast showing minimal heterogeneous enhancement of solid component and non enhancing cystic spaces of fluid.

Antero-superiorly: lesion is seen indenting greater curvature of stomach, gastro-duodenal junction and 1st part of duodenum.

Right laterally: it is seen displacing 2nd part of duodenum, celiac trunk and superior mesenteric artery causing kinking of splenic artery at its origin from celiac trunk.

Posterior and left laterally: Lesion is seen abutting abdominal aorta, splenic vessels, anterior surface of left kidney. Infrarenal portion of the lesion is seen extending till left paraspinal muscles and posterolateral abdominal wall.

Lesion is causing displacement of bowel loops anterolaterally.

### Histopathology Report

- Desmoid tumors are histologically qualified as benign and do not metastasize but they are often highly recurrent and locally invasive.
- 85% are FAP-associated (Familial adenomatous polyposis), and only 5% are sporadic.
- Repeated irritation or trauma to a certain part of the body, including surgical trauma, estrogen has been theorized to increase the risk of desmoid tumor occurrence. Desmoid tumors grow fast and depending on their localization can cause pain, nausea, vomiting, weight loss, obstruction or perforation of the small intestine, enterocutaneous fistula and intestinal bleeding.
- During gross histopathologic examinations desmoid tumors most often are seen as circumscribed lesions, but they also can have irregular borders. They are usually larger than 5 cm and can reach up to 15 cm. In 10 - 15% of cases they are multiple. Histologically desmoid tumors are lesions composed of bland spindled or stellate fibroblastic cells

embedded in a collagenous stroma, without evidence of muscular or neural differentiation and with little or no inflammatory component.

- The tumor may infiltrate adjacent viscera and tissues at the periphery. On a CT scan, most desmoid tumors appear as well-circumscribed homogeneous masses that may be iso-dense or hyper-dense relative to muscle. Some cases of heterogeneous masses with infiltrative outer margins are seen.
- Desmoid tumors may enhance after injection of IV contrast material, localizes the tumor and excludes metastasis.

### References

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