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Isolated pancreatic desmoid tumor, challenges in the diagnosis and management: a case report.

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Abstract: *Desmoid tumors are uncommon lesions involving the mesenchymal tissue. They commonly involve fascia, aponeurosis and periosteum. In the abdomen, they commonly involve mesentery and retroperitoneum. Isolated pancreatic desmoid is quite rare lesion. They can present as a solid or solid cystic mass. Here we report such a case in a 19 year old female who presented with pain abdomen and early satiety for a period of 2 months. On evaluation, we found a lesion arising from the body and tail of pancreas with combined solid and cystic areas. Considering the age, sex and imaging findings we planned for surgical resection suspecting solid pseudopapillary neoplasm of pancreas. However, intraoperative findings of stomach wall infiltration made us to perform wide local excision in the form of near total gastrectomy and distal pancreatosplenectomy. We present this case for the rarity of the lesion.*

Keywords- *Beta-catenin, Cystic neoplasm of pancreas, Familial adenomatous coli, Gardner syndrome, Solid pseudopapillary neoplasm.*

I. INTRODUCTION

The prevalence of the cystic lesion of pancreas varies from 2.2%-15.9% in various literatures [1]. Solid pseudopapillary epithelial neoplasm (SPEN) is a rare lesion of pancreas most commonly seen in females at their 30's, few of them present as early as 13years or as late as 75years [2]. These tumors present most commonly with pain abdomen. Desmoid tumor is an aggressive fibromatosis of mesenchymal cells incidence of which is around 0.03% of all tumors. They generally involve fascial and musculoaponeurotic tissues [3]. Isolated desmoid tumor of pancreas is uncommon. They present can as a solid mass, cystic or solid-cystic mass. Here we present a rare case of isolated desmoid tumor of pancreas which showed features of solid and cystic components on imaging mimicking SPEN.

II. CASE REPORT

A 19 year old female presented to us with history of dull aching left upper quadrant abdomen pain for duration of 2 months. She also developed early satiety, loss of weight, nausea over a period of 2 months. On examination, she was moderately built and nourished, hemodynamic stable. Mass lesion was palpable in the left

hypochondrium and umbilical region measuring 12*10cm, not moving with respiration. She was evaluated with ultrasound of abdomen which showed mass in the tail of pancreas. She underwent contrast enhanced computed tomography which showed 15.8*12.9 cm multiple cystic lesion replacing the body and tail of pancreas abutting posterior wall of stomach. MRI abdomen performed also showed similar findings. CT and MRI images are shown in Fig. 1. Endoscopic ultrasound was performed which showed homogenous hypoechoic solid and cystic lesion measuring 11*7cm in the body and tail of pancreas. Biopsy performed was inconclusive of the diagnosis. Tumor marker Carbohydrate antigen 19-9 was 5.7U/mL.

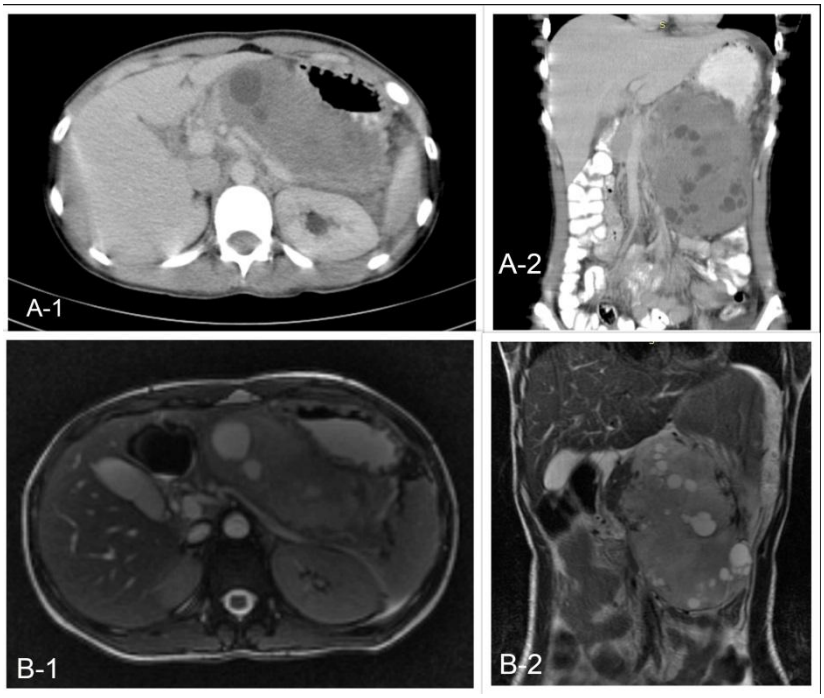

Figure 1: A-1, A-2 are CT images and B-1, B-2 are MRI images showing the solid lesion with cystic areas arising from body of pancreas abutting posterior stomach wall.

A working diagnosis of cystic neoplasm of pancreas favoring solid pseudopapillary neoplasm of pancreas was made and planned for distal pancreatosplenectomy. She underwent vaccination for Pneumococcal, Meningococcal and Hemophilus influenza. Diagnostic laparoscopy was performed to rule out metastatic disease followed by a laparotomy via a subcostal incision. Huge conglomerate of solid and cystic lesions arising from the body and tail of pancreas was noted to infiltrate the posterior wall of body of stomach, splenic vessels.

She underwent a distal pancreatosplenectomy, near total gastrectomy and Roux en Y gastrojejunostomy reconstruction with placement of Nasojejunal feeding tube. Fig. 2 is showing the picture of resected specimen. Fig. 3 is showing microscopy and immunohistochemistry picture. Post-operative period was uneventful. She was initiated on nasojejunal feeding by post-operative day2. Oral contrast study performed on day 10 to confirm the intactness of anastomosis following which she was started on oral diet. By the end of day 15, she was able to tolerate good oral diet and nasojejunal tube was removed.

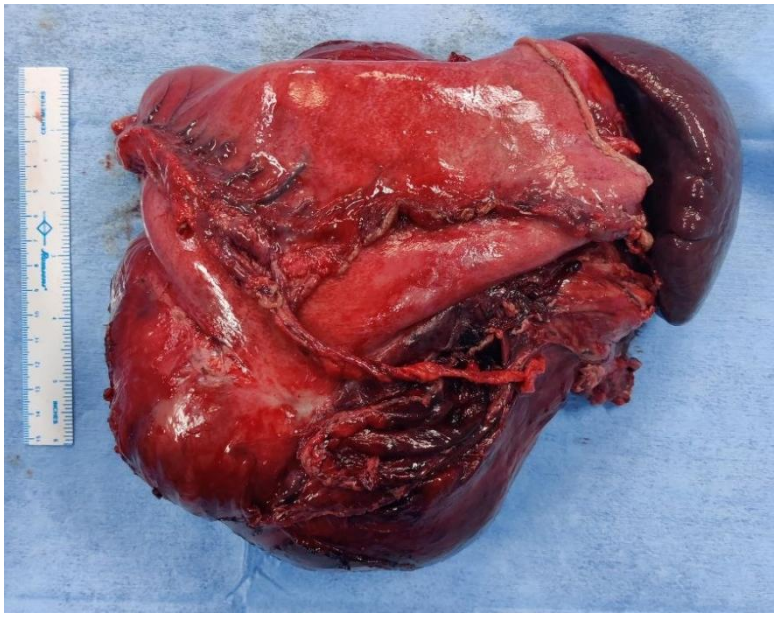

Figure 2: Picture of resected specimen showing the lesion with spleen and adherent stomach.

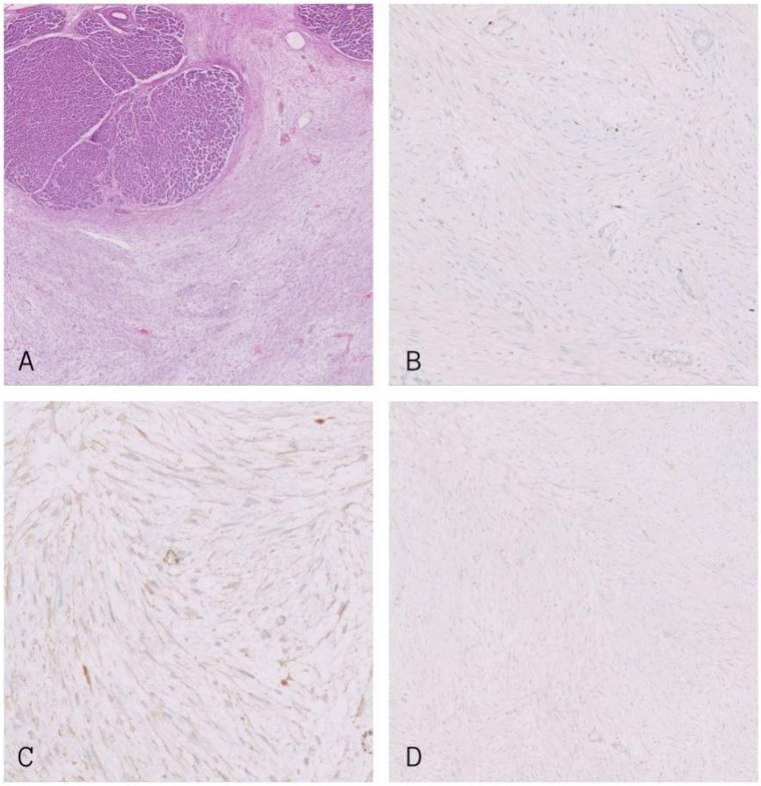

Figure 3: Microscopic and Immunohistochemistry images. A- Low power microscopy picture, B- Ki 67 staining, C- Beta-catenin staining, D- MUC4 staining.

The histopathological analysis of the lesion showed infiltrative lesion composed of spindle cells in elongated and focal whorls in a collagenous and myxoid stroma. Lesion is infiltrating the pancreatic parenchyma and stomach wall. Spleen is normal and free of infiltration. Immunohistochemistry performed showed nuclear positivity for Beta-catenin, Ki-67 3% and negative for MUC4, PgR, WT1 and CD 117.

Three months follow up following the surgery patient is doing fine, able to tolerate good oral diet and ultrasound imaging is suggestive of no recurrence. Currently she is on follow up every 3 months with the intent of performing cross sectional imaging once in 6 months or before if symptomatic.

III. DISCUSSION

Desmoid tumor is a rare tumor of mesenchymal cells. Incidence is around 0.01-0.03% of all tumors and 1-3% of all fibrous tumors [3, 4]. These tumors are locally invasive proliferation of well differentiated mesenchymal fibroblasts. They have no potential for distant metastasis [5]. These tumors primarily involve mesentery and retroperitoneal connective tissues and are often asymptomatic [3, 6]. Extra-abdominal location is not uncommon, mainly affecting skeletal muscle, fascia, aponeurosis and periosteum.

The most common presentation is in the form of pain abdomen, early satiety and weight loss. Lesions in the pancreas may present with epigastric pain radiating to back [4]. Radiologic appearance depends on the relative amounts of fibrosis, collagen and vascularity. They appear well or ill-defined masses with variable attenuation on CT, while on MRI they show low signal intensity on T1-weighted images and variable T2 signals[7, 8]. These tumors are made of spindle cells and immunohistochemistry shows positivity for vimentin and beta-catenin, negative for S100, CD117 and CD34 [9].

They are often found in patients with Familial adenomatous polyposis (FAP) or Gardner syndrome. The genes noted to be involved are adenomatous polyposis coli (APC) gene or beta-catenin gene [10]. Other risk factors noted are family history, surgery, oral contraceptive pills and trauma [5, 10].

The treatment of desmoid tumors are varies with the symptoms, site of the lesion and clinical status of the patients. Desmoid tumor working group has built a guideline based on the joint global consensus [11]. They have evaluated role of surgery vs observation, surgery plus radiotherapy vs surgery alone, radiotherapy alone vs radiotherapy plus surgery, radiotherapy vs surgery, medical therapy vs observation. Available medical therapies include Non-steroidal anti-inflammatory drugs (NSAIDs), anti-hormonal therapy, tyrosine kinase inhibitors and low dose chemotherapeutic agents [12, 13]. Desmoid tumor working group suggest wide local excision to achieve R0 margins should be preferred, R1 is acceptable when cosmosis is in question. However, if R1 resection is anticipated or if there is a high surgical morbidity, non-surgical options should be explored. There is no evidence in addition of radiotherapy or re-surgery in microscopic margin positive resection [11].

We have performed a wide local excision of the lesion with minimal morbidity. Even though it required resection of majority of stomach and spleen, there was not post-operative morbidity. We have vaccinated the patient against pneumococcal, meningococcal and Hemophilus influenza. She has been counseled regarding the need for genetic testing for FAP. In view of financial constraints, family has requested time for further evaluation. Presently she is performing well in academics and under regular follow up.

IV. CONCLUSION

Desmoid tumors are rare lesion to encounter and even rare is the isolated pancreatic desmoid. We present a lesion in the body and tail of pancreas with solid and cystic areas, which we initially thought to be a SPN based on the age, sex and imaging appearances. However, intraoperative picture of the lesion with infiltration of the stomach wall and final histopathological evaluation confirmed the lesion as desmoid tumor with free margins. Although various treatments for desmoid tumor are debatable, surgery with R0 resection is best possible way for cure. Syndrome association is to be evaluated in these patients.

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REFERENCES

[1.] Ip IK, Mortele KJ, Prevedello LM, Khorasani R. Focal cystic pancreatic lesions: assessing variation in radiologists' management recommendations. Radiology. 2011 Apr;259(1):136–41.

[2.] Reddy S, Cameron JL, Scudiere J, Hruban RH, Fishman EK, Ahuja N, et al. Surgical Management of Solid-Pseudopapillary Neoplasms of the Pancreas (Franz or Hamoudi Tumors): A Large Single-Institutional Series. J Am Coll Surg. 2009 May;208(5):950–9.

[3.] Sakorafas GH, Nissotakis C, Peros G. Abdominal desmoid tumors. Surg Oncol. 2007 Aug;16(2):131–42.

[4.] Torres JC, Xin C. An unusual finding in a desmoid-type fibromatosis of the pancreas: a case report and review of the literature. J Med Case Reports. 2018 May 12;12(1):123.

[5.] Gerleman R, Mortensen MB, Detlefsen S. Desmoid Tumor of the Pancreas: Case Report and Review of a Rare Entity. Int J Surg Pathol. 2015 Oct;23(7):579–84.

[6.] Kasper B, Ströbel P, Hohenberger P. Desmoid tumors: clinical features and treatment options for advanced disease. The Oncologist. 2011;16(5):682–93.

[7.] Amiot A, Dokmak S, Sauvanet A, Vilgrain V, Bringuier PP, Scoazec JY, et al. Sporadic desmoid tumor. An exceptional cause of cystic pancreatic lesion. JOP J Pancreas. 2008 May 8;9(3):339–45.

[8.] Rosa F, Martinetti C, Piscopo F, Buccicardi D, Schettini D, Neumaier CE, et al. Multimodality imaging features of desmoid tumors: a head-to-toe spectrum. Insights Imaging. 2020 Sep 25;11:103.

[9.] Jia C, Tian B, Dai C, Wang X, Bu X, Xu F. Idiopathic desmoid-type fibromatosis of the pancreatic head: case report and literature review. World J Surg Oncol. 2014 Apr 22;12(1):103.

[10.] Leal RF, Silva PVVT, Ayrizono M de LS, Fagundes JJ, Amstalden EMI, Coy CSR. Desmoid tumor in patients with familial adenomatous polyposis. Arq Gastroenterol. 2010;47(4):373–8.

[11.] Alman B, Attia S, Baumgarten C, Benson C, Blay JY, Bonvalot S, et al. The management of desmoid tumours: A joint global consensus-based guideline approach for adult and paediatric patients. Eur J Cancer. 2020 Mar 1;127:96–107.

[12.] Janinis J, Patriki M, Vini L, Aravantinos G, Whelan JS. The pharmacological treatment of aggressive fibromatosis: a systematic review. Ann Oncol Off J Eur Soc Med Oncol. 2003 Feb;14(2):181–90.

[13.] Al-Jazrawe M, Au M, Alman B. Optimal therapy for desmoid tumors: current options and challenges for the future. Expert Rev Anticancer Ther. 2015;15(12):1443–58.