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A Case of Diagnosis of Pseudomyxoma Peritonei

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Abstract

Objective: To report a case of pseudomyxoma peritonei accidentally discovered during preoperative examination and review relevant literature to improve the early diagnosis of the origin of pseudomyxoma peritonei. Methods: To analyze a case accidentally discovered due to abdominal distension in our hospital in October 2023. Combined with domestic and foreign literature, the etiology, clinical manifestations, diagnosis, differential diagnosis and treatment of PMP were discussed. Results: The patient was admitted due to abdominal distension with no other specific discomfort. Mucinous tumors were found during imaging examinations. Conclusion: PMP is a rare disease, and clinical manifestations often include abdominal distension. It is usually first discovered through imaging examinations such as ultrasound, CT or MRI. CRS and HIPEC are still recommended as the standard treatment for PMP.

Keywords

Pseudomyxoma Peritonei, Low-Grade Mucinous Tumor

1. Introduction

Pseudomyxoma Peritonei is a relatively rare abdominal disease. The notable characteristic of pseudomyxoma peritonei is that the abdominal cavity is filled with a large amount of jelly-like mucus, and tumor cell clusters can be seen floating in the mucus. There are scallop-shaped indentations around the compressed viscera [1]. Ultrasound presents a clear image of ascites and indentations, yet there are few cases of pseudomyxoma peritonei initially diagnosed by ultrasound. Ultrasound can preliminarily determine the source and nature of larger pelvic masses and provide a basis for the formulation of clinical treatment plans. Previous studies have mainly focused on the pathogenesis and treatment strategies of

pseudomyxoma peritonei. Due to the similarity of the ultrasound manifestations of pseudomyxoma peritonei with other abdominal diseases, it is often misdiagnosed. However, cases diagnosed with pseudomyxoma peritonei are relatively rare. We present a clinically incidentally discovered case of pseudomyxoma peritonei and discuss its differential diagnosis.

2. Case Presentation

A 63-year-old female patient was admitted to the hospital mainly due to abdominal distension for more than ten months and the discovery of a pelvic mass for one day. The patient developed abdominal distension and poor appetite without obvious incentives ten months ago, without nausea, vomiting, abdominal pain, diarrhea, fever, or night sweats. She lost more than 10 catties in weight. The patient's previous constitution was average. She underwent appendectomy and left adnexectomy in 1993 and right adnexectomy + hysterectomy in 2019. The pathological results indicated: Mature teratoma of the right adnex with some components of low-grade mucinous tumor. In July 2019, B-ultrasound examination suggested a pelvic 30 × 34 × 21 mm hypoechoic mass. In August 2019, B-ultrasound examination showed no obvious mass. In 2020, B-ultrasound examination suggested a cystic mass in the right adnex area, approximately 32 × 29 × 17 mm. The patient denied a history of hypertension, diabetes, coronary heart disease, etc. There was no history of drug or food allergies. The patient was conscious, autonomous in position, and cooperated well. The skin and mucous membranes showed no signs of jaundice, and no superficial lymph nodes were touched. There was no vascular murmur in the bilateral carotid arteries, the lung breathing sounds were clear, and there were no obvious dry or wet rales. The heart size was normal, the rhythm was regular, and there was no murmur. Abdominal examination showed significant abdominal bulging, in the shape of a frog's abdomen, positive shifting dullness, and weakened bowel sounds. A mass was touched on the right side of the pelvic cavity during palpation, with poor mobility and no tenderness.

At 8:38 on October 28, 2023, abdominal, urinary, and gynecological outpatient ultrasound examination showed: Sonographic image after hysterectomy. A cystic mass of approximately 67 × 37 × 39 mm on the right side of the pelvic cavity: Originating from ovarian mucinous tumor? Others? A large number of dark areas in the abdominal cavity on sonographic image: Consider: Jelly-like effusion, rupture of ovarian mucinous tumor, Others (**Figure 1**). At 11:27 on October 28, 2023, gynecological vaginal ultrasound examination showed: The uterus has been removed (patient's self-report). A low and no-echoic mass of approximately 129 × 50 × 60 mm was detected in the pelvic cavity, with a relatively clear boundary, an irregular shape, a honeycomb-like interior, and CDFI: No obvious blood flow signal was seen. The mass in the pelvic cavity: Originating from ovarian mucinous tumor? Pseudomyxoma peritonei (PMP) in the abdominal cavity? Others? Further examination is recommended (**Figure 2**). Lower abdominal MRI plain scan + enhancement + functional imaging examination showed: Cystic space-occupying

lesion in the pelvic cavity, considering mucinous tumor, the nature is undetermined, and the possibility of malignancy is not excluded; abdominal and pelvic effusion, thickening of the peritoneum and multiple nodules, peritoneal metastasis? Multiple small lymph nodes in the bilateral groin, considered mostly benign lymph nodes (Figures 3-6). Laboratory tests: No obvious abnormalities were found in blood routine, liver and kidney function, electrolytes, coagulation function, etc. The tumor marker CEA was 13.64 ng/ml (normal reference value < 10.0 ng/ml), and CA125, AFP, and CA199 were normal.

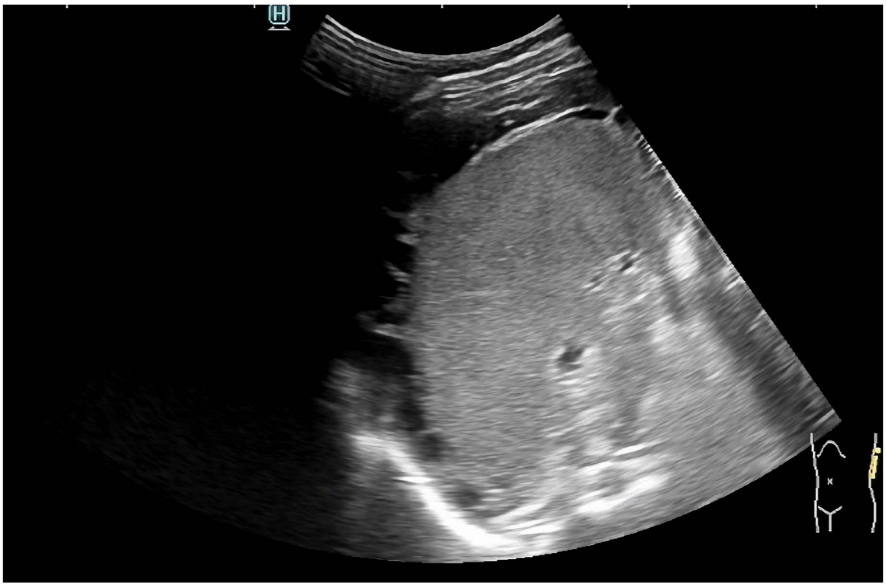

Figure 1. Ultrasound image: The peritoneal colloid-like effusion compresses the spleen periphery and presents a scallop-like indentation.

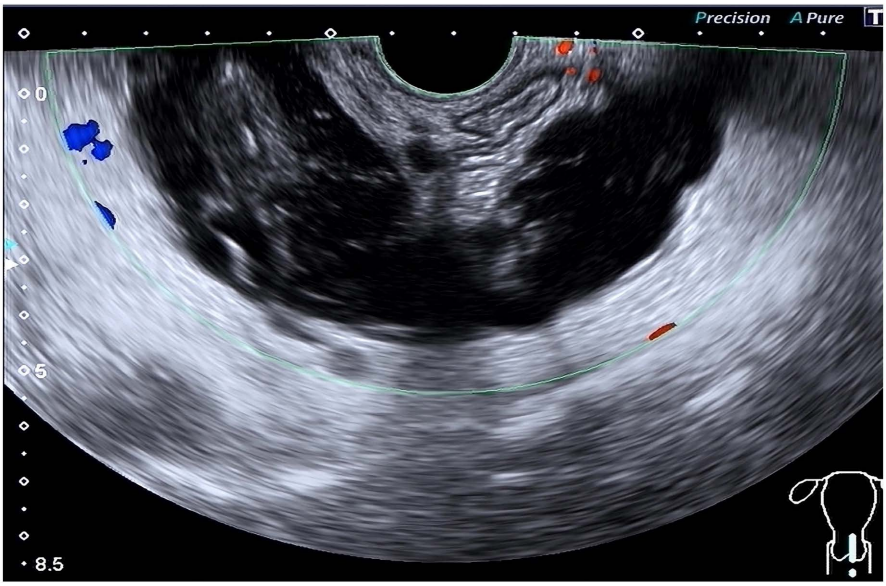

Figure 2. Color ultrasound image: The pelvic colloid-like effusion shows multilocular changes, and no obvious blood flow signals are detected inside.

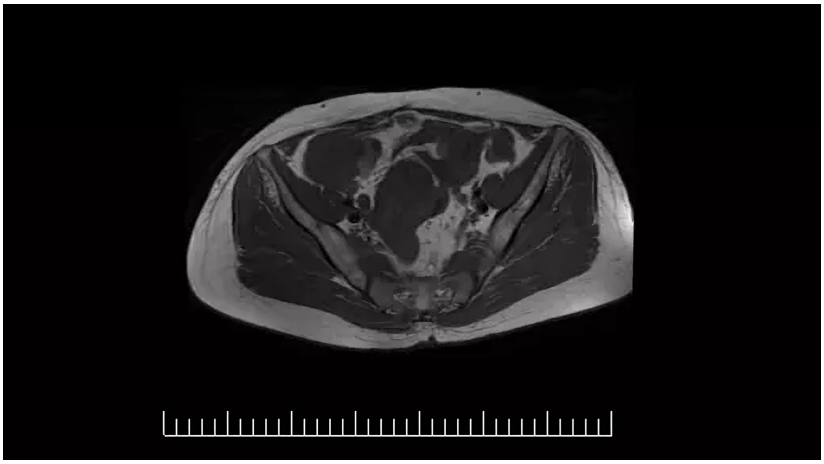

Figure 3. MRI Ax T1 FSE image: Strip-shaped high-signal shadows can be seen in the low-signal lesion, presenting a multilocular structure.

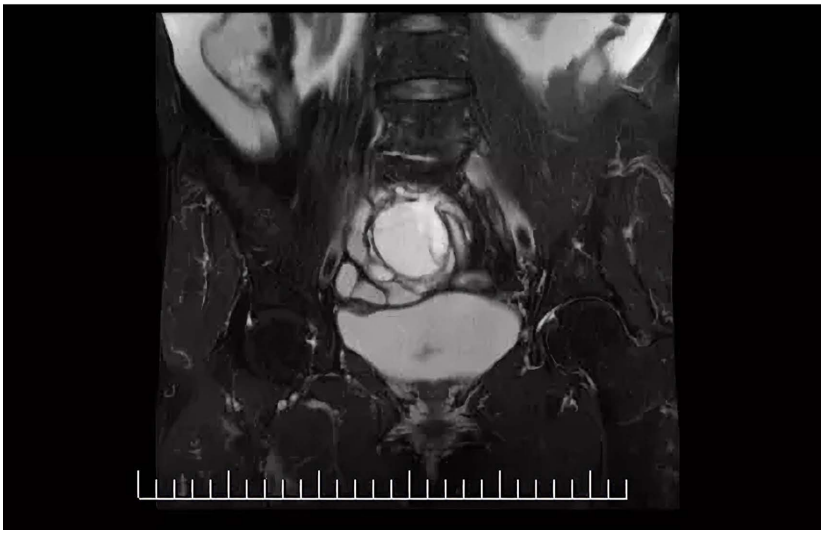

Figure 4. MRI OCor T2 fs FSE image: High-signal shadows can be seen in the abdominal and pelvic cavities.

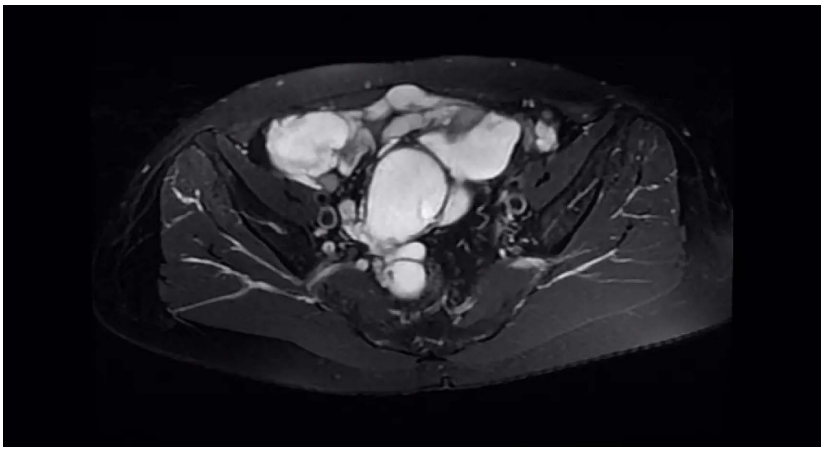

Figure 5. MRI Ax T2 FSE image: Small nodular and small strip-shaped low-signal shadows can be seen in the high-signal shadow.

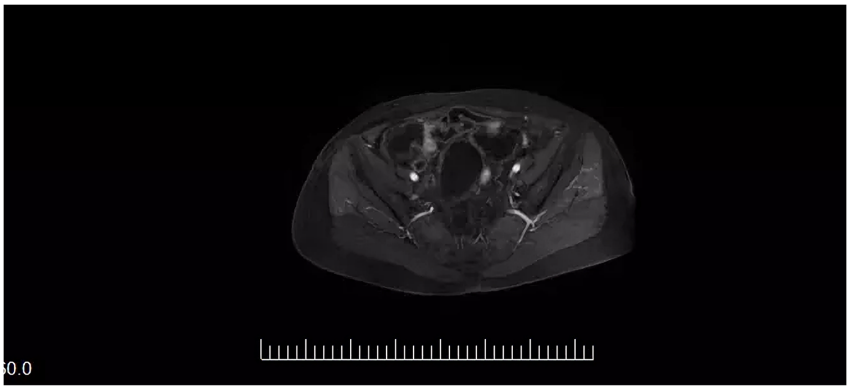

Figure 6. MRI Ax LAVA+C image: The small nodular and small strip-shaped low-signal shadows are enhanced, and the diffusely thickened peritoneum is enhanced.

Laparoscopy was performed to take a large yellow-white tissue on the peritoneum above the left sacroiliac joint for rapid pathological examination, and the report was: Low-grade mucinous tumor. Postoperative pathological examination showed: (Abdominal mass) The submitted tissue was a large amount of mucus and a small amount of mucous glandular epithelial cells, with mild atypia of the cells, consistent with low-grade mucinous tumor (Figure 7). Immunohistochemistry: CK7 (weak +), CDX2 (+) (Figure 8), CK20 (+) (Figure 9), ER (-) (Figure 10), KI-67 (about 50% +) (Figure 11), PR (-) , P53 (-), WT-1 (+). Based on various examinations, the diagnosis was pseudomyxoma peritonei.

The patient underwent laparoscopic peritoneal lesion resection + pelvic adhesion lysis + intestinal adhesion lysis under general anesthesia on November 3, 2023, and the tumor tissues visible to the naked eye were removed as much as possible. After the operation, chemotherapy was performed.

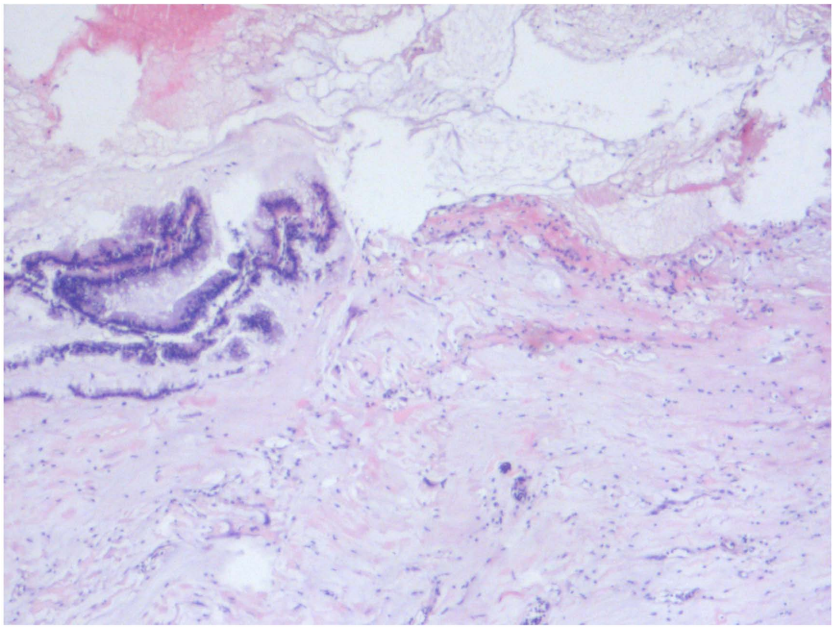

Figure 7. Pathological image.

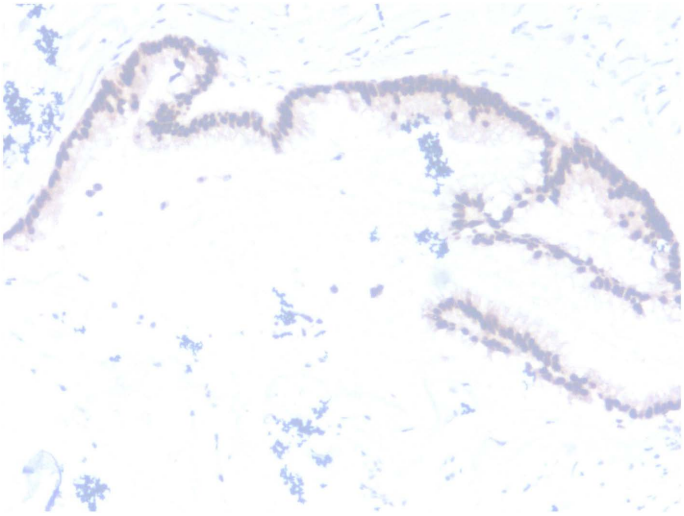

Figure 8. Immunohistochemical image: CDX2.

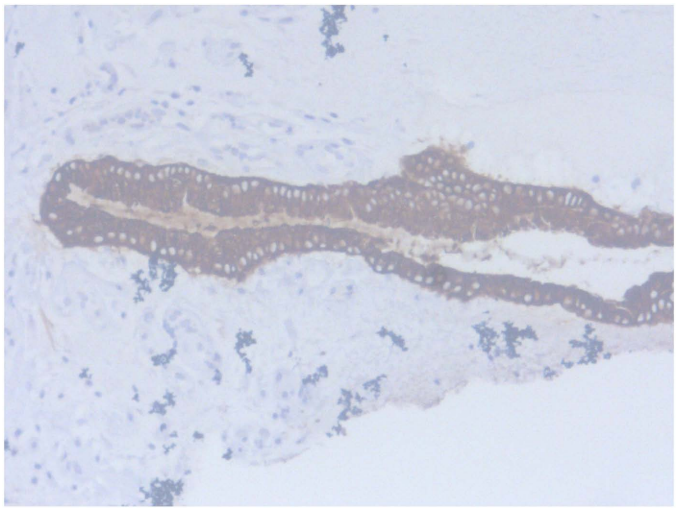

Figure 9. Immunohistochemical image: CK20.

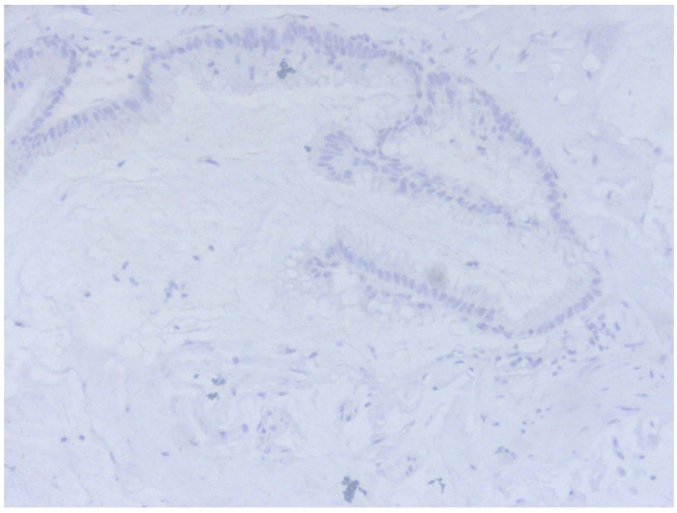

Figure 10. Immunohistochemical image: ER.

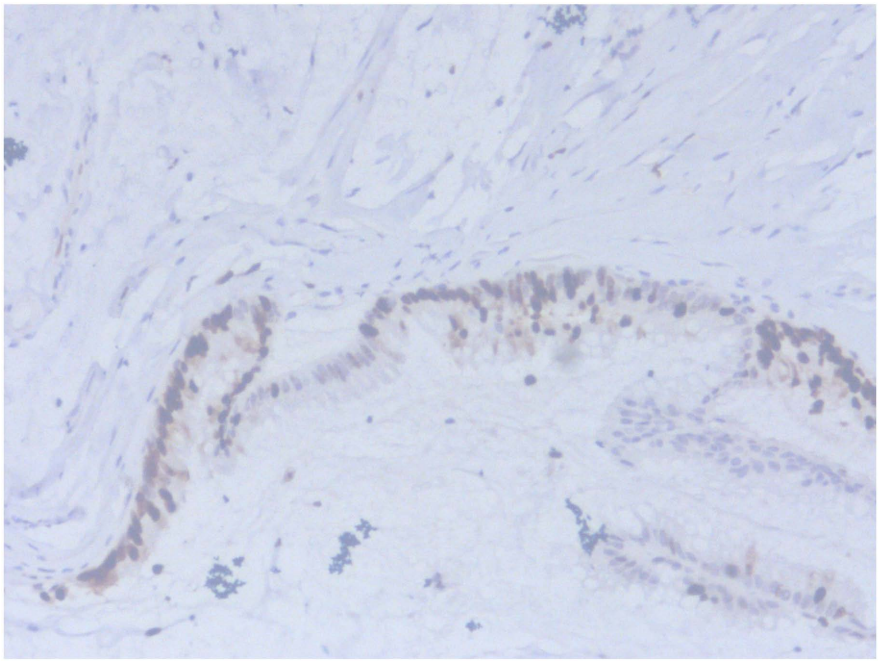

Figure 11. Immunohistochemical image: Ki-67.

Regular reexaminations were conducted after discharge. Abdominal ultrasound was reexamined at 1 month, 2 months, 3 months, and 4 months after the operation for recurrent ascites. The tumor marker CEA gradually returned to normal. The patient is still under follow-up at present.

3. Discussion

The pathogenesis of pseudomyxoma peritonei is still unclear. Currently, it is believed that it may be related to the rupture of appendiceal mucinous tumors, ovarian mucinous tumors, or other intra-abdominal mucinous tumors, releasing tumor cells into the abdominal cavity [2]. The mucus secreted by tumor cells accumulates in the abdominal cavity, forming a jelly-like substance, leading to compression and dysfunction of abdominal organs. Among adults, its incidence rate is relatively low.

PMP has uniqueness in aspects such as symptom manifestations and individual factors. The clinical manifestations of pseudomyxoma peritonei are non-specific, mainly manifested as gradual abdominal distension, abdominal bloating, abdominal pain, abdominal masses, ascites, etc [3]. Some patients may be accompanied by nausea, vomiting, loss of appetite, weight loss and other different symptoms. Due to the atypical symptoms, it is easily misdiagnosed as liver cirrhosis ascites, ovarian cancer, gastrointestinal tumors and other diseases. The location and growth rate of tumors in the pelvic and abdominal cavities are different, the invasiveness and differentiation degree of tumor cells are different, and the basic conditions, psychological and social factors of patients are different. However, there are certain commonalities in the pathophysiological mechanism, imaging features and treatment direction of the disease. For example, the secretion

function of tumor cells will lead to abnormal accumulation of mucus. The imaging manifestations are cystic or solid masses, ascites and peritoneal thickening, resulting in symptoms such as abdominal distension and abdominal distention. Most PMP cases originate from the appendix, ovaries or other gastrointestinal mucinous tumors. Surgery combined with hyperthermic intraperitoneal chemotherapy is the main treatment method. This patient has a previous history of appendectomy, oophorectomy, bilateral adnexectomy and pathological indications of adnexal mature teratoma and some components of low-grade mucinous tumors, undoubtedly casting a mysterious veil over the diagnosis of this patient. Understanding these uniquenesses and commonalities helps with earlier suggestive diagnoses and formulates personalized treatment plans for each patient to improve the treatment effect.

The diagnosis of pseudomyxoma peritonei mainly relies on clinical manifestations, imaging examinations, laparoscopy, laboratory tests and pathological examinations. Ultrasound, CT, and MRI examinations are commonly used imaging methods. In terms of diagnosis, ultrasound can clarify the involved area and scope of the lesion. The typical ultrasound manifestation of PMP is a large sheet-like anechoic area in the abdominal and pelvic cavity, in which a large number of disordered and irregular medium and low echoes and fine septa can be seen, and the mobility in ascites is poor, without obvious floating sensation; at the same time, diffuse cake-shaped thickening of the greater omentum in the abdominal cavity may be found, with a reticular structure, unclear boundaries, and some adhesions to the posterior intestinal tubes; changes in compression of parenchymal organs such as the liver, spleen, and kidneys, and serrated or scallop-like impressions can be seen on the surface [1]. Through these characteristic ultrasound manifestations, doctors can make a preliminary diagnosis of PMP and differentiate it from other similar diseases, such as peritoneal mesothelioma, tuberculous peritonitis, metastatic abdominal tumors, and ovarian cysts. CT examination can show the size, shape, location of the tumor and its relationship with surrounding tissues more clearly, and has advantages in evaluating the anatomical structure and the overall condition of the lesion in the abdominal cavity. MRI is superior to ultrasound in showing the contrast of soft tissues, and is clearer in distinguishing the boundary between the tumor and normal tissues, especially having high accuracy in judging whether the tumor invades the subperitoneal layer and whether it contains mucin, etc. [4]. Laparoscopy can directly observe the lesions in the abdominal cavity and perform biopsy, which is of high value for a clear diagnosis, such as visually seeing the mucinous nodules and masses on the peritoneum. Laboratory tests such as tumor markers CEA, CA125, etc. may have a mild increase. The final diagnosis requires pathological examination. The patient's CK20 (+) and CDX2 (+) suggest that the tumor may have a tendency of intestinal origin. WT-1 (+) can be positive in ovarian tumors, but it is not specific. CK7 (weak +) may be expressed in various tumors. KI-67 (about 50% +) indicates that the tumor cells proliferate more actively. ER (-) and PR (-) are insufficient supporting evidence for judging ovarian origin. CEA (+) can be positive in various adenocarcinomas.

Overall, these immunohistochemical results tend to suggest that the tumor may originate from the intestine, and the possibility of the appendix is relatively large, but the possibility of ovarian origin cannot be completely ruled out. Low-grade PMP is more likely to have reticular anechoic areas, jelly-like ascites and "fan-shaped impressions on the liver edge" than high-grade PMP, while peritoneal nodules and smaller single anechoic areas can be seen in high-grade PMP [5].

The significance of the multidisciplinary approach in diagnosing PMP is evident. Firstly, it improves diagnostic accuracy. By integrating the professional knowledge and techniques of multiple disciplines such as imaging, pathology, oncology, and gastrointestinal surgery, the patient's condition can be evaluated from different perspectives, reducing the possibility of misdiagnosis. Secondly, it formulates personalized diagnostic plans. The multidisciplinary team can establish the most suitable diagnostic processes and methods based on the specific circumstances of the patient. Thirdly, it promotes early diagnosis. Fourthly, it provides a basis for treatment. Accurate diagnosis is helpful in providing a basis for subsequent treatment options, including surgical methods, chemotherapy regimens, and the timing of radiotherapy and chemotherapy. In conclusion, diagnosing PMP faces many challenges. The adoption of the multidisciplinary approach can fully leverage the advantages of each discipline, enhance the accuracy and timeliness of diagnosis, and bring positive impacts on the treatment and prognosis of patients.

Pseudomyxoma peritonei often needs to be differentiated from liver cirrhosis ascites, tuberculous peritonitis, ovarian cysts, etc. 1) Ascites from liver cirrhosis is mostly light yellow and clear liquid, without septa and flocculent echoes, and patients often have a history of liver cirrhosis. 2) Tuberculous peritonitis often has systemic symptoms such as low fever and night sweats. Ascites is mostly straw-yellow and mycobacterium tuberculosis can be found. 3) Ovarian cysts are mostly unilateral, with clear boundaries, thin and smooth cyst walls.

4. Conclusion

In conclusion, pseudomyxoma peritonei is a rare abdominal tumor. It is often discovered incidentally through imaging examinations and generally has no specific clinical manifestations. Only by improving the understanding of the pathogenesis of pseudomyxoma peritonei, improving the early diagnosis of appendiceal, ovarian and gastrointestinal mucinous tumors, CRS and HIPEC, can the quality of life and survival of patients be improved. Ultrasound examination plays an important role in the diagnosis of pseudomyxoma peritonei, but it needs to be combined with other examinations and clinical manifestations for comprehensive judgment to avoid misdiagnosis. This depends on the doctor's understanding of pseudomyxoma peritonei and rich experience in ultrasound diagnosis. However, there are still three specific challenges in diagnosing PMP. First, the non-specificity of PMP symptoms. Ovarian cancer, colon cancer, peritonitis, etc. can all present as abdominal distension, abdominal pain, weight loss, etc. Second, the

limitations of imaging examinations. The small mucinous masses formed by PMP are similar to tumors and are easily misdiagnosed as normal abdominal structures. Finally, the complexity of pathological diagnosis. Obtaining sufficient and representative pathological tissue samples is sometimes challenging. Moreover, the subtypes of PMP need to be differentiated in pathological diagnosis, which requires accurate judgment by experienced pathologists.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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