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Case Report

Epidermoid Cyst arising within an Intrapancreatic Accessory Spleen [ECIPAS] mimicking a pancreatic mucinous cystic neoplasm-a case report with literature review

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Abstract

Background: Accessory spleens are congenital embryological aberrations usually found within the splenic hilum with no clinical significance. An Intra Pancreatic Accessory Spleen (IPAS) is an uncommon benign pancreatic lesion encountered clinically. The occurrence of an Epidermoid Cyst arising within an IPAS (ECIPAS) is exceedingly rare and is often misdiagnosed as a pancreatic pathology such as mucinous cystic neoplasm, cystic degeneration within a solid tumor such as a neuroendocrine tumor, or a lymph node.

Case report: A 68-year-old male presented with intermittent post-prandial abdominal pain for over 2 years. Abdominal computer tomography identified a 5.2 cm calcified cyst within the pancreatic tail and a mucinous pancreatic neoplasm/pancreatic pseudocyst was considered in the differential diagnosis. The results of endoscopic ultrasound and fine needle aspiration were indeterminate. Due to persistent abdominal pain, the patient underwent a laparoscopic distal pancreatectomy with splenectomy. A gross examination of the distal pancreatic/splenectomy specimen confirmed a well-demarcated cystic lesion with brownish fluid within the pancreatic tail. Microscopic examination revealed a nonpathological pancreas separated by a fibrous capsule with a large cyst arising within an intrapancreatic accessory spleen. The cyst was lined with multilayered non-keratinized stratified squamous epithelium positive for pancytokeratin, CA 19-9, CK5/6, and p63 with no lymphocytic infiltrates and absent hair/ dermal appendages confirming an epidermoid cyst. CD8 outlined the dendritic network of the littoral cells of the splenic tissue in the cyst wall. Post-operative follow-up at 6 weeks was uneventful.

Conclusion: The preoperative diagnosis of ECIPAS is extremely difficult as this entity shares overlapping radiological features with other cystic lesions such as mucinous pancreatic cysts. It is important for anatomic pathologists to recognize and consider the intrapancreatic compartment as a possible site for accessory spleen. As histopathology is the final determinant of this diagnosis, increased clinical awareness with an accurate diagnosis of this entity may prevent patients from unnecessary surveillance and/or extensive oncological-based surgical resection.

Introduction

Accessory spleens are incidentally found at autopsy in 12.1% of cases [1]. Approximately, 9.3 - 22% of these accessory spleens were identified within the pancreatic tail [1-3]. Epidermoid cyst within an intrapancreatic accessory spleen (ECIPAS) is a rare cystic neoplasm first described by Davidson, et al. in 1980 [4]. ECIPAS predominantly behaves as a benign lesion and to date, there is only one documented case of ECIPAS transformation to squamous cell carcinoma [5].

Radiological diagnosis of this entity is extremely challenging as it is difficult to differentiate ECIPAS from other 'pancreatic cystic neoplasms' such as Mucinous Cystic Neoplasm (MCN), cystic degeneration within a Neuroendocrine Tumour (NET) and Solid Pseudopapillary Tumour (SPT) [6,7]. Due to the malignant neoplastic potential of these entities, most of the patients with ECIPAS undergo surgical resection. Contrastingly, if a cystic lesion is identified within an accessory spleen preoperatively then the differential diagnosis is similar to that of splenic cysts as outlined in Figure 1.

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In this study, we report the case of a 68-year-old with ECIPAS who underwent distal pancreatectomy and splenectomy due to suspicion of MCN. A comprehensive literature review is also performed to summarize the clinical and pathological insights into cases with ECIPAS. The ancillary objective of our case report is to highlight this entity for a surgical pathologist to consider within their differential diagnosis of cystic lesions within the pancreas and to encourage clinical awareness for exploration of surgical alternatives such as cyst excision with splenic preservation in lieu of distal pancreatectomy with total splenectomy [7].

Case presentation/report

A 68-year-old otherwise healthy male presented with dull intermittent post-prandial epigastric pain radiating to the left flank for over 2 years. His physical examination was unremarkable. Biochemical tests were within normal limits except for elevated ferritin. Serum carbohydrate antigen 19-9 (CA 19-9) and Carcinoembryonic Antigen (CEA) tests were not performed. His routine laboratory data was within normal limits. Abdominal Computed Tomography (CT) showed a 5.4 x 4.9 cm cystic mass with partial calcification within the pancreatic tail (Figure 2). The radiology-based differential diagnosis included mucinous pancreatic neoplasm or pancreatic pseudocyst.

Fine needle aspiration via endoscopic ultrasound was attempted with indeterminate results. The patient underwent a laparoscopic distal pancreatectomy and splenectomy for query mucinous neoplasm of the pancreas. On gross examination, a cystic, partially calcified well-demarcated mass measuring 5.7 x 5.3 x 5.2 cm containing red-brown fluid was identified within the tail of the pancreas separate from the native spleen as seen in Figures 3A-C. Additional three firm well-defined nodules measuring 0.3 cm, 0.9 cm and 0.4 cm were identified abutting the splenic capsule and around the cyst as highlighted by arrows in Figure 3C.

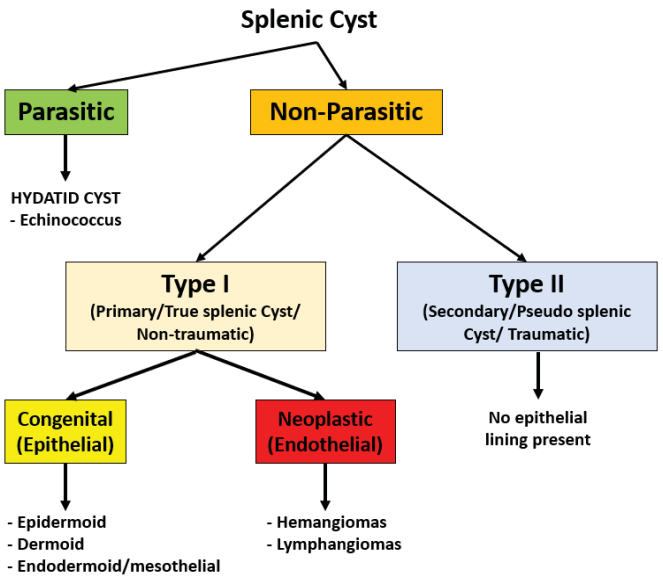

Figure 1: Schematic flow chart summarizing the stepwise differential diagnosis approach for a splenic cyst.

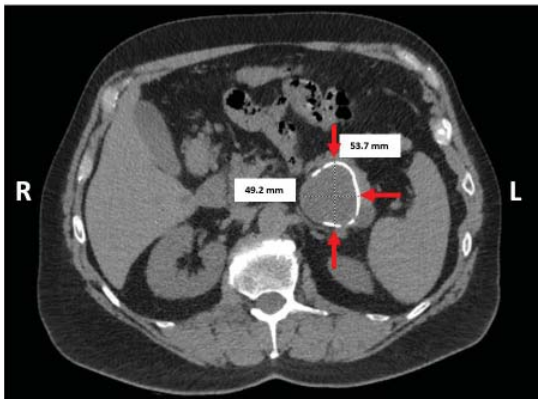

Figure 2: Abdominal Computer tomography showing a cystic mass (red arrows) with partial calcification within the pancreatic tail.

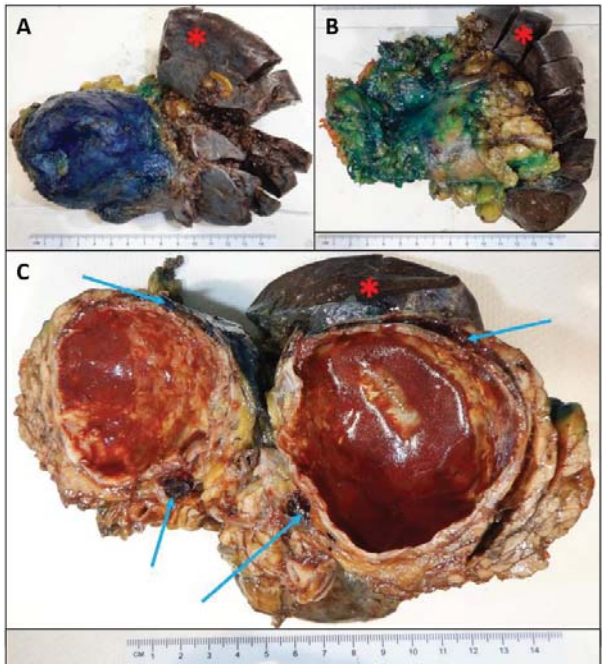

Figure 3: Distal pancreatomy and splenectomy specimen showing the relationship between the cyst mass and native spleen (red asterisks). [A]: Anterior surface of the cystic mass (inked blue). [B]: Distal pancreatic tail margin (inked orange) and posterior surface of the cystic mass (inked green). [C]: Cystic mass bisected into anteroposterior leaflets. The cyst shows a white wall lining with red-brown soft fluid. Cystic mass with peripheral intrapancreatic accessory splenic tissue (blue arrows).

Microscopic examination revealed an unremarkable pancreas with the presence of splenic tissue [*] inside the pancreas [#] separated by a thick fibrous capsule [+] confirming the presence of an intrapancreatic accessory spleen containing a cyst with a thick fibrous wall with foci of dystrophic calcifications (Figure 4A). The cyst was lined by non-keratinized Stratified Squamous Epithelium (SSE) (Figure 4B). In other areas, the cyst lining was denuded to a cuboidal monolayer with subepithelial fibrosis, and underlying accessory splenic tissue [*] was identified within the cyst wall (Figure 4B,C). No ovarian stroma was identified. The cyst wall showed no evidence of lymphocytic infiltrates or dermal appendages. There was evidence of cyst rupture with chronic inflammation and cholesterol clefts [^] (Figure 4D). No hair was present within the cyst. The nodules identified on gross

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were three accessory spleens. The native spleen had large coalescing granulomas (Figure 4E) containing multinucleated giant cells (Figure 4F). Acid-fast bacilli (AFB) stain, periodic acid-Schiff (PAS), and Grocott's methenamine silver (GMS) special stains showed no evidence of acid-fast bacilli or fungal microorganisms.

The Immunohistochemistry (IHC) stains for CD8 confirmed the classic dendritic pattern of staining of the littoral cells within the accessory splenic tissue (Figure 5A). The cyst SSE lining showed block positivity for pancytokeratin and CK5/6 (Figure 5B). P63 was also expressed with a preferential gradient pattern of stronger basal to suprabasal (Figure 5C). The superficial layers of the SSE lining showed positivity for CA 19-9 and MUC4 (Figure 5D and 5E). D2-40 expression was limited to the basal layer of cells with no expression in superficial layers (Figure 5F). There was no expression of Calretinin and WT1. The overall histology and IHC immunophenotype confirmed the presence of an Epidermoid Cyst arising within an Intrapancreatic Accessory Spleen [ECIPAS]. The patient remained asymptomatic, and his post-operative follow-up was unremarkable.

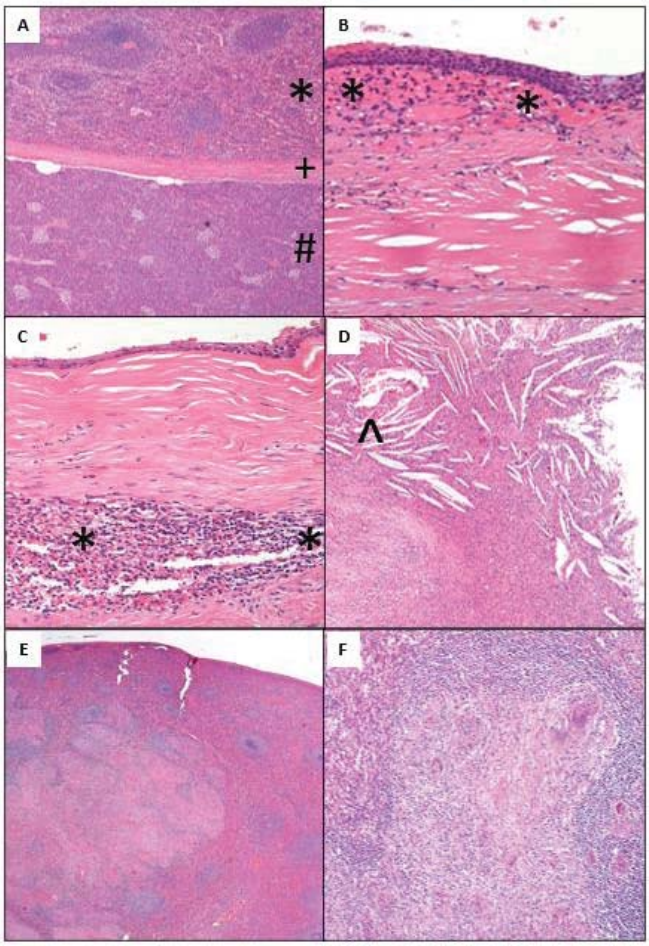

Figure 4: Characteristic microscopic findings of ECIPAS on hematoxylin and eosin. A: Hyalinized fibrous capsule [+] separates non-pathogenic pancreas [#] from accessory spleen [*]. B: Cyst lined by stratified squamous epithelium with splenic tissue in the wall [*]. C: Cyst lined by denuded cuboidal monolayer with subepithelial fibrosis and underlying splenic tissue [*]. D: Cyst rupture with Cholesterol clefts [^]. Native spleen specimen with coalescing granulomas [E]: with multinucleated giant cells [F].

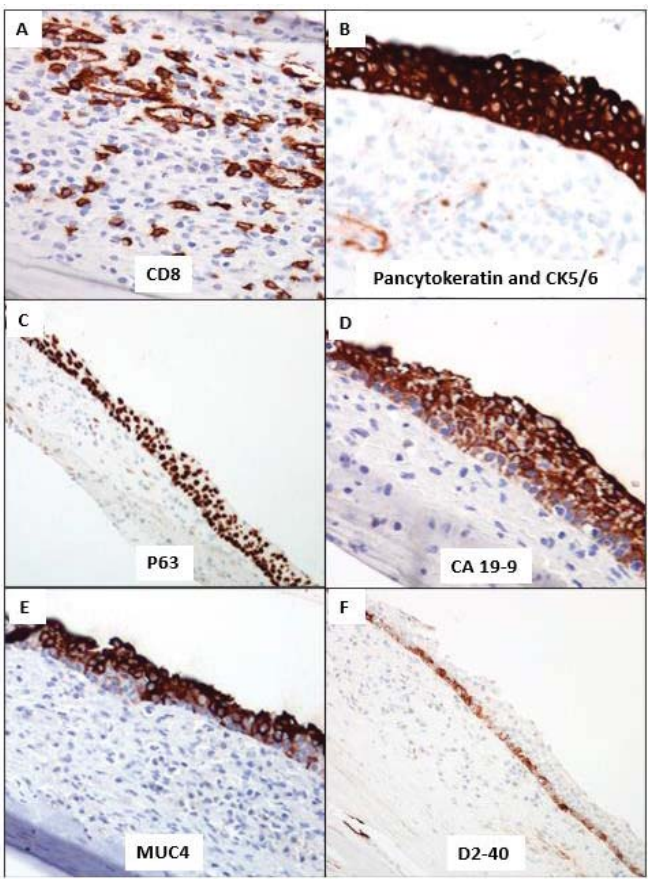

Figure 5: IHC results for ECIPAS. [A]: CD8 positive littoral cells showing classic dendritic pattern of staining. [B]: Cyst lined by Pancytokeratin and CK5/6 positive stratified squamous epithelium (SSE). [C] P63 shows a strong basal to weaker suprabasal gradient staining pattern. Stratified superficial layers express CA 19-9 [D]: and MUC4 [E]: predominantly in the suprabasal cell layers, while D2-40 staining [F]: is restricted to the basal cells.

Discussion

ECIPAS is a rare entity and has been reported in 59 patients within the existing English literature (Table 1). It is also notable these 49 publications analyzed in our literature review were from Japan (25), United States (9), South Korea (5), China (4), United Kingdom (1), Australia (1), Greece (1), Netherlands (1), and Singapore (2). To the best of our knowledge, this is the first case report from Canada.

ECIPAS was commonly identified in the 47 ± 13 years-old population [Range: 12-70] and shows a female gender predilection (F:M = 1.4: 1). These patients were either asymptomatic with incidental cystic lesions on their radiological imaging (55.9%, *n* = 33 out of 59) or presented with abdominal pain (38.9%, *n* = 23 out of 59). In cases with localized abdominal pain (*n* = 15), left-sided (42.8%, *n* = 6 out of 14) or epigastric (35.7%, *n* = 5 out of 14) pain was common. Abdominal ultrasound and computer tomography (CT) were first-line radiological studies ordered in these patients. Notable findings included cystic lesions within the pancreas (86.4%, *n* = 51 out of 59) with rim enhancement, calcifications, and no anatomic continuity with the spleen (27.2%, *n* = 12 out of 44). The majority of the ECIPAS were located within the pancreatic tail (98.3%, *n* = 58 out of 59). As radiological findings lack

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Table 1: Summary of clinical variables including Clinical presentation, preoperative diagnosis, Serum CA19.9, Serum CEA, management and their reference number as reported in 45 cases of ECIPAS in the English Literature. These cases are listed chronologically based on their date of publication.

Age (M/F)	Clinical Presentation	Preoperative diagnosis	Serum CA 19-9 (U/mL)	Serum CEA	Mx	Date	Ref.
40 (M)	WL, n/v, CP (24 months)	Pancreatic pseudocyst, cystadenocarcinoma	NA	NA	DPS	1980	[4]
51 (M)	Abdominal pain (RLQ)	Pancreatic pseudocyst	NA	NA	DPS	1981	[8]
32 (F)	Abdominal pain (LUQ)	Benign Pancreatic cyst	Normal	Normal	Cys	1991	[9]
37 (F)	Abdominal pain (Epigastric)	Malignant Pancreatic Cystic neoplasm	NA	NA	DP	1991	[10]
38 (M)	ASx, incidental imaging	NA	NA	NA	DPS	1994	[11]
45 (M)	ASx, incidental imaging	Malignant Pancreatic Cystic neoplasm	NA	NA	DP	1998	[12]
46 (F)	Abdominal pain (LF)	Malignant Pancreatic Cystic neoplasm	Elevated (201)	NA	DPS	1998	[13]
67 (F)	Abdominal pain	Pancreatic cyst	Elevated	Elevated	DPS	1998	[14]
49 (F)	ASx, incidental imaging	Pancreatic cyst	NA	NA	DPS	1999	[15]
51 (M)	ASx, incidental imaging	Benign Pancreatic cyst	Normal	Normal	DPS	2000	[16]
48 (M)	ASx, incidental imaging	MCN	Elevated	NA	DPS	2001	[17]
45 (F)	Abdominal pain (Epigastric)	Malignant Pancreatic Cystic neoplasm	Elevated (159)	NA	DPS	2002	[18]
12 (F)	Fever	Infected pancreatic/ enteric duplication cysts	NA	NA	DP	2002	[19]
38 (M)	ASx, incidental imaging	MCN, cystadenocarcinoma	Elevated (410)	NA	DPS	2002	[20]
58 (F)	ASx, incidental imaging	MCN	Elevated (62)	NA	DP	2004	[21]
55 (F)	Abdominal pain (Epigastric)	MCN, cystadenocarcinoma	Elevated (176)	Normal	DPS	2004	[22]
32 (M)	Abdominal pain (LUQ)	Pancreatic pseudocyst	Elevated (53)	NA	DP	2005	[23]
49 (F)	Abdominal pain	MCN	Normal	Normal	DPS	2005	[23]
41 (M)	ASx, incidental imaging	Malignant Pancreatic Cystic neoplasm	NA	NA	DPS	2007	[24]
40 (M)	ASx, incidental imaging	ECIPAS	Normal	Normal	DPS	2008	[25]
52 (F)	ASx, incidental imaging	Malignant Pancreatic Cystic neoplasm	Elevated	Elevated	DPS	2008	[26]
32 (F)	Abdominal pain (RUQ)	Malignant Pancreatic Cystic neoplasm	NA	Elevated	DPS	2008	[27]
26 (F)	ASx, incidental imaging	MCN	Normal	Normal	DP	2009	[28]
49 (M)	ASx, incidental imaging	MCN	Elevated	Elevated	DPS	2009	[29]
54 (F)	Abdominal pain (LUQ), WL	NA	NA	NA	DPS	2000	[30]
57 (F)	ASx, incidental imaging	Malignant Pancreatic Cystic neoplasm	Elevated (1880)	Normal	DPS	2010	[31]
70 (F)	ASx, incidental imaging	MCN	Elevated (48)	Normal	DPS	2010	[31]
37 (M)	ASx, incidental imaging	Pancreatic cystic lesion. Serous cyst, lymphoepithelial cyst	Elevated (647)	Normal	DP	2010	[31]
67 (M)	Abdominal pain (Epigastric), WL	MCN. cystadenocarcinoma	Elevated (182)	Elevated	DPS	2010	[32]
36 (F)	ASx, incidental imaging	MCN	Elevated (79)	NA	DPS	2011	[33]
55 (F)	ASx, incidental imaging	MCN	Elevated (90)	Normal	DPS	2011	[6]
50 (F)	ASx, incidental imaging	Malignant Pancreatic Cystic neoplasm	NA	NA	DP	2011	[34]
49 (F)	Abdominal pain	Pancreatic NET	NA	NA	DP	2011	[35]
62 (M)	Abdominal pain (LUQ)	NA	NA	NA	DP	2011	[36]
39 (M)	ASx, incidental imaging	Malignant Pancreatic Cystic neoplasm	NA	Normal	DPS	2012	[37]
54 (F)	Abdominal pain	Malignant Pancreatic Cystic neoplasm	NA	NA	DP	2013	[38]
41 (F)	ASx, incidental imaging	Cystic pancreatic lesions	Elevated (259.7)	Normal	DPS	2013	[39]
63 (F)	Abdominal pain, n/v	Pancreatic cystic tumour	Elevated (222)	NA	DPS	2014	[40]
50 (F)	ASx, incidental imaging	MCN	Elevated (43.1)	NA	DP	2016	[41]
60 (F)	Abdominal pain (LF)	IPMN	Elevated (52.9)	NA	DP	2016	[41]
33 (F)	ASx, incidental imaging	SPN, NET with cystic degeneration	Normal	Normal	DP	2016	[42]
21 (F)	Abdominal pain (RUQ)	MCN, SPN, NET with cystic degeneration	NA	NA	DP	2016	[7]
17 (F)	Abdominal pain, n/v	MCN, IPMN, SPN, NET	NA	NA	DPS	2018	[43]
33 (M)	ASx, incidental imaging	MCN	Elevated (50)	Normal	DPS	2019	[5]
56 (M)	Abdominal pain (Epigastric)	MCN	NA	NA	DPS	2019	[44]
51 (F)	Asx, incidental imaging	Pancreatic malignant neoplasm	NA	NA	DP	2019	[44]

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59 (F)	ASx, incidental imaging	MCN	Normal	Normal	DPS	2020	[45]
68 (M)	Abdominal pain	Colloid carcinoma, acinar cell carcinoma, SPN	Normal	Normal	PPPD	2020	[46]
42 (F)	Abdominal pain (Epigastric)	Pancreatic malignant neoplasm	Normal	Normal	DPS	2020	[47]
51 (M)	Abdominal pain (6 months)	Malignant Pancreatic Cystic neoplasm	Elevated (55)	Normal	DP	2021	[48]
34 (M)	ASx, incidental imaging	IPMN, PDAdc, NET	Normal	Normal	DPS	2022	[49]
57 (M)	ASx, incidental imaging	IPMN, PDAdc, NET	Normal	Normal	DP	2023	[50]
63 (F)	ASx, incidental imaging	IPMN, PDAdc, NET	Normal	Normal	DP	2023	[50]
55 (M)	ASx, incidental imaging	IPMN, PDAdc, NET	Normal	Normal	DP	2023	[50]
67 (F)	Abdominal pain (LF)	IPMN, PDAdc, NET	Elevated (52)	Normal	DP	2023	[50]
41 (F)	ASx, incidental imaging	IPMN, PDAdc, NET	Normal	Normal	DP	2023	[50]
64 (M)	ASx, incidental imaging	IPMN, PDAdc, NET	Normal	Normal	DP	2023	[50]
25 (M)	ASx, incidental imaging	SPN, MCN, IPAS with lymphatic cyst	Elevated (43.5)	Normal	DP	2024	[51]
28 (F)	ASx, incidental imaging	IPAS, ECIPAS	NA	NA	No surgery	2024	[52]

Abbreviations: M: Male; F: Female; CA 19-9: Carbohydrate Antigen 19-9; CEA: Carcinoembryonic Antigen; Mx: Management; Ref.: Reference; WL: Weight Loss; n/v: Nausea and Vomiting; CP: Chest Pain; ASx: Asymptomatic; NA: Not Available; RUQ: Right Upper Quadrant; RLQ: Right Lower Quadrant; LUQ: Left Upper Quadrant; LF: Left Flank; DP: Distal Pancreatectomy; DPS: Distal Pancreatectomy and Splenectomy; Cys: Cystectomy; PPPD: Pylorus Preserving Pancreaticoduodenectomy; MCN: Mucinous Cystic Neoplasm; PNET: Pancreatic Neuroendocrine Tumor; SPT: Solid Pseudopapillary Tumor; IPMN: Intraductal Papillary Mucinous Neoplasm; PDAdc: Pancreatic Ductal Adenocarcinoma; ECIPAS: Epidermoid Cyst Intrapancreatic Accessory Spleen; IPAS: Intrapancreatic Accessory Spleen **Reference range** [CA 19-9: 0-27 U/mL, CEA: <5 ng/mL]

specificity, the preoperative diagnosis is varied as seen in Table 1 with pancreatic malignant cystic neoplasms (74.6%, *n* = 44 out of 59) and mucinous cystic neoplasm (42.8%, *n* = 18 out of 42) being the predominant preoperative diagnoses.

The majority of these ECIPAS patients underwent extensive surgical resection in the form of distal pancreatectomy with splenectomy (54.2%, *n* = 32 out of 59) or distal pancreatectomy (38.9%, *n* = 23 out of 59). Two patients had ECIPAS treated by robot-assisted spleen-preserving distal pancreatectomy while another ECIPAS within the head of the pancreas underwent a pylorus-preserving pancreaticoduodenectomy (PPPD) [7,44,46]. Our patient underwent oncological-based surgery of distal pancreatectomy with splenectomy since MCN was the preoperative diagnosis. A possible suggestion to circumvent proceeding to these radical invasive surgical procedures is to obtain an intraoperative frozen section: biopsy the cyst wall to obtain a tissue diagnosis of ECIPAS to guide further management that may result in non-oncological resections with splenic preservation. However, parameters such as cyst size, location, and clinical presentation with symptoms will need to be considered together with an acknowledgement of resection acting as a preventative intervention against future cyst rupture, hemorrhage, infection, or recurrence for developing management plans.

In the last 5 years, there has been an increasing utilization of endoscopic ultrasound (EUS) procedures multiplexed with contrast enhancement or sulfur hexafluoride microbubbles to assist with the delineation of the margin of this pancreatic tail cystic lesion from the spleen. This preoperative assessment for clear margin delineation resulted in spleen-preserving distal pancreatomy in nine patients [44,48,50,51]. Interestingly, one patient underwent non-operative management based on the radiological findings in EUS and superparamagnetic iron oxide imaging (SPIO) which highlighted the subjacent parenchyma of the cyst showed signal hyperintensity similar to the spleen [52]. Utilizing these novel contrast-enhanced EUS and adequate fine needle sampling of the cystic lining that confirmed benign squamous cells, and CD8 positive splenic microvessel clusters with numerous lymphocytes in two cases did yield a preoperative diagnosis of ECIPAS [47,51,52].

The ECIPAS gross specimens resected in the literature review were distributed between multilocular (46.5%, *n* = 27 out of 59) and unilocular (47.4%, *n* = 28 out of 59) cysts with white to brown serous fluid contents, and a median largest cyst diameter of 3.0 cm [Range: 1-15 cm]. The histological findings in these cases showed a characteristic triad of non-neoplastic pancreatic tissue with hyalinized fibrous capsule and accessory splenic tissue identified by histology and the dendritic staining pattern of CD8 highlighting the littoral cells of the spleen. The cysts were lined with stratified squamous or cuboidal monolayer cells, and these cyst-lining cells were positive for CA 19-9 and cytokeratins (CK7, CK19, CK5/6, AE1/AE3). The majority of the cysts were lined by non-keratinizing squamous cells apart from 4 cases with keratinizing squamous cells [13,14,30,37]. However, it is to be noted that the cyst lining can be denuded and therefore the above triad cannot be mandated for accurate diagnosis of ECIPAS [53]. Many cases also report histological evidence of cyst rupture such as hematoma, cholesterol clefts, and giant cells [4,11,36,42,50]. As the cyst wall lining and contents are key determinants for cyst classification, the absence of hair and skin appendages in the cystic lesion and no lymphocytic infiltrates are the key histopathological features that differentiate an epidermoid cyst from a dermoid cyst and lymphoepithelial cyst respectively.

The Immunohistochemistry (IHC) profile for ECIPAS was reported in 33 cases in the literature. A summary of the pattern of staining in the ECIPAS lining for each reported IHC for patients in our literature review is shown in Figure 6. Overall, stratified squamous epithelium (SSE) of the ECIPAS stained positive predominantly with CA 19-9 (*n* = 8), CEA (*n* = 7), and CK7 (*n* = 6). The suprabasal SSE was notably positive for CA 19-9 (*n* = 7), CK7 (*n* = 6), CK 20 (*n* = 6) and MUC4 (*n* =

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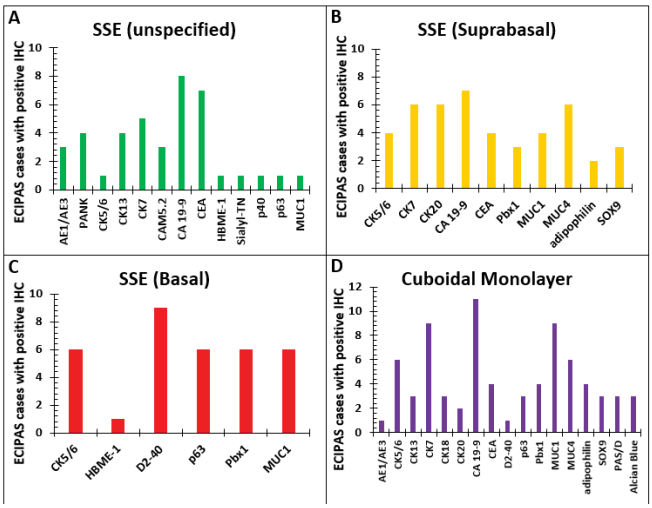

Figure 6: Summary of the pattern of staining and positivity for each reported IHC in ECIPAS patients in our literature review. [A]: Cases with IHC positivity with unspecified distribution within the stratified squamous epithelium (SSE) are shown in green solid bars. [B]: Cases with IHC positivity in the suprabasal SSE are shown in yellow solid bars. [C]: Cases with IHC positivity in the basal layer of the SSE are shown in red solid bars. [D]: Cases with IHC positivity in the cuboidal monolayer are shown in purple solid bars.

6). Contrastingly, basal SSE was commonly positive for D2-40 (n = 9), CK5/6 (n = 6), p63 (n = 6), Pbx1 (n = 6) and MUC1 (n = 6). Lastly, the cuboidal monolayer was predominantly positive for CA 19-9 (n = 11), CK7 (n = 9), CK5/6 (n = 6), MUC1 (n = 6), and MUC4 (n = 6). ECIPAS cases were predominantly negative for Calretinin (n = 7), WT-1 (n = 6), and vimentin (n = 4). We utilized Pancytokeratin, CK5/6, CK19, CK7, CA 19-9, MUC4, p63, Calretinin, WT1, and D2-40 for our IHC workup of ECIPAS and the pattern of staining was concordant with the reported literature. It is interesting to note that though Calretinin and WT1 are not expressed in this lesion there is basal expression of D2-40. Although D2-40 is a selective marker for germ cells and the lymphatic endothelium it is also a novel marker for cells with a mesothelial phenotype [54]. This is in keeping with the postulated congenital, primary, non-traumatic origin of these cysts that are favored to occur as a result of mesothelial cell misfolding in utero with invagination of the capsular surface mesothelium with subsequent cystic expansion and squamous metaplastic changes over time [55,56]. The rupture of the splenic capsule and small cystic spaces covered by mesothelium is probably one of the first steps in the genesis of the epithelial cyst of the spleen. This incorporation of mesothelial cells is a prominent determining factor in identifying splenic epidermoid cysts and is seen in their positive immunohistochemistry pattern of staining [56]. Our case report shows that the basal pattern of D2-40 indicates a primitive mesothelial cell phenotype. Contrastingly, the lack of Calretinin or WT1 expression indicates the absence of a more differentiated mesothelial phenotype.

It has been theorized that CA 19-9 and CEA are produced by the ECIPAS lining and disseminate into systemic circulation via trauma or elevated intracystic pressure. The changes in the serum CA 19-9 were recorded in our literature review (n = 36) highlighting the possible utility of this test in the preoperative work-up of ECIPAS. Elevated (61.1%, n = 22 out of 36) or normal (38.9%, n = 14 out of 36) levels of serum CA 19-9 were detected in these ECIPAS patients. There was no correlation between the gross size of ECIPAS and the elevation of CA 19-9. Interestingly, 10 of these patients with elevated serum CA 19-9 had a normalization of their serum CA 19-9 post-resection [6,13,14,17,18,20,22,33,39,40]. This post-operative decrease in CA 19-9 can be used to evaluate the extent of resection. Contrastingly, elevated levels of serum CEA were only concordant with CA 19-9 in 4 cases [13,14,30,37]. Therefore, intraoperative cyst fluid assessment for elevation in CA 19-9 paired with frozen section diagnosis can be considered as additional options to promote management options of spleen preserving surgical resections. All patients in the literature review were stable during their postoperative follow-up period. There is only one documented case of ECIPAS transformation to squamous cell carcinoma after 6 years from initial diagnosis [5]. Therefore, ECIPAS is predominantly a benign entity, and long-term follow-up with abdominal ultrasound paired with CA 19-9 may be utilized in monitoring these patients when clinically warranted.

The histogenesis of ECIPAS is yet to be discerned but the literature proposes two theories. The first theory suggests that similar to the histogenesis of splenic epidermoid cysts (SEC) there is an invagination of capsular mesothelial cells which undergo metaplastic change and cyst remodeling [55,56]. A genetic defect of mesothelial migration is considered the cause. While most are sporadic, rarely there is a familial occurrence [57]. The second theory suggests that these epithelial cysts are teratomatous derivatives or inclusion of fetal SSE [58]. Therefore, an exploration into the molecular mechanism of SEC may provide further insights into the histogenesis of ECIPAS. A genome-wide linkage and exome analyses study identified variants within the calcium-binding Epidermal Growth Factor (cb-EGF) like the domain of the fibulin 6 (HMCN1) protein in cases of SEC [59]. Fibulin 6 is an extracellular protein that is involved in cell adhesion, migration, and proliferation and is highly expressed in splenic tissue [60,61]. It is possible that instability in the HMCN1 structure can result in the misplacement of epithelial cells during developmental stages leading to SEC formation [57]. These variants in HMCN1 are also associated with age-related macular degeneration [62]. No biological studies exploring the function of human HMCN1 are available in the literature. Studies in the Zebrafish show that Hmcn1 mutation results in fin blistering due to cysts lined with epithelial cells [63]. HMCN1 could be a potential molecular candidate for future studies to explore the underlying molecular mechanism underlying the pathogenesis of ECIPAS.

Conclusion

The preoperative diagnosis of ECIPAS is extremely difficult as this entity shares overlapping radiological features with malignant pancreatic neoplasms and other cystic lesions such as mucinous pancreatic cysts thus resulting in oncological-based surgical resections. It is important for anatomic pathologists to recognize and consider the intrapancreatic compartment as a possible site for accessory spleen. This together with the

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evaluation of the cyst lining at the frozen section may facilitate consideration of cystectomy in relevant cases. The extent of surgical resection can be evaluated by measuring serum CA 19-9 in select cases. As histopathology is the final determinant of this diagnosis, increased clinical awareness with an accurate diagnosis of this entity may prevent patients from unnecessary surveillance and/or extensive oncological-based surgical resections.

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