Case Report

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Unveiling the Uncommon: A Rare Encounter of Solid Pseudopapillary Neoplasm of Testis

Pathology Section

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ABSTRACT

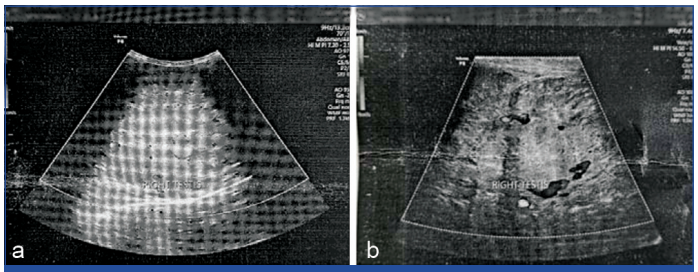
Solid Pseudopapillary Neoplasm (SPN) of the pancreas is a rare and intriguing entity within the spectrum of pancreatic neoplasms, accounting for approximately 1-2% of all pancreatic tumours. Extra-pancreatic SPN is even rarer. It has been reported in locations such as the mesocolon, ovary, retroperitoneum, paratesticular region, and others. A case of extra-pancreatic SPN of the testis was reported in the Department of Pathology, Government Medical College, Kottayam, in 2024. A 48-year-old male presented with a right hemiscrotal swelling of one-year duration, with normal serum tumour markers and laboratory investigations. Radiological evaluation (MRI and Doppler ultrasound) suggested a neoplastic aetiology, and the patient underwent high inguinal orchidectomy. Histopathological examination revealed morphological features consistent with SPN, supported by immunohistochemical findings.

Keywords: Beta catenin, Hypointense, Inguinal orchidectomy, Surrogate marker

CASE REPORT

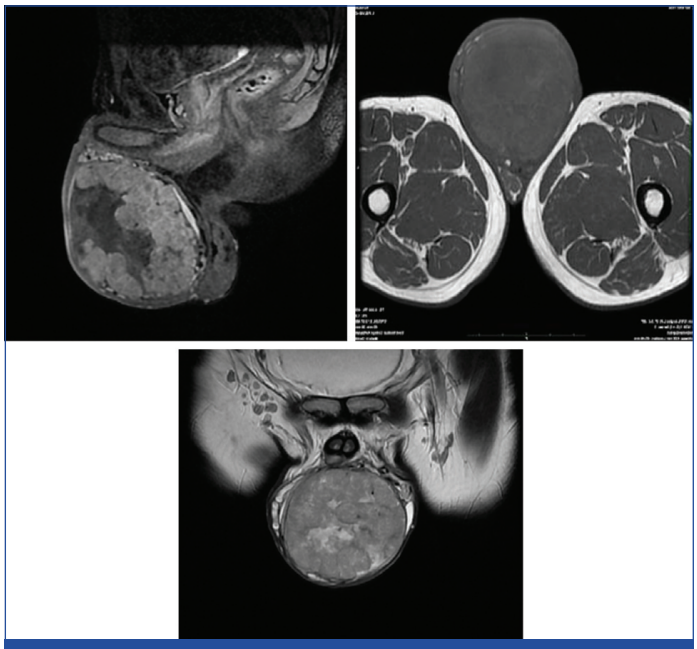
A 48-year-old male with no known co-morbidities presented to the Department of Urology with complaints of right hemiscrotal swelling for the past one year. Initially, the swelling was small and gradually increased over time. There was no history of pain, fever, trauma, or sudden increase in size. On clinical examination, a hard mass measuring approximately 10×10×7 cm was palpated in the right hemiscrotum. The overlying skin appeared normal with no tenderness or localised rise in temperature. The right testis was not separately palpable, although it was possible to get above the swelling. The left testis was normal. No abdominal masses or lymphadenopathy were detected.

Routine laboratory investigations were within normal limits. Serum lactate dehydrogenase (LDH), alpha-fetoprotein (AFP), and beta-human chorionic gonadotropin (β-hCG) levels were within normal limits. Ultrasonography of the abdomen and pelvis revealed cholelithiasis and right renal microlithiasis. The pancreas appeared normal, with no ductal dilatation or peripancreatic collection. Scrotal Doppler ultrasonography [Table/Fig-1] demonstrated a large heteroechoic mass in the right scrotal sac, with the right testis and epididymis not visualised separately.



[Table/Fig-1]: Scrotal doppler study showing a large heteroechoic mass lesion in the right scrotal sac.

Magnetic Resonance Imaging (MRI) of the scrotum [Table/Fig-2] showed a large, relatively well-defined heterogeneous mass in the right testis. The lesion appeared heterogeneously hyperintense on the T2-weighted images and hypointense on the T1-weighted images. Solid areas demonstrated intense post-contrast enhancement, suggestive of a neoplastic aetiology. The patient subsequently underwent high inguinal orchidectomy. Intraoperatively, a large mass measuring approximately 10×10×10 cm was noted, extending up to the root of the right scrotum, without infiltration of the scrotal wall and with no clear distinction of the epididymis.

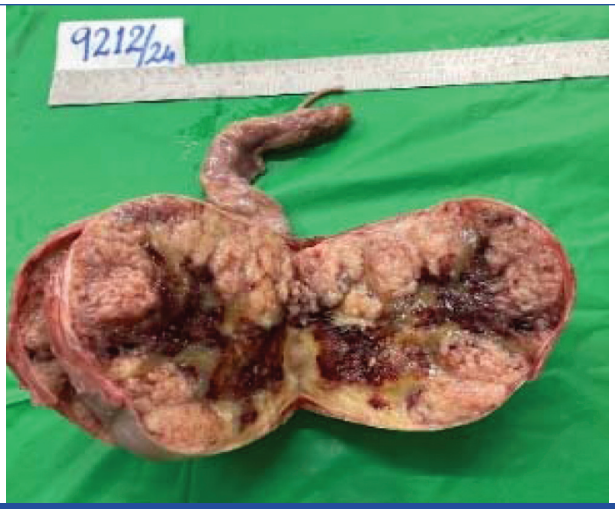


[Table/Fig-2]: MRI scrotum showing large well-defined heterogenous lesion in the right testis.

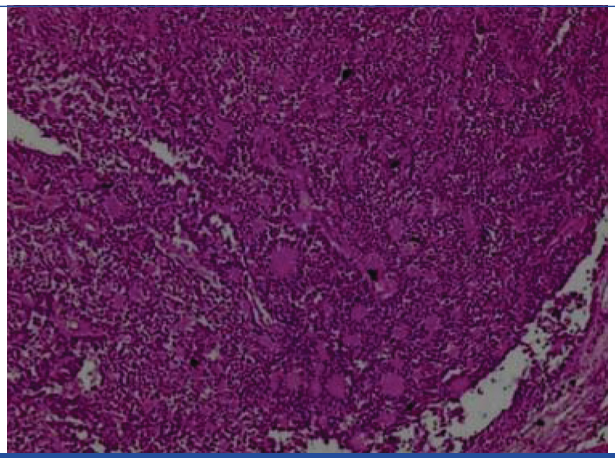
The postoperative period was uneventful. The excised specimen, consisting of an enlarged testis with attached spermatic cord, weighed 632 g. The testis measured 12×10×9 cm, and the spermatic cord measured 13.5 cm in length. The external surface appeared congested. On sectioning, a grey-white solid lesion with haemorrhagic and myxoid areas measuring 12×10×9 cm was identified. Normal testicular tissue and epididymis were not grossly identifiable [Table/Fig-3].

Histopathological examination revealed a neoplasm composed of monomorphic cells arranged in nests and solid patterns with fibrovascular cores. The tumour cells showed vacuolated cytoplasm and vesicular nuclei, with focal clear cell areas. Areas of necrosis and eosinophilic acellular hyaline globules were also noted [Table/Fig-4-7]. The epididymis was involved by the neoplasm, and seminiferous tubules were observed at the periphery. A provisional diagnosis of extra-pancreatic solid pseudopapillary neoplasm was made.

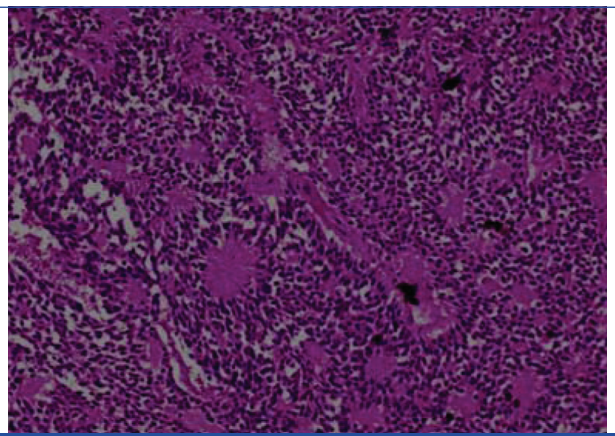
Immunohistochemical studies [Table/Fig-8-15] demonstrated positivity for beta-catenin (nuclear), Cluster of differentiation 10 (CD 10), vimentin, and Neuron-Specific Enolase (NSE). The tumour



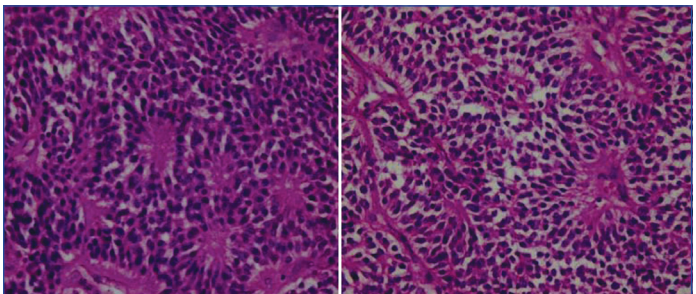
[Table/Fig-3]: Gross specimen of testis showing extensive areas of haemorrhage and grey white cystic lesion.



[Table/Fig-4]: Neoplasm composed of cells arranged in pseudopapillary pattern, nests and sheets (H&E, 4x).

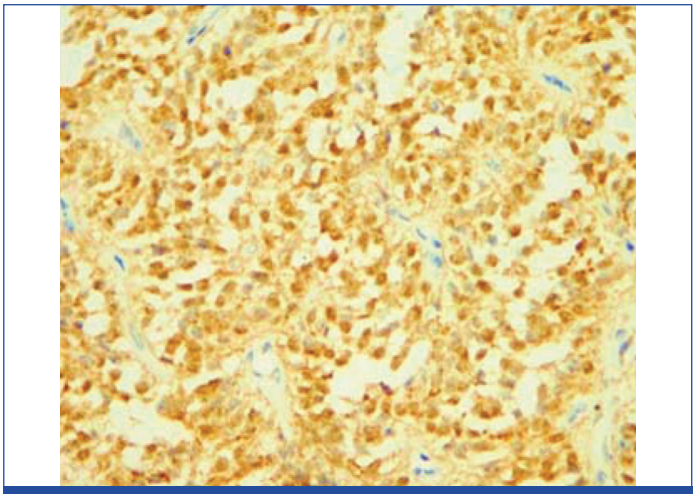


[Table/Fig-5]: Neoplastic cells in pseudopapillary pattern (H&E, 10x).

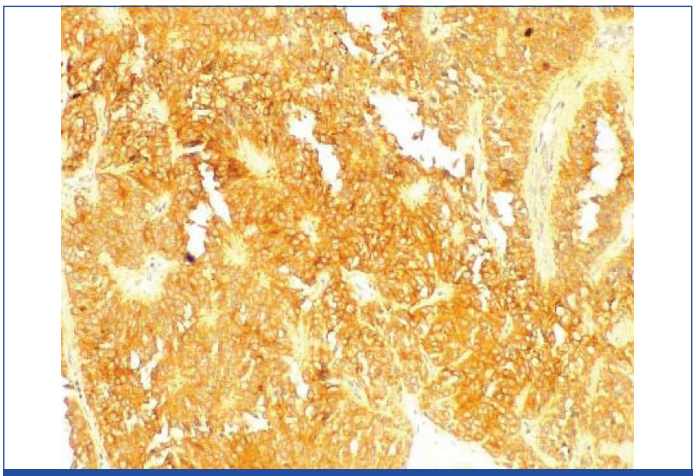


[Table/Fig-6,7]: Neoplastic cells are round with scant cytoplasm, round nucleus with fine chromatin (H&E, 40x). Images from left to right.

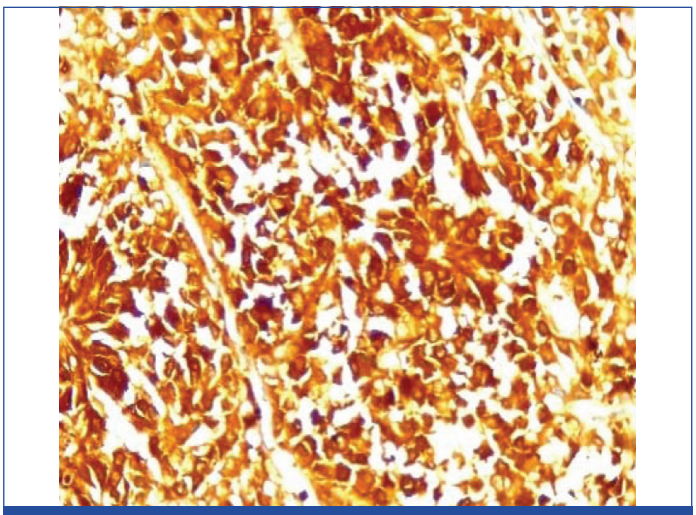
cells were negative for inhibin, calretinin, and CD117. Based on the histomorphological features and immunoprofile, a final diagnosis of extra-pancreatic SPN of the testis was established.



[Table/Fig-8]: Neuron Specific Enolase (NSE) positive cells (IHC, 40x).



[Table/Fig-9]: CD 10 positive in the neoplastic cells (IHC, 40x).



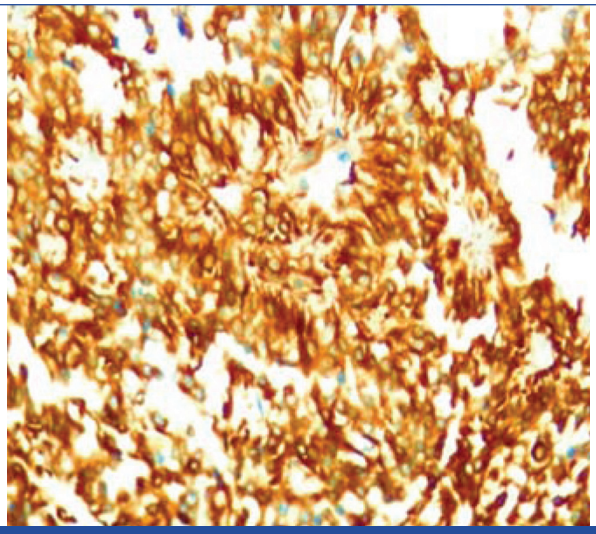
[Table/Fig-10]: Beta catenin showing nuclear positivity (IHC, 40x).

The patient was advised to undergo genetic testing for mutations in exon 3 of the CTNNB1 (beta-catenin) gene. Due to financial constraints, the patient declined testing. However, nuclear positivity for beta-catenin on immunohistochemistry serves as a reliable surrogate marker, supporting the diagnosis [1].

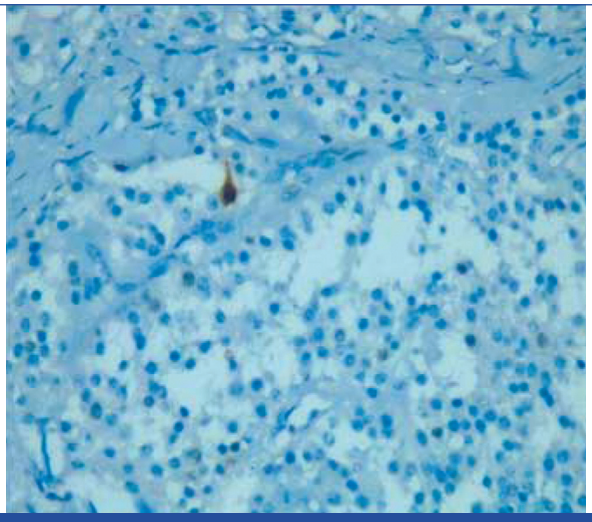
The patient remains under regular follow-up to date.

DISCUSSION

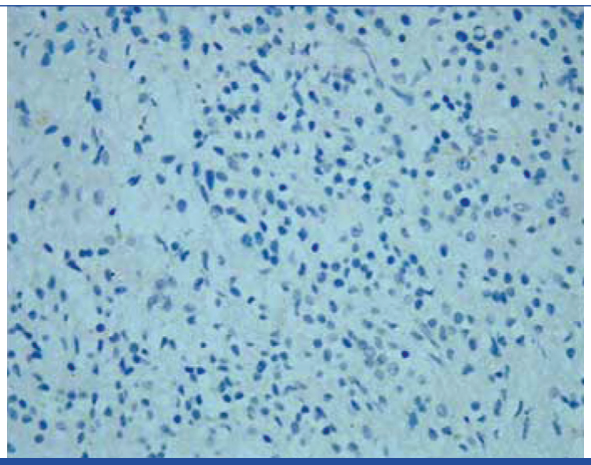
The SPN is a rare pancreatic tumour of unknown histogenesis. It was first described in 1959 by Franz as a distinct clinicopathological entity. The current World Health Organisation (WHO) classification retains the widely accepted term “SPN,” which reflects the two most prominent histological features of the tumour: solid and pseudopapillary areas [2].



[Table/Fig-11]: Vimentin positive cells (IHC, 40x).

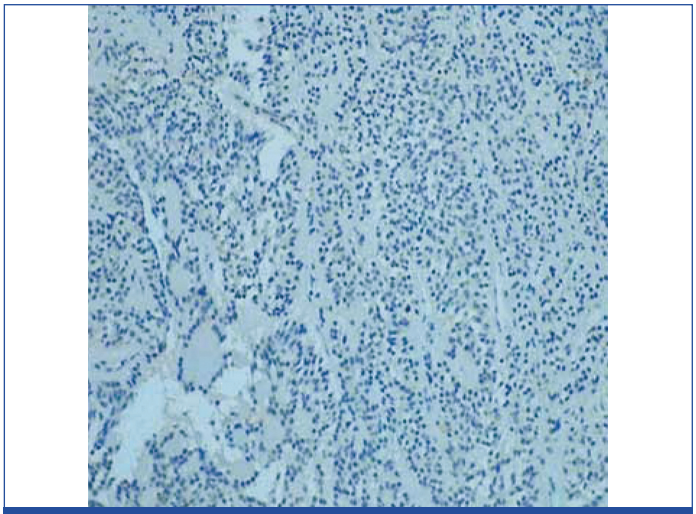


[Table/Fig-12]: Ki-67 index low (1%) (IHC, 40x).

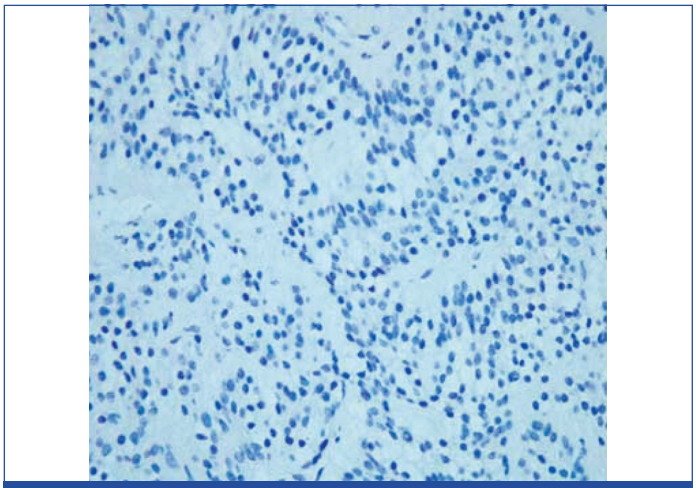


[Table/Fig-13]: Calretinin negative (IHC, 40x).

This neoplasm shows a strong female predominance, with a reported female-to-male ratio of approximately 10:1. Around 50 cases of extra-pancreatic SPN have been documented in the literature to date [2]. In some extra-pancreatic SPN cases, ectopic pancreatic tissue has been identified, suggesting that these tumours may arise from pancreatic progenitor cells within such tissue [2]. However, in most cases—including the present case—ectopic pancreatic tissue was absent. This observation supports the hypothesis that SPNs may originate from genital ridge-related cells, possibly derived from ectopic stem cells migrating during embryogenesis. Most patients with SPN are asymptomatic, and lesions are often detected incidentally during imaging for unrelated conditions. Extra-pancreatic presentations typically produce site-specific symptoms.



[Table/Fig-14]: Inhibin negative (IHC, 10x).



[Table/Fig-15]: CD117 negative (IHC, 40x).

In the present case, the patient was asymptomatic apart from scrotal swelling. In appropriate clinical settings, imaging studies can be highly suggestive of SPN [3]. However, in this case, radiological diagnosis was challenging due to the unusual location.

Typical Computed Tomography (CT) findings include a heterogeneous mass with solid and cystic components, peripheral arterial enhancement, and central calcification. MRI provides further detail, demonstrating heterogeneous signal intensity and areas of haemorrhage [4]. Grossly, SPNs are solitary, often haemorrhagic, round masses ranging from 0.5 to 25 cm in size, with an average diameter of approximately 9 cm [5,6]. They are well-circumscribed and frequently encapsulated, displaying a variegated cut surface with solid regions intermixed with haemorrhage, necrosis, and cystic degeneration [6]. The cystic spaces may contain necrotic debris. Although typically soft in consistency, some tumours may be fibrotic. Due to their cystic and haemorrhagic nature, SPNs may resemble pancreatic pseudocysts, necessitating thorough histological sampling for accurate diagnosis. Histologically, SPNs demonstrate a heterogeneous architecture comprising solid areas, pseudopapillary structures, and necrotic debris. Tumour cells surround delicate blood vessels within hyalinised and myxoid stroma, forming pseudopapillae as cells detach. The neoplastic cells may mimic ependymal rosettes; however, true glandular formations are absent. Solid areas contain loosely cohesive cells, multinucleated giant cells, cholesterol crystals, and periodic acid-Schiff-positive diastase-resistant hyaline globules. Although SPNs are considered tumours with low malignant potential, aggressive dedifferentiated variants have been described.

Given their rarity, a broad panel of immunohistochemical stains is often required to exclude differential diagnoses. The hallmark immunohistochemical feature of SPN is aberrant nuclear

expression of beta-catenin, reflecting underlying CTNNB1 (Catenin beta) gene mutations [2]. There is some overlap between SPN and pancreatic neuroendocrine tumours (PanNETs); however, immunohistochemistry aids differentiation. SPNs commonly show variable positivity for neuroendocrine markers such as synaptophysin, CD56, and NSE [7]. In contrast, chromogranin is typically negative in SPNs, as observed in the present case [8]. Immunostaining for beta-catenin and E-cadherin is particularly helpful in distinguishing SPN from neuroendocrine tumours. Most SPNs demonstrate nuclear beta-catenin positivity and loss of membranous E-cadherin expression, whereas neuroendocrine tumours show cytoplasmic and membranous beta-catenin staining along with strong E-cadherin expression [9]. These findings were consistent with the present case. Additional markers such as progesterone receptor, alpha-1 antitrypsin, CD10, and CD56 may also show positivity. Genetically, SPNs are characterised by somatic hotspot mutations in exon 3 of the CTNNB1 gene, leading to stabilisation and nuclear accumulation of beta-catenin detectable by IHC [10]. Beta-catenin is a key component of the Wnt signalling pathway and regulates transcription of downstream target genes. Mutations result in abnormal nuclear accumulation and pathway activation.

The primary differential diagnosis in this case includes Sertoli Cell Tumours (SCTs), including the sclerosing variant, which may share certain morphological and immunohistochemical features with SPN [11]. Notably, beta-catenin mutations and nuclear expression have also been reported in SCTs. However, SCTs typically show positivity for calretinin and inhibin, both of which were negative in this case. The signet-ring cell stromal tumour is considered a rare variant of SCT. The present case findings concur with Michal et al., that signet-ring cell stromal tumours of the testis may represent a distinct neoplasm of uncertain origin and possibly a morphological variant of testicular SPN [12]. The prognosis of SPN is generally excellent following complete surgical excision. Approximately 95% of patients achieve cure when the tumour is confined to the pancreas. Extra-pancreatic SPNs are considered tumours of low malignant potential; however, cases with metastasis and mortality—particularly in male patients—have been reported [13].

CONCLUSION(S)

The SPN of the testis is an exceptionally rare entity, with only a limited number of cases reported in the literature. Diagnosis requires a high index of suspicion, as clinical and radiological findings are often non specific and may mimic other testicular tumours. Histopathological examination supported by IHC remains the gold standard for definitive diagnosis. Although SPN generally carries a favourable prognosis, a long-term follow-up is recommended due to its uncertain malignant potential and the scarcity of data regarding its natural history in extra-pancreatic locations. Reporting rare cases improves understanding of the disease spectrum and aids refinement of diagnostic and therapeutic strategies.

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