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Case Report

High-grade Peritoneal Pseudomyxoma of Primary Ovarian Origin Revealed by a Borderline Mucinous Tumor in a 26-year-old Woman: A Case Report and Literature Review

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Abstract: Pseudomyxoma peritonei (PMP) is a rare condition predominantly originating from appendiceal neoplasms, making true primary ovarian PMP an exceptional and widely debated clinical entity. Establishing a primary Müllerian origin requires exhaustive histological and immunohistochemical evaluation to exclude an occult appendiceal tumor. A highly unusual clinical scenario involves a 26-year-old nulliparous woman presenting with a large borderline mucinous ovarian tumor paradoxically associated with high-grade peritoneal implants. Comprehensive pathological workup (CK7+, PAX8+, CK20−, CDX2−) and completely normal appendiceal serial sections definitively confirmed the primary ovarian origin. Standard-of-care management relies on complete cytoreductive surgery (CRS) combined with hyperthermic intraperitoneal chemotherapy (HIPEC). Successful CC0 cytoreduction and HIPEC were achieved, integrated with proactive ovarian cortex cryopreservation. This unusual presentation highlights the striking biological discordance that can exist between a primary tumor and its peritoneal dissemination, emphasizing the critical role of specialized multidisciplinary management, rigorous pathology, and early oncofertility integration for young patients.

Keywords: Pseudomyxoma Peritonei, Primary Ovarian Origin, Borderline Mucinous Tumor, High Grade, Complete Cytoreductive Surgery.

INTRODUCTION

Peritoneal pseudomyxoma (PMP) is a rare condition characterized by the progressive accumulation of gelatinous mucin within the peritoneal cavity, associated with multifocal mucinous epithelial implants. Its incidence is estimated at 1 to 2 cases per million inhabitants per year. In more than 70 to 90% of cases, the origin is appendiceal, most often secondary to the rupture of a low-grade appendiceal mucinous neoplasm (LAMN), through a mechanism of peritoneal dissemination described by Sugarbaker [1].

Primary ovarian origin of PMP remains exceptional and controversial. Indeed, numerous studies have demonstrated that the majority of mucinous ovarian tumors associated with PMP actually correspond to metastases from an occult appendiceal tumor. Complete histological analysis of the appendix and immunohistochemical study (CK7, CK20, CDX2, SATB2, PAX8 profile) are therefore essential to establish the diagnosis of primary ovarian origin [2].

Histologically, the PSOGI group classification distinguishes between low-grade and high-grade forms, whose prognosis and therapeutic strategy differ significantly. Current management is based on complete cytoreductive surgery combined with hyperthermic intraperitoneal chemotherapy (CRS + HIPEC), allowing substantial improvement in survival.

We report the case of a 26-year-old patient presenting with a borderline mucinous ovarian tumor associated with high-grade peritoneal pseudomyxoma, whose primary ovarian origin was confirmed after complete histological and immunohistochemical analysis. This case is remarkable due to the rarity of ovarian origin and the unusual discordance between the borderline grade of the primary tumor and the high grade of the peritoneal implants.

CASE REPORT

Ms. G.M., aged 26 years, nulligravida nullipara (G0P0), with no notable personal or family oncological history, consulted for progressive abdominal distension evolving over approximately six months, associated with increasing pelvic pain and menometrorrhagia. Clinical examination revealed diffuse dullness on percussion, without collateral venous circulation, and vaginal examination revealed a painful left adnexal mass on mobilization. General condition was preserved.

Morphological assessment revealed, on transvaginal ultrasound, a retro-uterine mass with mixed liquid and solid components with endocystic vegetations richly vascularized on Doppler examination. Pelvic MRI confirmed a mass arising from the left ovary, measuring 200 × 130 × 90 mm, cystic, solid and hemorrhagic in nature, septated, with heterogeneous enhancement after gadolinium injection, highly suspicious for malignancy, associated with pelvic-peritoneal effusion of mucinous appearance in T1 and T2 hypersignal. Abdominopelvic CT scan completed the assessment by demonstrating septated ascites of high density (> 20 HU), hepatosplenic scalloping and central displacement of the small bowel loops, constituting signs suggestive of peritoneal pseudomyxoma. No deep lymphadenopathy or distant metastatic lesions were identified. Biologically, standard workup was unremarkable. Tumor marker assay revealed isolated elevation of CA125 at 97 IU/ml, while other markers CEA, AFP and CA 19-9 were strictly normal.

After discussion at a multidisciplinary tumor board (MTB), exploratory laparoscopy converted to midline laparotomy allowed left salpingo-oophorectomy with peritoneal cytology, peritoneal biopsies and mucin sampling. Intraoperative exploration revealed a large left ovarian mass measuring 20 × 14 × 8 cm weighing approximately 100 grams, with spontaneous capsular rupture releasing approximately 70 ml of abundant gelatinous mucinous material. The appendix and right ovary were macroscopically normal. Yellowish mucinous and inflammatory-appearing lesions were observed on the small bowel peritoneum and sigmoid, without associated occlusion or perforation.

Anatomopathological examination concluded a borderline mucinous tumor of the left ovary containing foci of carcinoma in situ without stromal invasion, estimated at 10% of the tumor surface, without vascular or lymphatic emboli, associated with high-grade peritoneal pseudomyxoma (HG-PMP) lesions in the peritoneal implants. Analysis of pelvic mucin revealed mucus with rare non-invasive tumor cells. Peritoneal biopsies demonstrated mucinous peritonitis without serosal invasion of the small bowel, and intraperitoneal cytology showed atypical glandular cells embedded in abundant mucinous material. Complete and exhaustive histological analysis of the appendix, performed in serial sections, revealed no epithelial abnormality, formally excluding any occult primary appendiceal mucinous tumor. Immunohistochemical workup showed positive expression of CK7 and PAX8, with complete negativity of CK20, CDX2 and SATB2, a profile compatible with a primary Müllerian ovarian origin.

After presentation at a specialized peritoneal surface malignancy tumor board, the patient underwent complete cytoreductive surgery combined with hyperthermic intraperitoneal chemotherapy (HIPEC), in accordance with international PSOGI group recommendations, performed via xipho-pubic midline laparotomy. Intraoperative exploration according to Sugarbaker's method established a PCI score of 14/39, indicating moderate tumor burden predominantly pelvic with associated mesenteric and small bowel implants (LS score between 1 and 2). The surgical procedure included right salpingo-oophorectomy, appendectomy, total omentectomy, targeted parietal and pelvic peritonectomies, electrofulguration of residual mesenteric lesions, as well as right ovarian cortex sampling for fertility preservation purposes. Complete cytoreduction status CC0 was achieved, corresponding to the absence of any macroscopically visible tumor residue. HIPEC was administered immediately after cytoreduction at a temperature of 41–42°C for 60 minutes, using the open "coliseum" technique, with mitomycin C (3.3 mg/m²) and cisplatin (25 mg/m²), according to the institutional protocol.

Definitive anatomopathological examination of cytoreduction specimens confirmed the absence of primary appendiceal mucinous tumor, with a histologically normal appendix. Analysis of the omentum, peritonectomies and mesenteric samples demonstrated high-grade mucinous deposits with marked nuclear atypia and increased mitotic activity, confirming the diagnosis of high-grade peritoneal pseudomyxoma of primary ovarian origin.

Postoperative course was uneventful, without major complications according to the Clavien-Dindo classification. Hormone replacement therapy was initiated due to bilateral surgical castration. At 6 months follow-up, the patient is in good general condition, without clinical or radiological signs of recurrence. Follow-up tumor markers (CA125, CEA, CA 19-9) are normalized. Close clinical and radiological follow-up has been established, including regular tumor marker assays and thoraco-abdominopelvic CT scan every six months for the first three years, then annually, as well as specialized oncofertility management for discussion of perspectives related to the harvested ovarian cortex. Although our patient's 6-

month follow-up is short given the often slow natural history of peritoneal pseudomyxoma, rigorous long-term follow-up will be crucial to confirm the effectiveness of this management.

DISCUSSION

1. Epidemiology and Definition

Peritoneal pseudomyxomas (PMP) are rare conditions characterized by the progressive accumulation of gelatinous-appearing mucin within the peritoneal cavity, lining all peritoneal surfaces, hence the historical term "gelatinous disease of the peritoneum." Their incidence is estimated at approximately 2 cases per 10,000 laparotomies, with a prevalence in the general population of approximately 1 to 2 cases per million inhabitants per year [1]. A clear female predominance is observed, with a sex ratio of approximately 1:2, although this predominance is partially related to the frequency of mucinous ovarian tumors in women [2].

Despite their macroscopically benign appearance in low-grade forms, PMPs are characterized by progressive local evolution, tendency to recurrence and risk of potentially fatal complications, justifying specialized management in expert centers [3].

2. Etiopathogenesis

The etiopathogenesis of PMP most often involves rupture of a mucinous tumor into the peritoneal cavity, with secondary dissemination of mucinous epithelial cells onto peritoneal surfaces. The vast majority of cases, estimated between 70 and 90% according to series, are of appendiceal origin, secondary to a low-grade appendiceal mucinous neoplasm (LAMN: Low-grade Appendiceal Mucinous Neoplasm) or, more rarely, high-grade (HAMN) [4, 3].

Other primary locations, much rarer, have been reported in the literature, notably:

- Ovarian: representing approximately 5 to 10% of cases, although most mucinous ovarian tumors associated with PMP are actually metastases from an occult primary appendiceal tumor [5].
- Colorectal
- Biliary and gallbladder
- Pancreatic
- Urothelial (exceptional cases)

The question of ovarian versus appendiceal origin is fundamental and the subject of numerous debates in the literature. Indeed, several studies have demonstrated that up to 70 to 80% of mucinous ovarian tumors associated with PMP present [6], on immunohistochemical analysis, a profile compatible with appendiceal or colorectal origin (CK7−/CK20+/CDX2+/SATB2+) [7], arguing more for secondary ovarian involvement than for true ovarian primary [8]. Thus, even in the presence of a bulky mucinous ovarian tumor, occult appendiceal origin must be systematically sought by complete and meticulous histological analysis of the appendix, as well as by exhaustive immunohistochemical workup [9, 10].

3. Anatomopathology and Classification

Anatomopathologically, PMP corresponds to diffuse peritoneal involvement combining mucinous ascites and multifocal mucinous epithelial implants. The fundamental histological element is the presence of extracellular mucin, containing or not mucinous epithelial cells whose degree of atypia and differentiation conditions biological behavior and prognosis [13].

Histological classification has evolved over time. The international PSOGI group (Peritoneal Surface Oncology Group International) has proposed a consensus classification mainly distinguishing [14, 15]:

- **Low-grade PMP**: characterized by well-differentiated mucinous cells, minimally atypical, embedded in abundant extracellular mucin, with low mitotic activity. This subtype is associated with favorable prognosis, with 5-year survival rates exceeding 75 to 85% after optimal treatment.
- **High-grade PMP**: characterized by higher cell density, marked nuclear atypia, increased mitotic activity and more complex architecture. Prognosis is significantly less favorable, with 5-year survival estimated between 40 and 60% according to series.
- **PMP with signet ring cells**: rare subtype but with markedly poor prognosis, with 5-year survival below 30%, due to marked biological aggressiveness and relative resistance to conventional treatments.

This histological stratification is essential as it directly conditions therapeutic strategy and postoperative surveillance.

4. Clinical Presentation

Clinically, PMP symptomatology is long non-specific, which explains the frequently observed diagnostic delay, sometimes estimated at several years. The most frequently reported signs are:

- Progressive abdominal distension, often described by the patient as weight gain or bloating sensation [13]
- Abdominal pain of variable intensity, often diffuse
- Deterioration of general condition at advanced stage
- Bowel disorders (constipation, diarrhea or alternation)
- Umbilical hernia related to increased intra-abdominal pressure

In women, discovery is sometimes incidental during pelvic ultrasound performed for an adnexal mass. In some cases, the tumor is discovered incidentally during appendectomy or laparoscopy for another indication [14].

At advanced stage, serious complications may occur, notably:

- Intestinal obstruction by compression or infiltration of small bowel loops, constituting a poor prognostic factor requiring urgent surgical management
- Urinary tract compression causing hydronephrosis
- Severe malnutrition due to malabsorption [15].

5. Radiological Assessment

Imaging plays a fundamental role in positive diagnosis, extension assessment and surgical planning.

Abdominal ultrasound is often the first examination performed. It may reveal heterogeneous hypoechoic collections, septated, sometimes with hyperechoic spots corresponding to mucinous deposits. However, its sensitivity remains limited for precise extension assessment [16].

Abdominopelvic computed tomography (CT) with contrast injection represents the reference examination. It allows visualization of:

- Septated mucinous ascites, of higher density than simple liquid ascites (> 20 HU)
- Multilocular thin-walled masses, sometimes calcified, arranged peripherally in the abdominal cavity
- Hepatic and splenic scalloping, pathognomonic of PMP, corresponding to contour indentations related to compression by mucinous masses
- Characteristic omental cake
- Central displacement of small bowel loops, sign of diffuse peritoneal dissemination
- Peritoneal calcifications in some advanced forms [17].

MRI advantageously complements the assessment, particularly in case of diagnostic doubt or for better lesion characterization. It offers superior contrast resolution to CT, allowing more precise analysis of extension to peritoneal surfaces, mesentery and adjacent structures. It is particularly useful for assessing small bowel involvement and lesion resectability [18, 19].

Quantification of peritoneal extension is based on the Peritoneal Cancer Index (PCI), validated score from 0 to 39, assessing tumor burden in 13 abdominopelvic regions. This score conditions resectability and represents a major prognostic factor: high PCI (> 20) is associated with incomplete resection and significantly reduced survival [20].

6. Biological Assessment and Tumor Markers

Assessment of biological aggressiveness is partly based on serum tumor marker assay, notably:

- **CEA (Carcinoembryonic Antigen)**: elevated in advanced forms and high-grade histological subtypes
- **CA 19-9**: particularly relevant in mucinous tumors of digestive origin
- **CA 125**: useful in forms of ovarian origin or in case of diffuse peritoneal involvement

Elevation of at least one of these markers is found in more than 80% of patients with PMP, regardless of histological grade [20]. These markers are correlated with tumor burden, resectability and overall survival. They also constitute essential tools for postoperative follow-up, allowing early detection of progressive recurrence even before clinical or radiological expression [21].

7. Treatment

7.1. General Principles

PMP management was revolutionized by the combined approach initially proposed by Sugarbaker, combining complete cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC). This strategy currently constitutes the standard treatment recommended by international learned societies, notably PSOGI [12].

7.2. Cytoreductive Surgery

The objective of cytoreduction is to obtain macroscopically complete resection, defined by CC0 score (Completeness of Cytoreduction), corresponding to the absence of visible tumor residue. This CC0 status represents the most important perioperative prognostic factor, with significantly superior long-term survival compared to incomplete resections (CC1, CC2 or CC3) [22].

Cytoreduction generally includes:

- Parietal and visceral peritonectomies (pelvic, diaphragmatic, parietal peritoneum)
- Total omentectomy
- Systematic appendectomy (if not previously performed)
- Adapted visceral resections according to extension: right colectomy, sigmoidectomy, hysterectomy with bilateral salpingo-oophorectomy, splenectomy, partial hepatic resections or even cholecystectomy[23, 24]

The morbidity of this surgery is significant, with major complication rates (Clavien-Dindo grade III–IV) estimated between 20 and 40% in expert centers, and perioperative mortality below 2% in high-volume centers [25].

7.3. Hyperthermic Intraperitoneal Chemotherapy (HIPEC)

Immediately after cytoreduction, HIPEC is administered directly into the peritoneal cavity at a temperature of 41–42°C for 60 to 90 minutes, according to institutional protocols. Heat potentiates the cytotoxic effect of chemotherapeutic agents through several mechanisms: increased tissue penetration, inhibition of DNA repair mechanisms and direct cytotoxic effect on tumor cells.

First-line agents used in PMP are:

- **Mitomycin C**: alkylating agent, most used, particularly in North American and Australian protocols
- **Cisplatin or Oxaliplatin**: used alternatively or in combination, particularly in European protocols

HIPEC targets microscopic or millimetric tumor residues remaining after cytoreduction, inaccessible to surgery [25].

7.4. Strategy According to Tumor Grade

For Low-Grade Forms:

Complete cytoreduction combined with HIPEC is generally sufficient, without additional systemic chemotherapy. Long-term results are favorable, with 5-year survival rates exceeding 75 to 85% in expert centers [26].

For High-Grade Forms:

A more aggressive approach is necessary. In case of resectable disease, CRS + HIPEC remains the option of choice. However, in case of high tumor burden or borderline resectable disease, neoadjuvant systemic chemotherapy may be discussed, such as:

- FOLFOX (5-fluorouracil + leucovorin + oxaliplatin)
- CAPOX (capecitabine + oxaliplatin)
- With or without bevacizumab (anti-VEGF) in the absence of contraindication [27].

In situations of unresectable or distant metastatic disease, palliative systemic chemotherapy may provide benefit in terms of disease control, although the optimal protocol remains to be defined by prospective studies [28].

8. Surveillance and Recurrence

Peritoneal recurrences remain frequent in PMP, even after apparently complete cytoreduction, due to the often diffuse and multifocal nature of the disease. They may manifest clinically by progressive abdominal distension, occlusive syndrome or elevation of tumor markers, sometimes several years after initial treatment.

Surveillance is adapted to histological grade:

For Low-Grade Forms:

- Abdominopelvic CT scan and tumor marker assay (CEA, CA 19-9, CA 125) annually during the first 6 years, then spaced according to evolution [29].

For High-Grade Forms:

- Thoraco-abdominopelvic CT scan every 6 months during the first 3 years, then annual [30].
- Tumor marker assay at each consultation

In case of resectable recurrence, surgical reoperation such as debulking or new complete cytoreduction may be considered in selected cases, ideally in an expert center [31].

9. Particularity of Our Observation

In our observation, the occurrence of high-grade peritoneal pseudomyxoma in a young patient presenting with a borderline mucinous ovarian tumor constitutes an unusual and relatively rare clinical presentation in the literature. Several points deserve to be highlighted:

Regarding Tumor Origin:

Primary ovarian origin of PMP remains exceptional. As previously mentioned, the majority of apparently ovarian-origin PMPs are actually peritoneal metastases from an occult appendiceal tumor [32]. In our case, complete histological analysis of the appendix and thorough immunohistochemical workup were performed to formally eliminate primary appendiceal origin. Immunohistochemical results, combined with morphological data, oriented toward possible ovarian origin, although this diagnosis remains difficult to assert with certainty.

Regarding Histological Grade:

The presence of high-grade PMP associated with ovarian borderline tumor is particularly unusual. The discordance between the grade of the primary tumor (borderline, therefore low-grade) and the grade of PMP (high-grade) raises the question of transformation or tumor progression at the peritoneal implant level, a phenomenon described but rare in the literature [33].

Regarding Treatment:

Management by complete cytoreduction combined with HIPEC fully aligns with current recommendations for high-grade forms [34]. The decision to perform complete cytoreductive surgery in this young patient was essential, given its demonstrated prognostic impact.

This case illustrates the diagnostic and therapeutic complexity of PMP, highlighting the importance of multidisciplinary management in specialized centers with expertise in peritoneal surgery and oncology.

10. Oncofertility and Quality of Life

Complete cytoreductive surgery for peritoneal pseudomyxoma frequently involves bilateral salpingo-oophorectomy, resulting in definitive surgical castration and early iatrogenic menopause whose long-term consequences are well documented: increased risk of osteoporosis, cardiovascular diseases, cognitive decline and overall impairment of quality of life [37]. This issue is particularly concerning in young women of childbearing age, particularly in ovarian-origin forms of peritoneal pseudomyxoma.

ASCO recommendations, updated in 2025, stipulate that fertility preservation must be systematically addressed from diagnostic announcement with any patient likely to receive gonadotoxic treatment, regardless of tumor type and prognosis [38]. Among available techniques, cryopreservation of mature oocytes after ovarian stimulation represents the most established method, but requires a two to three week delay sometimes incompatible with oncological urgency. Ovarian cortex cryopreservation constitutes an alternative particularly adapted to surgical emergency situations, with ovarian function recovery rates exceeding 95% after transplantation and more than 200 live births documented worldwide [39]. In the specific context of peritoneal pseudomyxoma, Thomakos *et al*., reported 14 successful pregnancies after CRS and HIPEC, with an average interval of approximately 30 months between surgery and conception, confirming the feasibility of parental project in selected patients [40].

Furthermore, initiation of hormone replacement therapy is recommended in any woman who has undergone bilateral salpingo-oophorectomy before natural menopause age, to prevent deleterious consequences of prolonged estrogen deficiency, and should be continued until the theoretical age of physiological menopause [41]. These considerations highlight the importance of a multidisciplinary approach integrating reproductive medicine and gynecological endocrinology alongside peritoneal surgery and oncology from the initial phase of management.

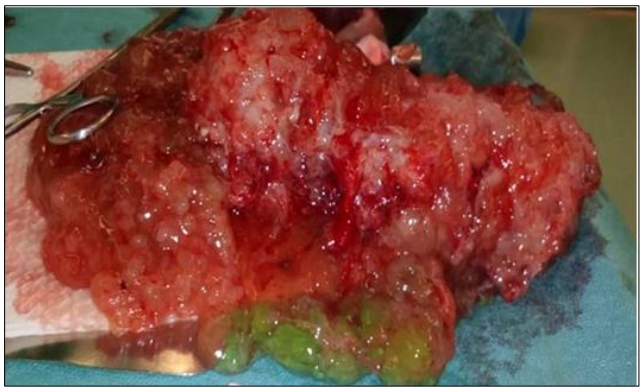

Figure 1: Surgical specimen of peritoneal pseudomyxoma with diffuse gelatinous deposits

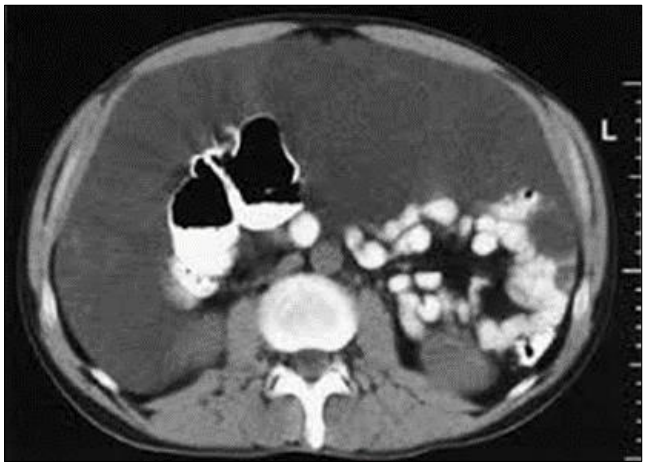

Figure 2: Pseudomyxoma peritonei. Intraperitoneal mucinous collections resulting in hepatic and splenic scalloping

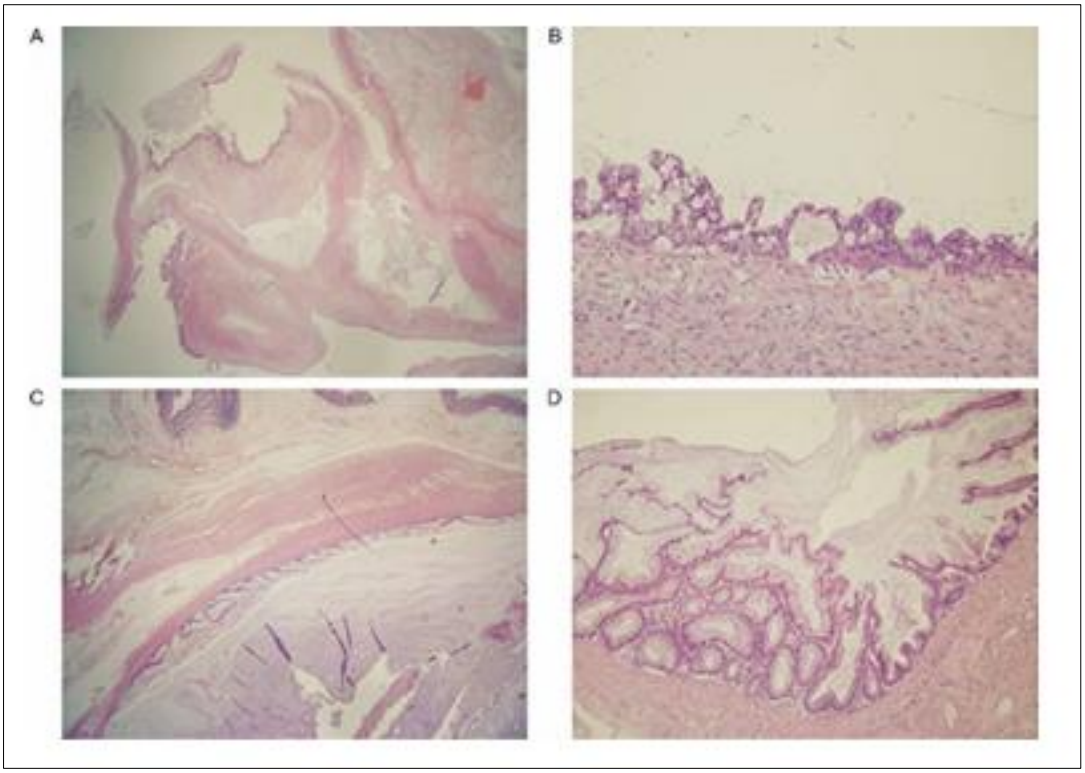

Figure 3: (A) Image showing the multicystic nature of the ovarian tumor, with mucinous epithelium and mucin accumulation within the wall. (B) The mucinous epithelial layer exhibited nuclear enlargement, stratification, and complex architecture. (C) The ovarian mucinous tumor contained large mucin pools, and (D) its lining epithelium displayed low-grade dysplasia without infiltrative growth. All images were stained with hematoxylin and eosin (H&E, original magnification ×100).

CONCLUSION

Pseudomyxoma peritonei (PMP) is a rare condition characterized by progressive accumulation of gelatinous mucin in the peritoneal cavity, primarily of appendiceal origin in 70 to 90% of cases. Primary ovarian origin remains exceptional and controversial, as the majority of ovarian involvement corresponds to metastases from an occult appendiceal tumor.

We report the case of a 26-year-old nulliparous patient presenting with progressive abdominal distension and pelvic pain evolving over six months. Morphological assessment revealed a large 200 mm left ovarian mass associated with mucinous peritoneal effusion suggestive of pseudomyxoma peritonei. Initial surgical exploration identified a 20 cm ovarian mass with spontaneous capsular rupture and abundant mucinous ascites. Anatomopathological examination concluded a borderline mucinous tumor of the left ovary associated with high-grade pseudomyxoma peritonei lesions in the peritoneal implants. Complete histological analysis of the appendix in serial sections revealed no abnormalities, formally excluding any appendiceal origin. The immunohistochemical profile (CK7+, PAX8+, CK20−, CDX2−, SATB2−) confirmed a primary müllerian ovarian origin.

Following discussion in a specialized multidisciplinary tumor board, the patient underwent complete cytoreductive surgery combined with hyperthermic intraperitoneal chemotherapy (CRS-HIPEC) according to PSOGI group recommendations. Intraoperative exploration established a PCI score of 14/39. The surgical procedure included bilateral salpingo-oophorectomy, appendectomy, total omentectomy, and targeted peritonectomies, achieving complete cytoreduction status CC0. HIPEC was administered using the coliseum technique at 41-42°C for 60 minutes with mitomycin C and cisplatin. A right ovarian cortex biopsy was performed for fertility preservation purposes. The postoperative course was uneventful. At 6 months follow-up, the patient remains asymptomatic without clinical, biological, or radiological signs of recurrence.

This case illustrates the rarity of primary ovarian pseudomyxoma peritonei, whose diagnosis requires complete histological analysis of the appendix and comprehensive immunohistochemical workup. The major distinctive feature lies in the unusual discordance between the borderline grade of the primary ovarian tumor and the high grade of the peritoneal implants, suggesting tumor progression within the peritoneal microenvironment. This observation underscores the importance of multidisciplinary management in an expert peritoneal surgery center, combining complete cytoreduction and hyperthermic intraperitoneal chemotherapy, the only therapeutic strategy that can significantly improve prognosis. It also emphasizes the necessity of early integration of oncofertility considerations in young patients, with ovarian cortex preservation constituting a realistic option for maintaining reproductive potential after treatment.

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