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CASE REPORT

Primary ovarian solid pseudopapillary neoplasm associated with familial adenomatous polyposis

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Abstract

Background: Solid pseudopapillary neoplasms (SPNs) are rare tumours, most often present in the pancreas. Extra-pancreatic SPNs are especially rare. Although FAP has not classically been associated with ovarian neoplasms, here we present the first documented case of a primary ovarian SPN in a patient with a known history of FAP.

Case presentation: We report a patient who developed ovarian SPN on follow-up for FAP. A 25-year-old female with a background of FAP presented with a large abdominal mass, raised serum Ca125. Subsequently, the right salpingoopherectomy microscopically and immunocytochemically confirmed a Solid pseudopapillary neoplasm.

Conclusion: This case highlights awareness of possible occurrence of solid pseudopapillary neoplasm in the ovary in FAP patients. It is therefore important that SPNs are considered in the clinical and pathological differential diagnoses of tumors presenting in FAP patients.

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Introduction

Solid pseudopapillary neoplasms (SPNs) are rare tumours that were first described in the pancreas in 1959 (1). These tumours most often present in the pancreas, although they represent fewer than 3% of all pancreatic neoplasms (2,3). They usually affect young patients (4,5) and there is a distinct preponderance in females, which account for nearly 90% of cases (3,5). Extra-pancreatic SPNs are especially rare (4).

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In 2010, Deshpande et al reported the first three primary ovarian SPNs, which morphologically and immunophenotypically resembled pancreatic SPNs (2). Since then, there have only been a few further case reports of 13 separate primary ovarian SPNs (6).

Majority of SPNs exhibit abnormal nuclear accumulation of beta-catenin, which is secondary to germline *APC* tumor suppressor gene mutation (7,8).

Although FAP has not classically been associated with ovarian neoplasms, rare occurrence of microcystic stroma tumour of the ovary have been described in patients with FAP.

Here we present a case of primary ovarian SPN in a patient with a known history of FAP, adding to the existing literature on this rare entity.

Case Report

A 25-year-old primigravid female presented clinically with a large abdominal mass. She had a raised serum Ca125 at 201kU/L. Her serum LDH, AFP and HCG were within normal limits. Her BMI was 37kg/m^2 and she had a background of FAP.

An initial pelvic ultrasound scan showed a 239mm complex cystic mass with septation and solid components within the pelvis and extending up to the xiphisternum. A subsequent CT scan of the thorax, abdomen and pelvis showed a multiloculated cystic mass that measured 322x255x206mm and had a likely right adnexal origin. There was no evidence of distant metastases.

Laparotomy was done. Intraoperatively, the right ovarian mass was identified and there was evidence of torsion. The uterus was unremarkable other than a fibroid and the left fallopian tube and ovary were normal. There was no evidence of ascites. The omentum looked inflammatory. Right salpingo oophorectomy, omentectomy and peritoneal washings were performed. One prominent supracolic lymph node was also excised.

Macroscopic measurement of the right ovary was 320x270x10mm. The capsule was intact and it had a dusky, smooth outer surface. Cut section of the ovary revealed a multiloculated cyst that was partially solid and partially haemorrhagic and necrotic *(Figure 1)*.

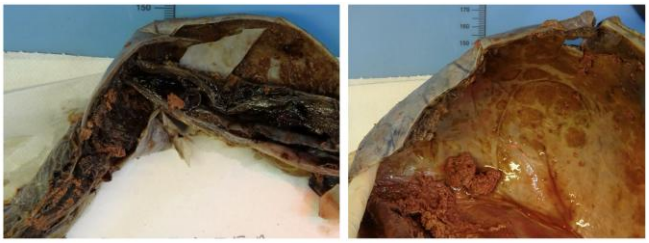


Figure 1: Macroscopic features of the ovary: An intact dusky capsule that on cut section revealed a multiloculated cyst that was partially solid, partially haemorrhagic and necrotic.

The associated fallopian tube was macroscopically normal. The omentum was associated with fibrinous material.

Microscopic examination revealed a tumour that showed a solid tumour with heterogenous morphology. It had a vaguely papillary architecture *(Figure 2A)*. There were areas of cellular nests separated by fibrous stroma. The stroma was also vascular in places and showed hyalinisation. The tumour cells had bland, uniform round to oval nuclei and there were no mitotic figures. Tumour cells contained dense, hyalinised intracytoplasmic eosinophilic granules in others *(Figure 2B)*.

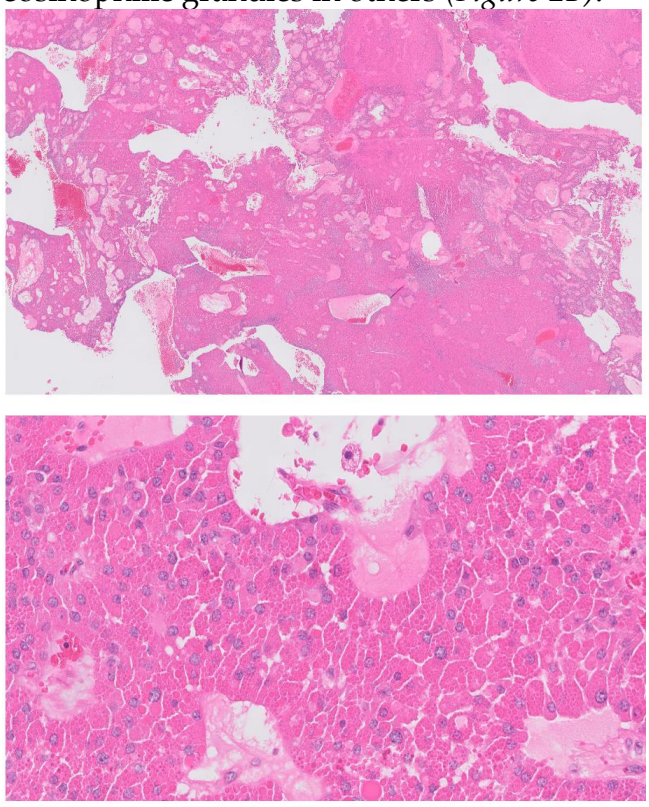


Figure 2: Microscopic features of the ovary: (A) show a solid tumour with heterogenous morphology. There is a papillary architecture separated by fibrous stroma. The stroma is also vascular in places and shows hyalinisation. (B) Tumour cells display bland, uniform round to oval nuclei with foamy cytoplasm in some areas and dense, hyalinised intracytoplasmic eosinophilic granules in others. Mitotic figures are absent.

Immunohistochemistry was performed and the tumour cells were positive for beta-catenin (nuclear and cytoplasmic) *(Figure 3A)*, CD56 *(Figure 3B)*, vimentin *(Figure 3C)*, CD10 *(Figure 3D)* and alpha 1 antitrypsin.

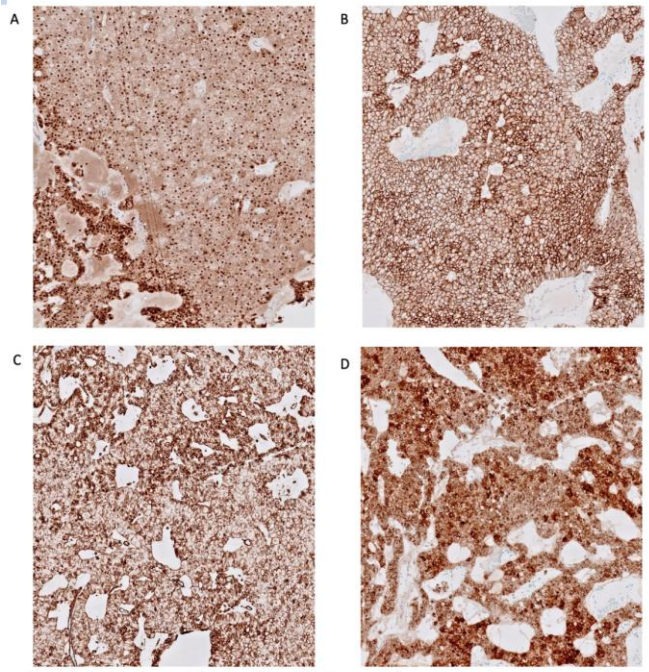
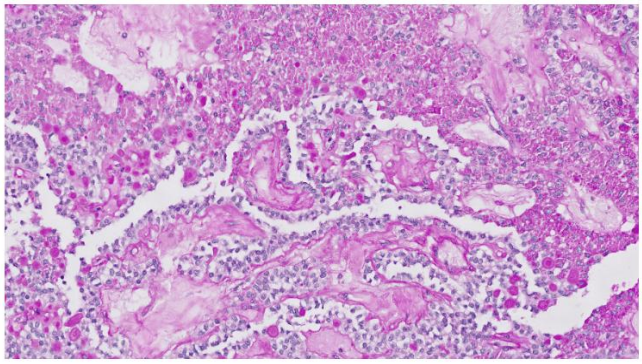


Figure 3: Positive immunohistochemistry of the tumour: (A) aberrant nuclear and cytoplasmic beta-catenin, (B) CD56, (C) vimentin, (D) CD10.



(Figure 4): PAS stain was positive in the intracytoplasmic globules

The tumour cells were negative for epithelial markers, S100, Melan A, CD30, AFP, HCG, Oct4, SALL4, inhibin, chromogranin and synaptophysin.

The supracolic lymph node was negative and the omentum showed areas of chronic inflammation and fibrosis. The peritoneal washings were negative for malignancy.

A follow up CT scan of the abdomen and pelvis that was carried out after the microscopic diagnosis of ovarian SPN showed a normal pancreas. Three years post-operation, the patient is well and follow up imaging to assess the left fallopian tube and ovary show findings consistent with polycystic ovarian syndrome only. Six monthly follow up serum CA125, CEA and CA19-9 tumour markers have been within normal range.

Given that this patient has FAP, she has also since undergone a prophylactic total colectomy for FAP, which demonstrated numerous characteristic adenomas *(Figures 5 and 6)*.

Figure 5: Total colectomy in this patient displaying multiple colonic polyps, characteristic of FAP

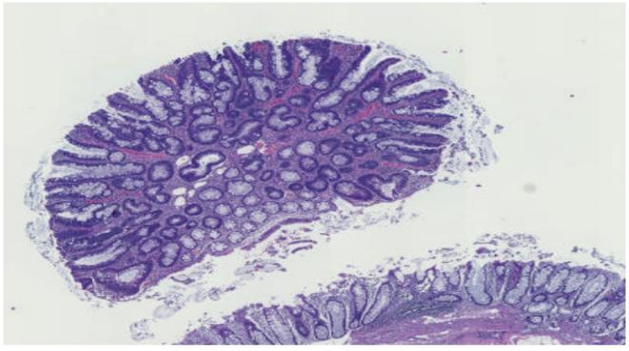


Figure 6: Tubular adenoma with low grade dysplasia

Discussion

FAP is caused by mutations of the APC gene. A loss of function mutation in the APC gene is associated with an accumulation of intra cellular beta-catenin signalling, which promotes colorectal adenoma formation.

Extracolonic manifestations are likely to occur in patients with FAP, most of which were reported to be ductal adenocarcinoma of the pancreas. A few reports of pancreatic SPN in patients with FAP has been noted (7).

We report a case of primary SPN in ovary that resemble the SPN of pancreas both morphologicallyandimmunohistochemically.

Grossly SPN of pancreas can vary from entirely cystic with small mural nodule to solid but most tumours are solid and cystic like the neoplasm we describe in the ovary. These neoplasms frequently undergo haemorrhagic cystic degeneration and occur predominantly in young women as seen in our case. Histologically the tumour closely resembled their pancreatic counterparts showing solid and pseudopapillary patterns. The nuclear features including oval nuclei, variable longitudinal nuclear grooves, and fine chromatin are identical to those seen in their pancreatic counterpart (2).

Given the negative findings in the pancreas on imaging studies and lack of tumour elsewhere, this neoplasm is a primary ovarian neoplasm, similar to the three cases reported by Deshpande et al. (2)

The neoplasms are diffusely positive for CD 56. This aberrant expression of neuro endocrine markers in this non-neuroendocrine tumour is well recognised. Significantly, this ovarian neoplasm was negative for chromogranin, like their pancreatic counterpart. The two recently described markers of pancreatic SPN, beta catenin and e-cadherin are now believed to be the most robust markers of this neoplasm. SBN of the pancreas invariably show nuclear reactivity for beta catenin, whereas pancreatic neuroendocrine tumours lack nucleus staining. This evident nuclear localisation of beta catenin is the result of mutation in Exon 3 of this gene (2).

Ovarian neoplasms that display solid and pseudopapillary or true papillary growth pattern should be included in the differential diagnosis such as carcinoid, struma ovarii, paraganglioma, granulosa cell tumour, Sertoli cell tumour, steroid cell tumour and Microcystic stromal tumour. The constellation of morphological features such as cellular dehiscence, rich vascularity, and hyaline globules together with nuclear beta catenin expression and lack of reactivity for chromogranin and inhibin should distinguish this tumour from the mimickers (4).

Microcystic stromal tumour is a subtype of ovarian stromal neoplasm that was first reported in 2009 and is characterised by prominent histological microcytic and solid patterns as well as diffuse CD10 and vimentin immunoreactivity. Histologically MCSTs and SPNs exhibit solid – cystic growth pattern and haemorrhagic changes, and the tumour cells have abundant eosinophilic cytoplasm. However, there are some significant differences between them. A pseudopapillary structure is characteristic of SPN, whereas the structure is absent in MCST. Additionally, immunoreactivity for markers such as WT-1 and CD 56 is observed in SPN. In recent report, MCST was found to exhibit beta catenin nuclear reactivity and to carry the point mutation c.98C >G in exon 3 of CTNNB 1, this mutation causes replacement of serine with cystine at codon 33. The mutation point of CTNNB1 defer between the SPN And MCST (9). SPNs are driven by dysregulated Wnt/beta-catenin signalling and we can easily detect the resultant aberrant intracellular accumulation of beta-catenin in these tumours by immunohistochemistry. There is evidence that in many SPNs this is a consequence of a somatic point mutation in exon 3 of the *CTNNB1* gene.

Abnormal accumulation of beta-catenin protein caused by *APC* tumour suppressor gene mutations, is the causative molecular abnormality in FAP. It is therefore surprising that so few cases of ovarian SPNs have been previously reported in FAP patients. This may be because ovarian SPNs have previously been underdiagnosed given that they have only been recently described in the literature and that the majority have exhibited an indolent course (10).

SPNs are considered tumours of low malignant potential, but 15% can develop metastasis mostly involving the liver and peritoneum. Tang et al have reported two cases of clinically aggressive SPN of the pancreas where the patient died within months after the initial diagnosis. Based on these two cases, the author suggested that histological features might be associated with the aggressive behaviour including perineural and vascular invasion, invasion of surrounding organs, nuclear atypia, elevated mitotic rate and necrosis (11).

Conclusion

This case not only contributes to our knowledge of this uncommon entity but also provides further insight into its potential association with FAP. SPNs should be considered in the clinical and pathological differential diagnoses of tumours presenting in FAP patients.

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All the authors agree to take responsibility for every facet of the work, making sure that any concerns about its integrity or veracity are thoroughly examined and addressed.	