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Cystic Lymphangioma of Pancreas - Case Series

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Highlights

- Cystic lymphangiomaof Pancreas is a rare entity.
- Eventhough imaging shows large size , mass wont be palpable because of yielding nature.
- Importance of this entity lies in the differential diagnosis from other cystic lesions of pancreas based on age at presentation, clinical & radiological characteristics.

I. Introduction

Cysticlymphangioma of pancreas is a very rare entity[1]. In 1913, Koch reportedthe first case of cystic lymphangioma of pancreas[1].Lymphangioma usually occurs in the neck, axillary region, and rarely in the mediastinum. Abdominal organs are uncommon sites of origin for lymphangioma[1]. Of all the lymphangiomas in the peritoneal cavity, about 70% have been found in the mesentery of small intestine[2]. Pancreas is one of the rarest site& it is to be considered as one of the differential diagnosis among cystic neoplasms of pancreas, pseudocyst of pancreas & also other retroperitoneal cystic tumours.

II. Case Series –Study

Herein, We report 6 cases of cystic pancreatic lymphangioma from march 2013 to june 2016 . All patients presented in 2nd decade as vague epigastric discomfort.Male : female ratio is 1:1; more commonly located in the region of body & tail of the pancreas (body & tail - 4 ; head – 2) occupying lesser sac & upper retroperitoneum. Upon physicalexamination, there was no palpable mass because of yielding nature though lesion was large.Size of the lesion vary from 6cm to 19cm (average- 12cm) with Hepatic artery &GDA seen coursing through the lesion in one case.

Radiological Imaging studies commonly revealed multiseptated cystic mass lesion located in the upper ventral pancreatic area (head – 2; body&tail -4)neighboring posterior stomach occupying upper retroperitoneum; Size of the lesion vary from 6cm to 19cm (average- 12cm) with Hepatic artery & GDA seen coursing through the lesion in one case.EUS guided cyst fluid aspiration done & level of CEA,Amylase,
CA 19-9 are found to be within normal limits.Complete excision preserving vessels was done in all the cases.Final pathological diagnosis was reported as cystic lymphangioma.All the patients made complete recovery & no recurrences on followup.

Table

	Case-1	Case-2	Case-3	Case-4	Case-5	Case-6
Age	16	12	20	13	19	16
Sex	Female	Male	Male	Female	Male	Female
Location	Head	Head	Body & tail	Body &tail	Body & tail	Body & tail
Extent	Lesser sac &HDL ligament	Lesser sac	Lesser sac Upper retroperitoneum	Lesser sac Upper retroperitoneum	Lesser sac Upper retroperitoneum	Lesser sac Upper retroperitoneum
Complication	Infection	-	-	-	-	-

Figure -1
CECT abdomen showing multiseptatedhypodense cystic lesion with nonenhancing thin delicate septa occupying the body & tail of the pancreas in the lesser sac.

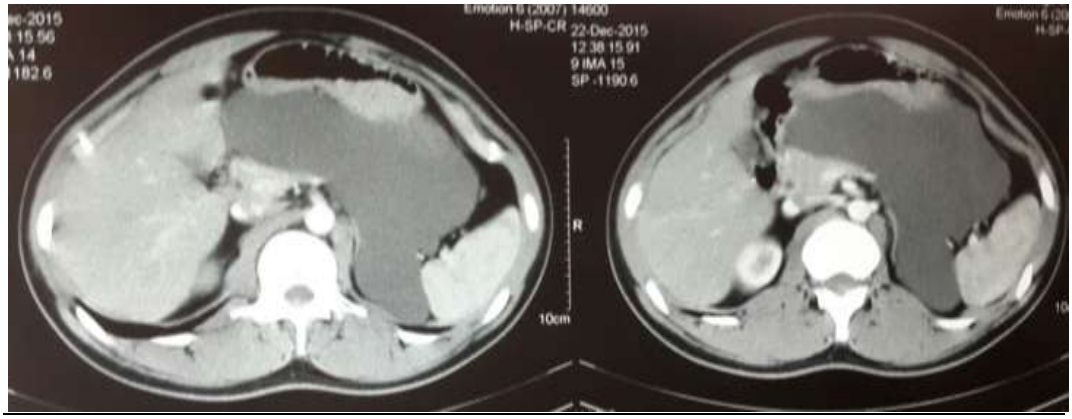

Figure-2
Intraoperative picture showing cystic lymphangioma occupying the body & tail of the pancreas in the lesser sac posterior to stomach.

III. Discussion

Cystic lymphangiomas are multi-loculated soft cystic masses, composed of dilated lymphatic channels, divided by thin septae because of insufficient drainage due to abdominal trauma, inflammatory process, surgery or radiation therapy[3].Pancreatic lymphangioma is a very rare entity less than 1%.In our study lesion more commonly located in body & tail of the pancreas (body & tail – 4; head – 2); male : female ratio-1:1; Age group – 2nd decade; Size of the lesion varying from 6cm to 19cm (average- 12cm) ; occupying upper retroperitoneum; not invading any vascular structures though imaging suggested hepatic artery & GDA coursing through the lesion in one case. Infection as a complication was found in one case althoughintracystic hemorrhage, cyst rupture, volvulus reported in literature[4]. Association of these lesions with blue rubber bleb nevus syndrome has also been described in literature.[5]

Importance of this entity is that this has to be differentiated from Pseudocysts, cystic neoplasm of pancreas (mucinous and serouscystadenomas, solid pseudopapillarytumors,IPMN),congenital cysts and ductal carcinoma& other retroperitoneal cystic tumours[6] .Imaging studies (CT) helped us to differentiate these lesions from others. These lesions usuallypresented asmultiloculated cystic tissue lesion with anechoic contents with nonenhancing thin delicate septa crisscrossing the lesion[7]; No true capsule; No mural nodules/ wall calcification or perilesional lymphadenopathy .Complete excision is the treatment of choice ; recurrence can occur in 7% of the cases[8] though no recurrences on follow up. Although lymphangioma rarely becomes malignant, its prognosis is generally good[9]

IV. Conclusion

Cystic pancreatic lymphangiomas are rarely seen tumors. Importance of this entity lies in the differential diagnosis from other cystic lesions of pancreas based on age at presentation,clinical & radiological characteristics.Complete surgical resection is curative.

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