

## Case Report

**Pseudomyxoma Peritonei**

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**ABSTRACT** The authors report a case of a 60 years old Romanian male with peritoneal pseudomyxoma that was discovered to have a primary location in the greater omentum. Pseudomyxoma peritonei arising into the greater omentum is very rare. It is an original case report and the new data represents a well-balanced summary of a timely subject, with reference to the literature. This it is a significantly advance in our understanding of a particular disease etiology. Until now, only a few primary greater omentum pseudomyxoma peritonei have been described. Clinical and pathologic findings of this case are presented and the topic of primary location in the great omentum of pseudomyxoma peritonei was reviewed. Preoperative ultrasound examination showed the presence of an "eyes of net" tumoral-mass formation, in the greater omentum, and a fluid collection was found in the abdomen (Morrison space and the pouch of Douglas). Notwithstanding, diagnosis of pseudomyxoma was preoperatively considered less probable due to the rarity of the affection and the advanced age. During laparotomy, the presence of the large mass observed at the ultrasound examination was revealed. Mucinous ascites was present with invasive mucinous implants in the peritoneum and in the greater omentum. Operation consisted in the total removal of the mass followed by evacuation of the mucinous ascites. Histological examination confirmed the clinical diagnosis of pseudomyxoma peritonei.

**KEY WORDS** *pseudomyxoma, peritoneum, ultrasound examination, fibrous*

**Introduction**

Pseudomyxoma peritonei (PMP) is a rare, chronic, relapsing, diagnostically challenging and poorly understood disease characterized by disseminated mucinous ascites and peritoneal implants. Difficulties exist with the definition of PMP.

Sugarbaker has suggested that "that the term pseudomyxoma peritonei syndrome be strictly applied to a pathologically and prognostically homogeneous group of cases characterized by histologically benign peritoneal tumors that are frequently associated with an appendiceal mucinous adenoma." This definition excludes all cases with mucinous adenocarcinoma.

Pseudomyxoma peritonei (PMP) is a rare disease that is characterized by a large amount of mucinous ascites with peritoneal and omental implants [10]. The etiology of the disease remains unclear. Histological, two main categories have been described: disseminated peritoneal adenomucinosis (DPAM) and peritoneal mucinous carcinomatosis (PMCA). It is commonly diagnosed incidentally at laparotomy. Most investigators agree that radical surgical debulking and appendectomy is the cornerstone of treatment, but the optimal management of the disease remains controversial [7, 10].

The optimal treatment is undoubtedly complete tumor excision, using complex

surgical peritoneotomy procedures [5, 6, 8, 9, and 11].

**Case Report**

The patient was a 60-year-old Romanian male, with 160 cm height and 95 kg weight who presented with fatigue and dyspnea. The patient is a moderate smoker and alcohol consumer (one or two bottles of beer and five cigarettes daily).

He was investigated for his anemia and the following results were found:

**Clinical data:** affected general state, the abdomen was diametrical increased, without collateral circulation, without icterus (sub-icterus), without hemorrhagic phenomena.

**Laboratory investigation showed:** The usual analyses were normal, cytotoxicity syndrome was absent, inflammatory syndrome was present. Erythrocyte Sedimentation Rate (ESR) was increased, the anemia was moderate (Hb 10% without modification of the formula). He had also no history of family malignancy.

The initial treatment was for liver cirrhosis and the patient received hepatic protection medication.

**Ultrasound examination** was helpful for the positive diagnosis - it confirmed the presence of the ascites in peritoneal cavity, and "net's eyes" tumoral formation, without sings of portal venous pressure.



**Figure1.** Ultrasound examination of the abdomen revealed the presence of the fluid in peritoneal cavity and the presence of a “net’s eyes” tumoral formation without sings of the portal venous pressure.

The patient was investigated through **laparotomy**. The “net’s eyes” tumoral formation was found in the greater omentum and the peritoneal cavity was filled with mucinous liquid.

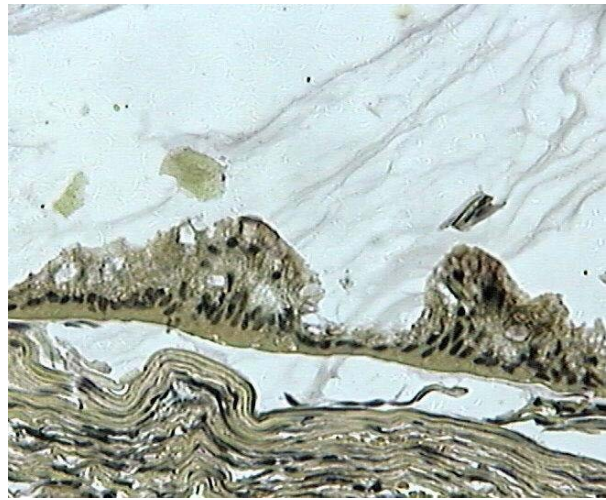


**Figure2.** Macroscopically, it was a round shaped formation of about 12 cm in diameter containing gelatinous substance like pseudomyxoma peritonei. Thus, we diagnosed it as retroperitoneal pseudomyxoma.

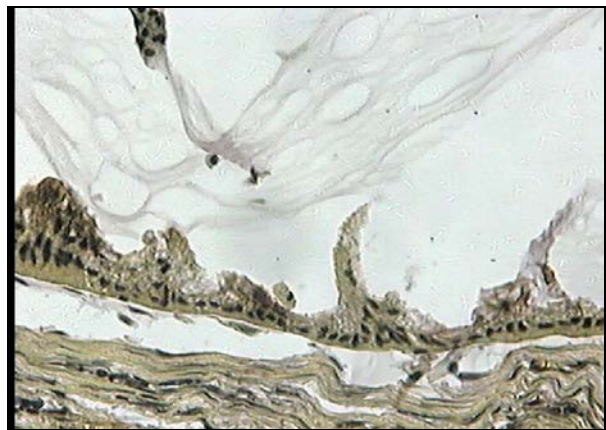
**Biopsy and histopathological** examinations of the tumoral mass confirmed the presence of PMP: fibroma with myxomatous aspect, a few cells are round shaped, others are stellar shaped with prolongation forming a net.

The surgical intervention consisted in total removal of the mass and evacuation of the mucinous ascites.

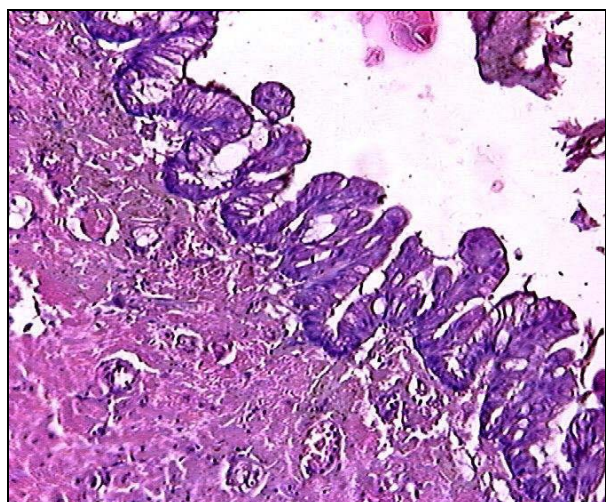
The patient’s evolution was good for 4 years. Then, he complained of increased abdominal circumference and coughing; the ultrasound examination showed the recurrence of extensive amount of multiloculated material throughout the peritoneal cavity.



**Figure3.** Pseudomyxoma peritonei characterized by pools of mucin containing very rare clusters of low-grade epithelium. (Hematoxylin-eosin, original magnification  $\times 200$ )

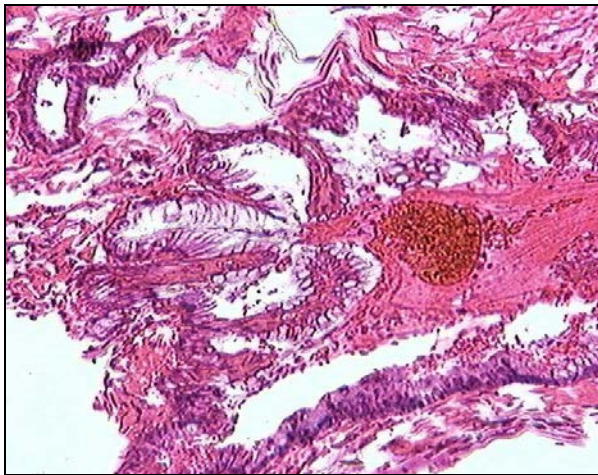


**Figure4.** Gelatinous aspect liquid (Hematoxylin-eosin, original magnification  $\times 200$ )

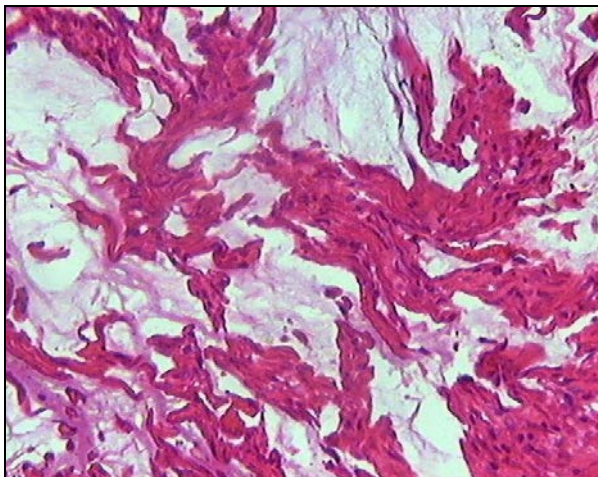


**Figure5.** Typical epithelium of a cyst adenoma with pseudo stratified, columnar cells containing elongated, crowded, hyper chromatic nuclei and scattered, well-defined goblet cells. (Hematoxylin-eosin, original magnification  $\times 200$ ).





**Figure6. Adenomatous epithelium within diverticulum with marked associated acute inflammation. (Hematoxylin-eosin, original magnification  $\times 200$ ).**



**Figure7. Pools of acellular mucin infiltrate through fibro-vascular tissue (Hematoxylin-eosin, original magnification  $\times 400$ ).**

Therefore, the second debulking surgery was performed. The patient had an uneventful recovery and was discharged in a stable condition. He had the third debulking surgery for recurrent disease. He died from an unrelated cause.

## Discussion

Pseudomyxoma peritonei is an indolent disease and preferentially affects women with an average age of 53 years [9]. It is traditionally believed that most cases of PMP originate from ovarian tumors. This belief is challenged recently by increased usage of immunohistochemical stains and molecular genetic studies, which showed a large proportion of these tumors to be secondary to appendiceal tumors in both men and women [7].

As symptoms remain non-specific the disease presents a great diagnostic challenge to clinicians.

Clinical presentation is late and patients usually experience a long course of health deterioration before an accurate diagnosis is made. Due to its indolent nature, advanced stages of the disease with generalized peritoneal tumor implants, fistula formation and adhesions are common. In this advanced stage, abdominal symptoms caused by partial or complete bowel obstruction are the main complaints.

A precise diagnosis is difficult due to the lack of specific symptoms in the early stage of the disease. Routine laboratory studies are seldom helpful in making this diagnosis.

In the majority of cases, it is often an unexpected finding at explorative laparoscopy, which remains the main diagnostic tool with the final diagnosis being confirmed by histopathology [15].

## Conclusions

We have described one male patient who developed pseudomyxoma peritonei arising in the greater omentum. We were diagnostically challenging and the positive diagnosis was accurately established at surgery and confirmed by histopathology. In order to remove the tumor surgical debulking procedures were used. Awareness of this indolent and rare condition is an important prerequisite for early diagnosis and appropriate management [6].

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