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REVIEW ARTICLE

Solid Pseudopapillary Neoplasm — Case Series and Review of Literature

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Abstract

Solid-pseudopapillary neoplasm (SPN) is a variety of solid and cystic tumors of the pancreas. It was first described by Frantz in 1959. It is an unusual form of pancreatic carcinoma, with unknown etiopathogenesis, which accounts for about 0.17 to 2.7% of all pancreatic tumors. Here, we are describing 5 cases of pancreatic solid pseudopapillary neoplasm, out of 180 pancreatic tumors, operated in our institution in the 5-year period (2015–2020). Also, we have reviewed all available case series (from 2006 to 2020) in the literature, of pancreatic pseudopapillary neoplasm, for demographic information, etiopathogenesis, diagnosis, and extent of operation to establish the optimal management of this condition. Retrospective analysis of pancreatic tumors was carried out from February 2015 to January 2020. A total of 180 patients underwent pancreatic resection in this period for pancreatic tumor, out of which, the solid pseudopapillary neoplasm was confirmed in 5 cases (2.76%). Among these 5 cases, 4 cases (80%) were female and one (20%) male, with age group range from 14 to 45 years (mean age — 28 years). Abdominal pain was the most frequent presenting symptom (60%). Mean tumor diameter was 6.9 cm (range, 2–18 cm). Two patients were diagnosed preoperatively by CECT and MRI findings, and three patients were diagnosed preoperatively by percutaneous/USG-guided and CT-guided FNA cytology. Two patients underwent pancreatoduodenectomy; one patient underwent enucleation; and two patients underwent spleen preserving distal pancreatectomy. Four patients are alive and on regular follow-up, while one patient died on the 5th post-operative day due to post-operative sepsis.

Keywords Solid pseudopapillary neoplasm · Pancreas · Immunohistochemistry · Pancreatoduodenectomy

Introduction

Solid-pseudopapillary neoplasm (SPN) is a variety of solid and cystic tumors of the pancreas. It is a rare tumor of low malignant potential. It was first described by pathologist Virginia Kneel and Frantz in 1959, in the Armed Forces Institute of Pathology (AFIP) band on tumors of the pancreas [1]. The patient was a 2-year-old boy who died during an attempted pancreatoduodenectomy [1]. In 1970, Hamoudi et al. described the ultrastructural features of the tumor, which led to its acceptance as a separate clinicopathological entity [2]. It is a rare but characteristic neoplasm, with unknown etiopathogenesis, accounting for 0.17 to 2.7% of all pancreatic tumors and less than 5% of pancreatic cystic tumors [3] [4]. Previously it was known by various names including papillary epithelial neoplasm of the pancreas, solid and papillary tumors of the pancreas, and Hamoudi or Frantz tumors. In 1996, the World Health Organization (WHO) classified them as a borderline tumor of the exocrine pancreas and named them Solid pseudopapillary neoplasm [5]. The term SPN is currently the most frequently used name for this entity [6]. SPN occurs most frequently in young women of the 2nd to the 3rd decade of life [7]. The most commonly affected areas of the pancreas are the body and tail of the pancreas, accounting to approximately 60% of the diagnosed cases [7]. It usually has a favorable prognosis,

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with approximately 95% of patients reported as without any disease after surgical resection and with less than 3% mortality [7]. They are classically present as a large, solitary, well-circumscribed lesion, which can have a completely cystic, mixed cystic and solid, or a purely solid appearance on abdominal imaging [8]. The majority of patients have a localized disease, with only 10–15% presenting with metastasis or local invasion [9]. The mainstay of treatment is surgical resection, and the reported 5-year survival rate is as high as 94–97% [10]. In this study, we have focused on the general descriptive features of SPN and arrange by age, gender, symptoms, diagnostic tools, pathological features, surgery, and outcome.

Method

We collected and analyzed retrospective data on the clinical presentation, laboratory investigations, radiologic imaging, pathology, and operative details of patients with SPN of the pancreas, diagnosed between February 2015 and January 2020. Also, we searched keyword "solid," "pseudopapillary," "tumor," "neoplasm" in various indexed journals like PUBMED, MEDLINE, EMBASE, and SCOPUS, and collected all relevant data like demographic profile, etiopathogenesis, diagnosis, management and prognosis from all available case series, compiled them and reviewed them in the tabulate form for better understanding of these parameters. In the available literature, more than 90% of the cases are recorded in the last two decades, as more cases are diagnosed due to better imaging techniques and recent advances in immunohistochemistry. Therefore, here, we have reviewed the literature of the last two decades only (2006–2020), as before that understanding about the disease was not optimum, and SPN was classified by WHO (in 1996) two decades before only. The clinical presentation, radiological details, clinicopathologic feature, type of surgical procedure, operative time, postoperative complications, and prognosis data were collected and concluded.

Case Presentation

Case 1

A 14-year-old girl presented with upper abdominal swelling for 2 months. On physical examination, an 18 × 18 cm well-defined, a non-tender, non-pulsatile mass was palpable in the epigastrium and right hypochondrium. Blood investigations were normal. The tumor markers (CA19-9, CEA, and AFP) were normal. Computed tomography (CECT) showed a circumscribed encapsulated heterogeneous mass with solid and cystic areas arising from the head of the pancreas,

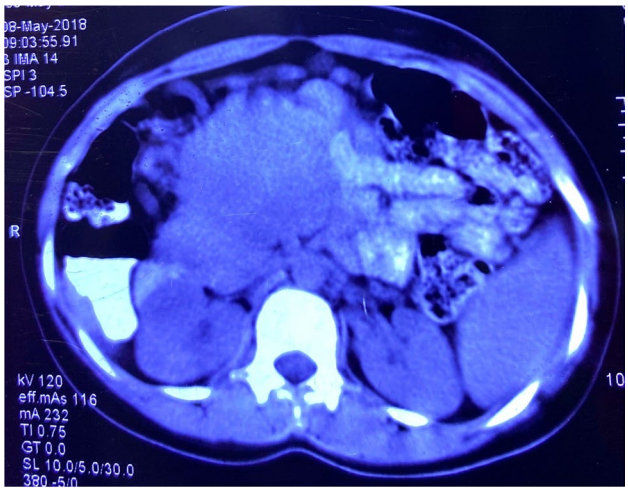

Fig. 1 CECT showing a heterogeneous mass with solid and cystic areas arising from the head of the pancreas

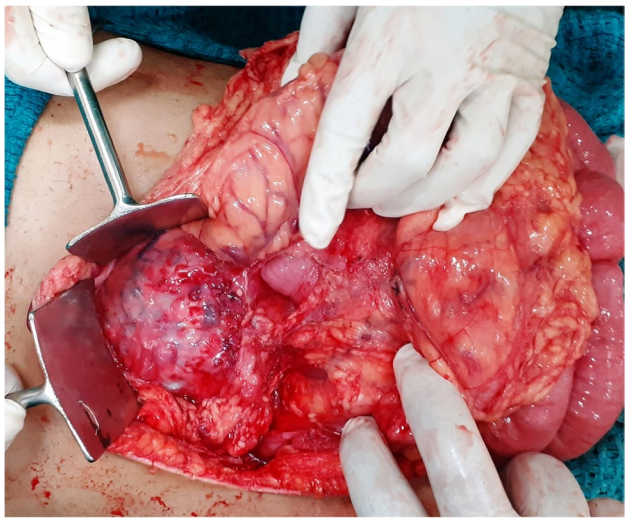

Fig. 2 Intraoperative picture showing pseudopapillary neoplasm in the pancreatic head region with solid and cystic consistency

measuring 18 × 15 × 16 cm (Fig. 1). No lymphadenopathy or other pathological findings were seen. USG-guided percutaneous core needle biopsy was done, which is consistent with SPN of the pancreas. Immunohistochemistry was positive for vimentin, progesterone-receptor, and beta-catenin with variable expression of pan keratin, synaptophysin, and neuron-specific enolase (NSE). Laparotomy confirmed a pancreatic head tumor (Fig. 2) without evidence of intraabdominal metastasis. The patient underwent a pylorus-preserving pancreatoduodenectomy. The resected margins were free of tumor, and all the 15 lymph nodes recovered were negative for malignancy. The final histological report confirmed the SPN (Fig. 3). The patient is currently disease-free 12 months after surgery.

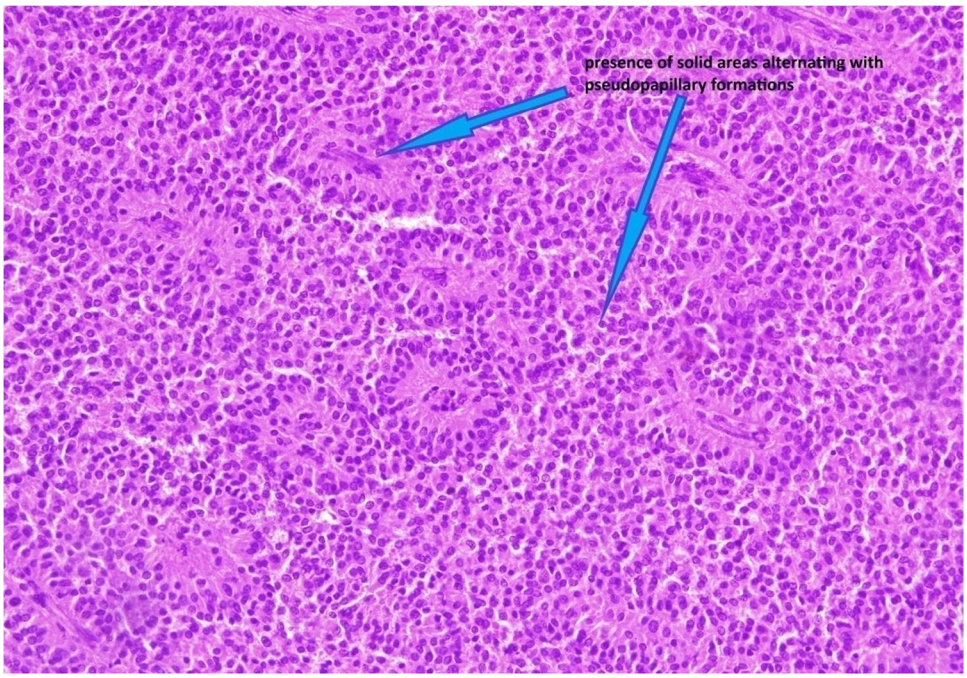

Fig. 3 Histopathological slide showing the presence of solid areas alternating with pseudopapillary formations in solid pseudopapillary neoplasm

Case 2

A 31-year-old lady presented with epigastric pain for 4 months. An abdominal ultrasound found a single 2-cm nodule in the body of her pancreas. CECT abdomen showed a regular, well-defined solid lesion with alternating cystic areas. Distal pancreatectomy without splenectomy was done. Histopathology confirmed the diagnosis. She is doing well after 3 years of follow-up.

Case 3

A 24-year-old lady presented with non-specific complaint of occasional nausea and epigastric discomfort for 3 months. USG showed a 6 × 6-cm lesion in the tail region of the pancreas. A CECT scan showed a solitary cystic mass in the pancreatic body and tail region. Ultrasound-guided fine-needle aspiration was performed, and cytopathological analysis was suggestive of a pseudopapillary solid tumor. She underwent a body-tail laparoscopic pancreatectomy without splenectomy. Ten months after the diagnosis, she remains asymptomatic, continuing regular follow-up in our out-patient clinic.

Case 4

A 45-year-old lady was admitted to the surgery department with severe epigastric pain over 3 days with a temperature of 38.5 °C. In the laboratory tests, slightly elevated white blood cells were found. An ultrasound (USG) examination and computed tomography (CECT) scan showed the presence of a tumor located in the head region of her pancreas (12 × 10 × 8 cm). The tumor markers carcinoembryonic antigen, carbohydrate antigen (CA 19–9), and alpha-fetoprotein (AFP) were all within the normal range. A CT-guided biopsy of the pancreatic mass showed a solid pseudopapillary tumor of the pancreas. Pancreaticoduodenectomy (Whipple procedure) was performed. Histopathology confirmed completely resected SPT. However, due to post-operative complications, she died on the 5th post-operative day due to uncontrolled sepsis.

Case 5

A 28-year-old gentleman was admitted to our department for an incidentally detected pancreatic mass revealed during USG. CECT scans showed a cystic and solid mass, 3 × 2.5 × 3 cm in size, arising from the body of the pancreas. MRI abdomen was done, which showed encapsulated, solid, and cystic tumors with intratumor hemorrhage in the body of the pancreas. Laparotomy showed a solid tumor 3 × 3 cm in diameter of the body of his pancreas. Enucleation with excision of 1-cm normal margin of the pancreas was performed. Histopathology revealed completely resected SPN with tumor-free margin.

Demographic Features

SPN occurs most frequently in young non-Caucasian women of the 2nd to the 3rd decade of life but can be seen rarely in men and children [7]. It seems to have a high incidence in Asian and African women [11]. It accounts

for approximately 8 to 15% of pancreatic tumors in children [9]. Our current review study suggested that SPN occurred in patients between 8.7 and 77 years of age, and the median age of diagnosis is 29.5 years. Female predominance (> 90%) has been attributed to the proximity of primordial pancreatic cells to the ovarian ridge during development [11].

Etiopathogenesis

The etiopathogenesis of SPN has not been fully understood [6]. The predominant occurrence of SPN in young women at the beginning of the reproductive period, along with the presence of progesterone receptors indicates the role of female hormones in the growth of this tumor [12]. Various theories have been given for its origin. The most accepted theory is SPN has been postulated to arise from primitive pancreatic cells (e.g., acinar cells, ductal epithelium, or endocrine cells) [13] or cell lines of the female genital bud [14]. Another hypothesis is an extrapancreatic origin from genital ridge–anlage-related cells [15]. Multiple mutations and alterations have been described for the development of SPN. Out of these, beta-catenin mutations, alterations of the wnt pathway, and disorganization of E-cadherin are the most important in the development of SPN [16]. The common expression of progesterone receptor and the strong predilection for females suggest that it might be a hormone-dependent tumor. However, estrogen receptors have not been demonstrated [17].

Clinical Features

The symptoms of SPN are usually non-specific. Most patients are asymptomatic and get diagnosed incidentally [18]. Abdominal pain or discomfort is the most common presenting symptom, accounting for approximately 37.6% of the cases [18]. Other signs and symptoms like jaundice, abdominal distention, abdominal mass, anorexia, nausea, vomiting, and weight loss may also be present [18]. Due to its slow growth and asymptomatic presentation, the tumor can reach to a large size at the time of identification in large proportion. Exocrine and endocrine insufficiencies have not yet been described. The mean size of the tumor is 6.5 cm; however, the tumor size range of 0.5 to 30 cm had been described [19]. The most common localization of SPN is the tail and body of the pancreas. In our study, the proportion of tumor location in the body and tail of the pancreas is 64%. Rarely, it may be found in the uncinate process also [20].

Histological Features

Grossly, the tumor is demarcated from the pancreatic tissue by the presence of a fibrous capsule [21]. Various solid, cystic, or mixed patterns with hemorrhagic and necrotic patches are seen on gross examination of SPN. SPN is commonly well-circumscribed and encapsulated with irregular degenerative cystic cavities and hemorrhages [22]. Microscopic examination reveals a mixed solid and cystic mass with hemorrhagic or necrotic cellular material in its center with lobules of solid tissue at its periphery. Characteristic findings include the presence of solid areas with pseudopapillary formations, foamy histiocytes, nuclear grooves, and cytoplasmic globules [23]. Malignant SPN, designated as a solid pseudopapillary carcinoma, occurs in 15% of adult patients [23]. According to the WHO classification system [5], these are:

1. Solid pseudopapillary neoplasms with borderline malignancy potential
2. Solid-pseudopapillary carcinoma

Criteria which distinguish potentially malignant tumors and which are classified as "solid papillary carcinoma" are [5]:

1. Angioinvasion
2. Perineural invasion
3. Deep invasion of the surrounding pancreatic parenchyma.
4. High nuclear grade and "necrobiotic nests."

Immunohistochemistry

This is the most important tool in the diagnosis of SPN. Beta-catenin has been regarded as a unique immunohistochemical feature of SPN as it underlies the genetic mutation of "catenin", found in more than 90% cases of SPN [24]. Beta-catenin and Wnt signalling pathway have been found to play an important role in the development of SPN [25]. In some cases, reactivity with epithelial markers and S-100 were described [22]. SPN cells also reveal immunoreactivity for neuroendocrine markers like S-100, vimentin, synaptophysin, neuron-specific enolase (NSE), and chromogranin. Abnormal nuclear and cytoplasmic beta-catenin expression and the presence of progesterone receptors are fairly common features of SPN. Also, SPN expresses galectin-3, CD-56, and CD10, all of which are useful in differentiating SPN from pancreatic neuroendocrine tumors. A low Ki-67 index (≤ 5%) indicates a slow growth of the tumor [22] [24] [25].

Metastasis

SPN is a rare tumor with a low malignant potential. Unlike adenocarcinoma and most other pancreatic tumors, SPN usually behaves in an indolent fashion, but with generally excellent prognosis. Malignant behavior with metastasis is observed in about 10–15% of the cases [23]. Some of them like liver and peritoneal metastasis can be treated with aggressive resection. Butte et al. suggested that larger tumors (> 6 cm) have more propensity of malignancy [26] while Park et al. suggested that deep parenchymal invasion is more consistent with malignant behavior of SPN [27]. Metastases were described in the liver, spleen, colon, peritoneum, and omentum [9]. Invasion of regional lymph nodes has been rarely reported [28]. Cai H et al. reported portal venous invasion and perineural invasion in 9 patients [29].

Differential Diagnosis

Because the 5-year survival rate of SPN is 94–97%, it is very important to differentiate it from other pancreatic lesions [6]. The typical presentation of a pancreas-associated solid and cystic upper abdominal mass with or without calcifications in a young woman should always raise the possibility of SPN. The differential diagnosis of SPN includes, solid and cystic lesions such as serous microcystic adenoma, cystadenocarcinoma, mucinous cystic neoplasms, solid and cystic neuroendocrine tumors, cystic acinar cell carcinoma, teratoma, pancreatoblastoma as well as a variety of congenital and acquired dysontogenic, and post-inflammatory and infectious cysts [30].

Diagnosis

There is no specific blood investigation to diagnose this condition. There are no specific tumor markers also. CA-19.9 is more specific for pancreatic adenocarcinoma.

Various imaging techniques can be used to diagnose pancreatic masses, such as abdominal ultrasound (USG), computed tomography (CECT), magnetic resonance imaging (MRI), and endoscopic ultrasound (EUS). SPN appears as a solid well-demarcated mass, usually heterogeneous in echotexture, sometimes containing hypoechoic fluid-filled cystic areas in USG [29]. CT scans show a heterogeneous mass, often with peripheral contrast enhancement corresponding to the fibrous pseudocapsule [31].MRI is superior to CT in distinguishing certain tissue characteristics, such as hemorrhage, cystic degeneration, or the presence of a capsule [32]. Typically, a large, well-defined, encapsulated lesion with heterogeneous high or low signal intensity on T1-weighted, heterogeneous high signal intensity on T2-weighted, and early peripheral heterogeneous enhancement with progressive fill-in are found on gadolinium-enhanced dynamic MRI. These features help differentiate this rare tumor from other pancreatic neoplasms [11].

Endoscopic ultrasound (EUS) has emerged as a very important tool in the diagnosis of pancreatic lesions, providing a better evaluation of the morphologic characteristics of the lesions, and the possibility of guided fine-needle aspiration cytology (FNAC), for tissue sampling, with a low risk of complications and increased diagnostic accuracy [16]. EUS-guided FNAC become the gold standard method for the diagnosis of pancreatic solid and cystic mass [33]. The sensitivity and specificity of EUS-FNAC for the diagnosis of pancreatic neoplasm range from 80 to 90% and from 85 to 96%, respectively [33]. SPN on EUS identifies as a well-circumscribed, solid, hypoechogenic, heterogeneous tumor with cystic components and calcifications [34]. Complications related to EUS-guided FNAC are only reported in approximately 1% of patients [33]. The most common complications are abdominal pain, fever, vomiting, bleeding, and rarely acute pancreatitis [16]. If EUS-guided FNAC not available, ultrasound or CT-guided percutaneous core needle biopsy could also be used with the same accuracy [35]. Laparoscopic or open biopsy may be done if guided-FNAC is not available or inconclusive [35].

Treatment

Complete surgical resection has been widely accepted as well as the standard of care in the treatment of SPN [36]. Incomplete excision should not be attempted due to the risk of tumor dissemination, development of pancreatic fistula, and the higher recurrence rate [37]. Complete surgical resection is the mainstay of treatment in all the patients with SPN, even in the presence of local invasion or distant metastasis [36]. The most common surgical procedures performed for pancreatic SPN are distal pancreatectomy with or without splenectomy, as the most common location of the tumor is the distal body or tail of the pancreas [21]. Pylorus preserving pancreatoduodenectomy or Whipple operation should be done for pancreatic head tumors [21]. Central pancreatectomy, segmental resection, and enucleation can be carried out for small tumors [36]. Extensive lymphatic dissection or resection of adjacent structures is not required, as lymph node metastasis is very rare [38]. There is debate on spleen preservation. Nakamura et al., after a case series on 14 patients, reported that a spleen-preserving operation is preferable for younger patients with SPN, as the tumor is mostly benign in nature [39]. Tumor size is not a criterion or predictor of unresectability because very large lesions may be resected without much problem [40]. Staging of the tumor also does not play any role in the treatment of SPN [6]. Local invasion and metastases are not a contraindication for resection [40]. Portal vein resection can be done when there is evidence of tumor invasion or malignancies [40].

Table 1 Comparison of demographic profile and presenting features of pseudopapillary tumor in various case series (2006–2020). Source: reference nos. [7] [8] [14] [15] [21] [23] [24] [26] [27] [28] [29] [31] [36] [37] [39] [42] [43]

Serial no	Year	Author	Total cases	Sex		Age group (mean)	Chief complaints			
				Male	Female		Asymptomatic	Pain abdomen	Abdomen mass	Others
1	2006	Dong et al	3	0	3	15–35 (24.5)	0	3	0	0
2	2006	Wang et al	9	2	7	15–44 (28.5)	NA	NA	NA	NA
3	2006	Tipton et al	14	1	13	30	NA	NA	NA	NA
4	2009	Yang et al	26	4	22	15–64 (32.3)	7	18	0	1
5	2011	Guo et al	24	1	23	11–61 (31)	5	10	8	1
6	2011	Frost et al	21	0	21	13–51 (24.6)	5	16	10	3
7	2011	Butte et al	45	7	38	10–63 (38)	6	35	3	1
8	2012	Song et al	3	2	1	29–44 (37)	1	2	0	0
9	2012	Lee et al	18	4	14	10–68 (32.4)	5	11	0	0
10	2012	Speer et al	11	1	10	9–17 (14)	2	9	0	0
11	2013	Wang et al	17	0	17	11–55 (26.6)	8	7	1	1
12	2013	Park et al	60	5	55	13–77 (34)	NA	NA	NA	NA
13	2013	Cai et al	33	2	31	12–59 (29.2)	8	20	4	1
14	2013	Vassos et al	4	0	4	15–42 (24.5)	0	2	1	1
15	2013	Yagci et al	10	1	9	18–71 (38.8)	3	7	0	0
16	2014	Law et al	34	3	31	16–81 (37)	14	19	0	1
17	2014	Park et al	11	3	8	10–18 (13.5)	1	6	4	0
18	2014	Manuballa et al	6	0	6	18–38 (27.7)	0	6	0	0
19	2014	Afridi et al	13	1	12	15–77 (21)	4	9	0	0
20	2014	Ren et al	19	2	17	29	5	6	4	4
21	2015	Uppin et al	33	1	32	12–62 (26.6)	1	29	3	0
22	2015	Zaneta et al	2	0	2	12–15 (13.5)	1	1	0	0
23	2016	Irtan et al	51	10	41	8.7–17.9 (13.1)	14	32	3	2
24	2016	Mirminachi et al	7	1	6	15–61 (29.4)	1	6	0	0
25	2016	Nakamura et al	14	1	13	14–45 (29.6)	NA	NA	NA	NA
26	2016	Nesrin et al	16	1	15	13–63 (35.7)	9	7	0	0
27	2017	Song et al	53	7	46	14–67 (35.4)	21	20	16	15
28	2017	Bhutani et al	11	1	10	17–41 (27.6)	3	8	0	0
29	2017	Xu et al	121	28	93	11–68 (33.7)	0	29	86	6
30	2017	Antoniou et al	5	0	5	13–47 (24.5)	1	4	4	2
31	2017	Lubezky et al	32	3	29	15–40 (28.4)	10	16	5	1
32	2018	Sachan et al	13	0	13	15–25 (20)	2	11	0	0
33	2018	De Moura et al	2	0	2	31–35 (33)	0	2	1	0
34	2019	Simona et al	2	0	2	36–51 (43.5)	1	1	0	0
35	March 2020	Cohen et al	35	3	32	18–64 (33.8)	15	11	0	11
36	April 2020	Priya et al	16	NA	NA	NA	NA	NA	NA	NA

NA Data not available

Neoadjuvant or adjuvant chemotherapy and radiotherapy have no role in the treatment of SPN [41]. Neoadjuvant chemotherapy was tried with some experimental regimes including 5-fluorouracil, doxorubicin, streptozocin, cisplatin, topotecan, iphosphamide, and etoposide in some series, but their role has not been established. Fried et al. observed a favorable response to radiotherapy in locally advanced unresectable disease with tumor shrinkage in one case series [41].

Prognosis and Mortality Rate

SPN carries an overall excellent prognosis, with a 5-year survival rate of up to 97% [6] [9] [11]. SPN is considered to be a tumor with low malignant potential; however, up to 10–15% of cases have been reported to be aggressive and can metastasize to the liver and/or peritoneum [42]. The Ki-67 index in immunohistochemistry is suggested as an indicator of malignant potential and

Table 2 Localization and malignant profile of pseudopapillary tumor in various case series (2006–2020). Source: reference nos. [7] [8] [14] [15] [21] [23] [24] [26] [27] [28] [29] [31] [36] [37] [39] [42] [43]

Serial no	Year	Author	Total cases	Size range/mean tumor size (cm)	Location			Metastasis/ malignancy	
					Head/uncinate	Body/neck	Tail	Yes	No
1	2006	Dong et al	3	NA	NA	NA	NA	NA	NA
2	2006	Wang et al	9	NA	NA	NA	NA	0	9
3	2006	Tipton et al	14	NA	3	1	10	2	12
4	2009	Yang et al	26	2–15 (6.25 cm)	14	4	8	2	24
5	2011	Guo et al	24	2.3–25.9 (7.5 cm)	8	5	11	2	22
6	2011	Frost et al	21	8–20 (12.5 cm)	13	2	6	1	20
7	2011	Butte et al	45	NA	10	12	23	9	36
8	2012	Song et al	3	1.7–5 (3.9 cm)	1	0	2	0	3
9	2012	Lee et al	18	NA	NA	NA	NA	1	17
10	2012	Speer et al	11	3.5–12 (5 cm)	NA	NA	NA	NA	NA
11	2013	Wang et al	17	2–10 (5.5 cm)	5	2	10	2	15
12	2013	Park et al	60	NA	NA	NA	NA	9	51
13	2013	Cai et al	33	2–15 (4.9 cm)	14	4	14	8	23
14	2013	Vassos et al	4	1–16 (5.5 cm)	3	0	1	1	3
15	2013	Yagci et al	10	NA	1	1	4	NA	NA
16	2014	Law et al	34	1.9–9.4 (4.2 cm)	NA	NA	NA	0	34
17	2014	Park et al	11	2.5–15 (7.9 cm)	4	4	3	1	10
18	2014	Manuballa et al	6	2–12 (5.7 cm)	1	2	3	0	6
19	2014	Afridi et al	13	1.5–11 (6 cm)	3	5	5	0	13
20	2014	Ren et al	19	6.3 cm	1	4	14	0	19
21	2015	Uppin et al	33	0.5–16 (8.6 cm)	8	6	19	6	27
22	2015	Zaneta et al	2	6–7 (6.5 cm)	1	0	1	1	1
23	2016	Irtan et al	51	NA	24	5	22	NA	NA
24	2016	Mirminachi et al	7	1.2–6 (4.2 cm)	2	3	3	2	5
25	2016	Nakamura et al	14	2–11 (4.8 cm)	0	2	12	0	14
26	2016	Nesrin et al	16	3–12 (6.9 cm)	6	0	10	NA	N
27	2017	Song et al	53	2–14 (6.4 cm)	20	2	31	10	43
28	2017	Bhutani et al	11	3–11 (6.9 cm)	2	1	8	3	8
29	2017	Xu et al	121	1.9–8.1 (4.9 cm)	32	19	70	19	102
30	2017	Antoniou et al	5	NA	1	0	4	0	5
31	2017	Lubezky et al	32	1–14 (5.9 cm)	NA	NA	NA	10	22
32	2018	Sachan et al	13	6.5 cm	10	0	3	2	11
33	2018	De Moura et al	2	1.4–2 (1.6 cm)	1	0	1	0	2
34	2019	Simonaet al	2	NA	0	1	1	1	1
35	March 2020	Cohen	35	0.9–14 (5.2 cm)	7	10	18	4	31
36	April 2020	Priya et al	16	NA	NA	NA	NA	NA	NA

NA data not available

poor outcome of SPN [43]. Some other features that may indicate poor prognosis are venous invasion, diffuse infiltrative growth pattern, extensive tumor necrosis, significant nuclear atypia, high mitotic count, nuclear pleomorphism, de-differentiation, DNA aneuploidy, double loss of X chromosomes, trisomy of chromosome 3, and unbalanced translocation between chromosomes 13 and 17 [24]. The overall mortality rate of SPN is about 2%, and the recurrence rate is almost 10–15% of patients after resection [44].

Conclusion

To conclude, SPN is a rare, but treatable pancreatic tumor. It occurs predominantly in young women in the 2nd to the 4th decade of life. The clinical manifestation of SPN is usually a slow-growing abdominal neoplasm with or without abdominal pain. It is of low malignant potential; however, some cases may be locally aggressive, with metastases to the liver, lung,

Table 3 Comparison of diagnosis, surgical procedure and prognosis of pseudopapillary tumor in various case series (2006–2020) Source: reference no. [7] [8] [14] [15] [21] [23] [24] [26] [27] [28] [29] [31] [36] [37] [39] [42] [43]

Serial no	Year	Author	Total cases	Preoperative diagnosis made		Treatment					Perioperative mortality (%)	Survival	
				Yes	No	DP	CP	PD	EN	Other		5 yr (%)	10 yr (%)
1	2006	Dong et al	3	1-CECT, 1-MRI	1	NA	NA	NA	NA	NA	NA	NA	NA
2	2006	Wang et al	9	2-EUS-FNAC, 7-CT FNAC	0	NA	NA	NA	NA	NA	0%	95	NA
3	2006	Tipton et al	14	7-CECT	7	9	0	3	1	1	9%	90	NA
4	2009	Yang et al	26	NA	NA	10	1	8	6	1	3%	97	NA
5	2011	Guo et al	24	18-CECT, 6-MRI	0	9	6	6	3	0	0%	91	NA
6	2011	Frost et al	21	10-CECT	11	8	4	5	3	1	5%	95	NA
7	2011	Butte et al	45	NA	NA	26	2	11	2	4	9%	95	NA
8	2012	Song et al	3	2-CECT, 1-EUS-FNAC	0	2	0	1	0	0	NA	NA	NA
9	2012	Lee et al	18	NA	NA	NA	NA	NA	NA	NA	5%	92	NA
10	2012	Speer et al	11	CECT-9, USG-2	0	NA	NA	NA	NA	NA	10%	90	NA
11	2013	Wang et al	17	8-CECT, 3-MRI, 2-USG-FNAC	4	7	1	4	5	0	0%	97	NA
12	2013	Park et al	60	NA	NA	NA	NA	NA	NA	NA	1.50%	97.3	NA
13	2013	Cai et al	33	NA	NA	14	4	15	0	0	3%	97	NA
14	2013	Vassos et al	4	2-Core needle biopsy	2	1	0	2	1	0	0%	96	NA
15	2013	Yagci et al	10	6-MRI	4	8	0	0	1	1	NA	NA	NA
16	2014	Law et al	34	18-CECT, 10-EUS, 6-EUS-FNAC	0	NA	NA	NA	NA	NA	NA	NA	NA
17	2014	Park et al	11	7-CECT, 3-MRI, 1-USG-FNAC	0	6	0	4	1	0	0%	95	NA
18	2014	Manuballa et al	6	0	6	0	0	2	0	4	0%	100	NA
19	2014	Afridi et al	13	8-CECT/MRI, 5-US/CT-guided biopsy	0	8	1	4	0	0	4%	96	NA
20	2014	Ren et al	19	CECT-2, MRI-3, USG-FNAC-2	12	12	1	4	0	2	0%	95	NA
21	2015	Uppin et al	33	23-CECT/MRI, 1-FNAC	9	23	2	6	0	2	NA	NA	NA
22	2015	Zaneta et al	2	1-CECT, 1-Lap biopsy	0	1	0	1	0	0	0%	NA	NA
23	2016	Irtan et al	51	39-CECT/MRI, 12-FNAC/Biopsy	0	22	5	19	0	5	0%	100	NA
24	2016	Mirminachi et al	7	7-EUS-FNAC	0	3	0	4	0	0	0%	NA	NA
25	2016	Nakamura et al	14	0	14	14	0	0	0	0	0%	NA	NA
26	2016	Nesrin et al	16	4-CECT	12	7	0	3	0	6	6%	NA	NA
27	2017	Song et al	53	50-CECT/MRI,3-EUS-FNAC	0	28	2	9	10	4	0%	NA	NA
28	2017	Bhutani et al	11	NA	NA	7	0	0	3	1	8%	NA	NA
29	2017	Xu et al	121	NA	NA	71	10	34	6	0	7%	NA	NA
30	2017	Antoniou et al	5	0	5	4	0	1	0	0	0%	NA	NA
31	2017	Lubezky et al	32	NA	NA	NA	NA	NA	NA	NA	0%	NA	NA
32	2018	Sachan et al	13	NA	13	3	0	10	0	0	0%	NA	NA
33	2018	De Moura et al	2	2-EUS-FNAC	0	1	0	1	0	0	50%	NA	NA

peritoneum, and skin. Exocrine and endocrine insufficiencies have not yet been described. The pathogenesis is still unknown. On histological examination, the tumor is a solid and cystic mass with pseudopapillary and pseudo cystic structures with rich microvasculature pattern. EUS/USG/CT-guided FNAC is the gold standard in the preoperative diagnosis of SPN of the pancreas. The typical radiological appearance of solid pseudopapillary tumor in CECT scan, and MRI is also diagnostic. Surgical resection is the treatment of choice. Prognosis of SPN is excellent, with a 5-year survival of about 90–95%. The prognosis is favorable even in the presence of distant metastasis. Surgical resection is generally curative. A close follow-up is advised to diagnose a local recurrence or distant metastasis and choose the proper therapeutic option for the patient.

We have summarized the demographic features of SPN in various case series described in recent literature in Tables 1 and 2.

We have summarized the diagnostic method and treatment of SPN in various case series described in recent literature in Table 3.

Abbreviations SNP: Solid pseudopapillary neoplasm; CT: Computed tomography; CECT: Contrast-enhanced computed tomography; EUS: Endoscopic ultrasound; FNAC: Fine-needle aspiration cytology; MRI: Magnetic resonance imaging

Declarations

Consent to Participate I declare that consent has been obtained from the patient or subject after full explanation of the purpose and nature of all procedures used. I also declare that approval is not required in our study as the patient is not harmed during all procedures.

Conflict of Interest The authors declare no competing interests.

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Table 3 (continued)

Serial no	Year	Author	Total cases	Preoperative diagnosis made		Treatment					Perioperative mortality (%)	Survival	
				Yes	No	DP	CP	PD	EN	Other		5 yr (%)	10 yr (%)
34	2019	Simona et al	2	1-CECT	1	0	0	0	2	0	0%	NA	NA
35	March 2020	Cohen	35	0	35		0	5	2	3	NA	NA	NA
36	April 2020	Priya et al	16	16-USG-FNAC	0	NA	NA	NA	NA	NA	NA	NA	NA

CECT contrast enhanced computed tomography, *MRI* magnetic resonance imaging, *EUS* endoscopic ultrasound, *USG* ultrasonography, *FNAC* fine needle aspiration cytology, *DP* distal pancreatectomy, *CP* central pancreatectomy, *PD* pancreatoduedenectomy, *EN* enucleation, *NA* data not available

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