

PSEUDOMYXOMA PERITONEI OF APPENDICEAL ORIGIN – AN UNUSUAL CAUSE OF ABDOMINAL COMPARTMENT SYNDROME: CASE REPORT.

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PSEUDOMYXOMA PERITONEI OF APPENDICEAL ORIGIN – AN UNUSUAL CAUSE OF ABDOMINAL COMPARTMENT SYNDROME: CASE REPORT (ABSTRACT): A 75-year old male patient was admitted with clinical features of tensioned ascites, dyspnea for the last two weeks. He had a history of appendectomy four years previously. During the first 24 hours in the intensive care unit (ICU) the patient required high volume of i/v infusion and vasopressors for correction of hypotension and oliguria. Computed tomography revealed characteristic signs for pseudomyxoma peritonei. Grade III Abdominal Compartment Syndrome (ACS) was detected by measuring the intra-abdominal pressure via the bladder and the patient was scheduled for decompressive laparotomy. During decompressive laparotomy surgical debulking, peritoneal stripping combined with intraoperative hyperthermic intraperitoneal chemotherapy with 5-FU were performed. After surgical decompression the patient's condition improved, the postoperative period was uneventful. During a 14 month follow-up period the patient was free of disease recurrence. To the best of our knowledge this is the first case of pseudomyxoma peritonei (PMP) induced intraabdominal hypertension and ACS successfully treated by surgical debulking, peritoneal stripping combined with intraoperative hyperthermic intraperitoneal chemotherapy with 5-FU.

KEY WORDS: PSEUDOMYXOMA PERITONEI • ABDOMINAL COMPARTMENT SYNDROME

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INTRODUCTION

Intra-abdominal hypertension (IAH) is defined as sustained or repeated pathologic elevation of abdominal pressure over 12 mmHg [1].

Abdominal compartment syndrome (ACS) is defined as the adverse physiologic effect of increased intraabdominal pressure (IAP) [2]. Elevated IAP can be induced by increased intra-abdominal volume (ascites, blood, tumors, and pregnancy) or conditions that limit the expansion of the abdominal wall (burns); the development of IAH-induced multiple organ dysfunction and failure appears when abdominal pressure is over 10 - 15 mmHg [1, 2]. Abdominal compartment syndrome in patients with malignancy such as large ovarian cystadenoma [3,4], massive neoplastic ascites [5], ovarian tumor (granulosa cell tumor) [6] and pseudo-Meig's syndrome [7] were recently published.

The authors present an additional case of ACS due to malignancy and the first Pseudomyxoma peritonei (PMP)-induced ACS case.

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CASE REPORT

In February 2008, a 75-year old male patient was admitted with clinical features of tensioned ascites, dyspnea for the last two weeks. He had a history of appendectomy in 2004 for mucinous cystadenoma. On physical examination, the abdomen was largely distended with a significant amount of ascites (Fig. 1).



Fig. 1 Largely distended abdomen with a significant amount of ascites.

During the first 24 h in the ICU the patient required 5300 ml infusion and vasopressors for oliguria (the urine output was 20.8 ml/h) and hypotension (systolic blood pressure 100 mm Hg). The ultrasound (US) and computed tomography (CT) revealed characteristic signs for pseudomyxoma peritonei [8]: low-attenuation mucinous ascites (10 HU) and associated scalloping of the liver margins and hypodense peritoneal implants with bowel implants (Fig. 2).



Fig. 2 Abdominal CT-scan demonstrating mucinous ascites and scalloping of the liver margins.

The IAP measured using a Foley catheter (according to the technique described by Kron IL. *et al*) [1, 9] was 40 cm H₂O or 29 mm Hg. With the diagnosis of pseudomyxoma peritonei complicated with ACS the patient was taken for decompressive laparotomy. During surgical debulking, peritoneal stripping combined with intraoperative hyperthermic intraperitoneal chemotherapy with 5-FU 750 mg/m² at 43°C was performed. Fifteen liters of mucinous component and 5 kg of solid component were removed from the peritoneal cavity (Fig. 3).

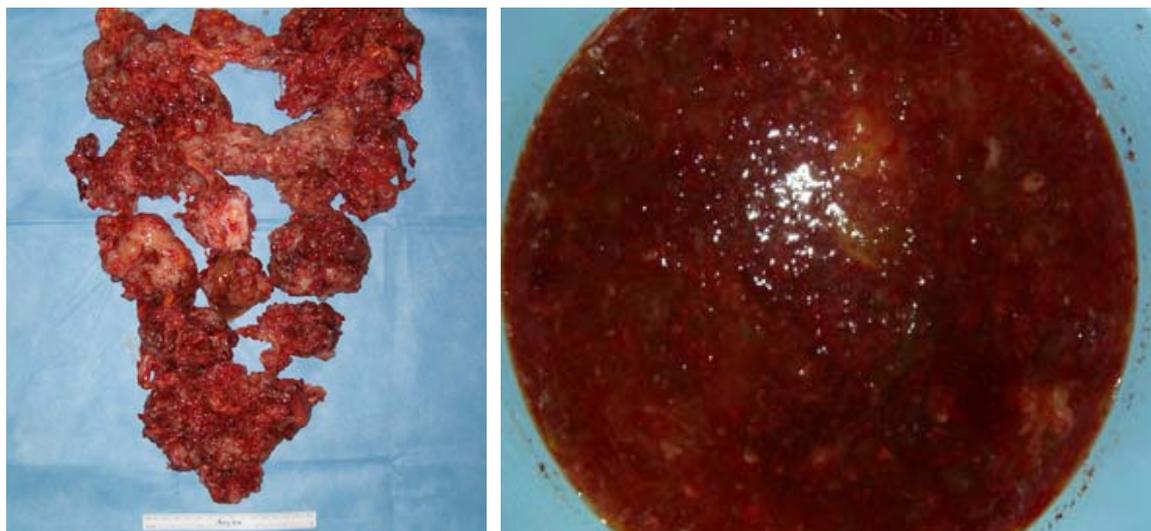


Fig. 3 Solid component removed from the peritoneal cavity (left) and mucinous component removed from the peritoneal cavity (right).

Immediately after surgery the urine output increased from 20.8 ml/h to 110 ml/h and dyspnea resolved. Since our patient developed ACS, we considered the extension of surgery to the Sugarbaker procedure unnecessary [10, 11]. Intraperitoneal hyperthermic chemotherapy with 5-FU 750 mg/m² was administrated until the 5th postoperative day. Histologically, the benign form of disseminated peritoneal adenomucinosis was diagnosed (Fig. 4) [12]. The postoperative period was uneventful and the patient discharged on the 15th postoperative day, the patient being scheduled for close follow-up.

DISCUSSION

Abnormally elevated intraabdominal pressure (over 12 mm Hg) is defined as intraabdominal hypertension; if unrecognized or untreated, it can lead to abdominal compartment syndrome – a condition defined as adverse physiologic consequence of acutely increased intraabdominal pressure [1,13,14].

Prolonged, unrelieved elevated intraabdominal pressure over 20 mmHg can induce cardiopulmonary dysfunction, renal failure, visceral ischemia, shock and death [13,14]. Under these circumstances urgent surgical decompression is recommended.

Intra-abdominal hypertension and abdominal compartment syndrome is frequently diagnosed in critically-ill patients following severe abdominal trauma, major abdominal surgery, ruptured abdominal aortic aneurysms, intra- or retroperitoneal hemorrhage, and thermal injury; less common causes include abdominal neoplasm, tensioned ascites, and pancreatitis or massive fluid resuscitation for sepsis [15-17].

Recently a few case reports of abdominal compartment syndrome associated with different malignancies were published [3-7].

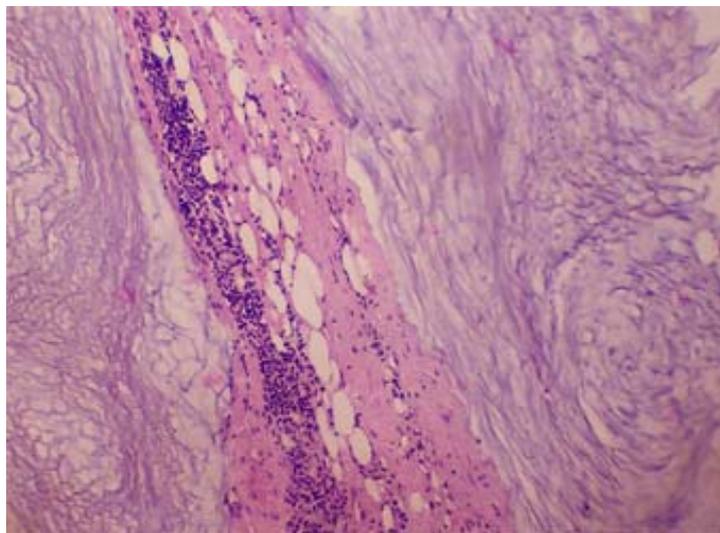


Fig. 4 Histology: disseminated peritoneal adenomucinosis (HEX40).

A PubMed search using “pseudomyxoma peritonei”, “intraabdominal hypertension”, “abdominal compartment syndrome” revealed no published cases of PMP-induced IAH and ACS.

To the best of our knowledge this is the first case described of PMP-induced ACS. In the present case, the excessive IAP induced oliguria, dyspnea and hypotension with rapid improvement in the patient’s condition after decompression indicated the existence of ACS.

Pseudomyxoma peritonei is a poorly understood disease. Literarily translated PMP means false mucinous tumor. Mucocele of the appendix was recognized as a pathologic entity by Rokitansky in 1842 and was formally named by Feren in 1876 [18], but it was R. Werth [19] to use the term “pseudomyxoma peritonei” for the first time in 1884 when he described its occurrence in association with a mucinous carcinoma of the ovary.

The incidence of PMP is approximately two in 10,000 laparotomies, and about 75% of patients are female with an average age of 53 years [20].

The unifying diagnostic feature of PMP is the presence of extracellular mucin in the peritoneal cavity. Patients with PMP develop a characteristic mucinous ascites associated with mucinous and cellular implants on the peritoneum [21]. The mucinous tumors accumulate at anatomic sites such as the pelvis, right retrohepatic space, the greater and lesser omentum and the abdominal gutters.

Tumor implants on the small bowel are extremely rare, most likely because peristaltic movements prevent the adherence and implantation of tumor cells on the bowel surface [10].

This large accumulation of mucinous ascites can theoretically induce IAH and ACS. Any increase in the volume of the abdominal or retroperitoneal contents increases IAP, thus it is only logical to presume that PMP associated with large accumulations of mucinous ascites could induce IAH and ACS.

In a large majority of patients (80%), PMP arises from appendicular disease and not ovarian disease [22]. According to the data published by Seidman *et al.* in 90% of female patients diagnosed with PMP, the ovarian seeding is observed [23].

The most common sites of origin are considered the appendix and the ovary, although other sites, such as the gallbladder, stomach, pancreas, colon, uterus, fallopian tubes, urinary bladder, breast and lung, have been described [24, 25].

The recent advances in the immunohistochemistry and molecular biology have greatly contributed to the debate concerning the appendicular or ovarian origin of PMP, thus the molecular profiles generally exhibit a colorectal rather than an ovarian pattern [26]. Up-to-date the origin of the disease is identified by immunohistochemical techniques [26].

Because of the rarity of this condition, the diagnosis of PMP is often difficult. Ultrasonographic findings are echogenic ascites with multiple semi-solid tumors and scalloping of the hepatic and splenic margins from external pressure of adjacent peritoneal implants [27].

On CT, PMP is characterized by low-attenuation mucinous ascites, associated scalloping of the liver margins and hypodense peritoneal implants with bowel implants [8].

Histology is always required and should be achieved by laparotomy. A laparoscopic approach should be avoided since all laparoscopic port sites develop tumor infiltration through the entire thickness of the abdominal wall [28].

Ronnett *et al.* histologically described three main diagnostic categories of PMP: (1) the benign form of disseminated peritoneal adenomucinosis (DPAM); (2) the malignant form of peritoneal mucinous carcinomatosis (PMCA), and (3) the intermediate form with features between DPAM and PMCA [12]. This classification has an important prognostic significance, because patients with DPAM have a significantly better prognosis than those with PMCA [29].

The therapeutic recommendations are controversial due to the rarity of PMP, the lack of randomized studies, and its complex biology.

Certain features such as low biological aggressiveness, rare metastasis to lymph nodes and liver, and accumulation in anatomically resectable sites (pelvis, right retrohepatic space, the greater and lesser omentum and the abdominal gutter) make the disease amenable to curative surgery [10].

Up to date, the standard treatment includes cytoreductive surgery, inclusively peritoneal stripping procedures and appendectomy, in combination with intraoperative/postoperative intraperitoneal chemotherapy [10] in our case, due to the poor patient's condition we performed surgical debulking, peritoneal stripping combined with intraoperative hyperthermic intraperitoneal chemotherapy with 5-FU 750 mg/m² at 43°C.

In summary, ACS is a surgical emergency frequently diagnosed in critically-ill patients and usually requires immediate decompression. We described the first PMP-induced ACS successfully treated by surgical decompression.

Abdominal hypertension and compartment syndrome must be considered in all the patients with PMP and if recognized immediate abdominal decompression by surgical debulking, peritoneal stripping combined with intraoperative hyperthermic intraperitoneal chemotherapy should be considered.

REFERENCES

1. Malbrain ML, Cheatham ML, Kirkpatrick A, Sugrue M, Parr M, De Waele J, et al. Results from the International Conference of Experts on Intra-abdominal Hypertension and Abdominal Compartment Syndrome. I. Definitions. *Intensive Care Med.* 2006; 32(11): 1722-1732.
2. Schein M, Wittmann DH, Aprahamian CC, Condon RE. The abdominal compartment syndrome: the physiological and clinical consequences of elevated intra-abdominal pressure. *J Am Coll Surg* 1995; 180(6): 745-753.
3. Chao A, Chao A, Yen YS, Huang CH. Abdominal compartment syndrome secondary to ovarian mucinous cystadenoma. *Obstet Gynecol.* 2004; 104(5 Pt 2): 1180-1182.
4. Giuliani A, Basso L, Demoro M, Scimo' M, Galati F, Galati G. Bilateral ovarian mucinous cystadenoma in an adolescent presenting as abdominal compartment syndrome. *Eur J Obstet Gynecol Reprod Biol.* 2008; 140(2): 278-279.
5. Etzion Y, Barski L, Almog Y. Malignant ascites presenting as abdominal compartment syndrome. *Am J Emerg Med.* 2004; 22(5): 430-431.
6. Haan J, Johnson SB, Scalea T. Ovarian Tumor Causing Abdominal Compartment Syndrome. *J Trauma.* 2007; 62(3): 768–769.
7. Peparini N, Di Matteo FM, Silvestri A, Caronna R, Chirletti P. Abdominal hypertension in Meigs' syndrome. *Eur J Surg Oncol.* 2008; 34(8): 938-942.
8. Sulkin TV, O'Neill H, Amin AI, Moran B. CT in pseudomyxoma peritonei: a review of 17 cases. *Clin Radiol.* 2002; 57(7): 608–613.
9. Kron IL, Harman PK, Nolan SP. The measurement of intra-abdominal pressure as a criterion for abdominal re-exploration. *Ann Surg.* 1984; 199(1): 28-30.
10. Sugarbaker PH. Cytoreductive surgery and peri-operative intraperitoneal chemotherapy as a curative approach to pseudomyxoma peritonei syndrome. *Eur J Surg Oncol.* 2001; 27(3): 239-243.
11. Sugarbaker PH. Cytoreduction including total gastrectomy for pseudomyxoma peritonei. *Br J Surg.* 2002; 89(2): 208–212.
12. Ronnett BM, Yan H, Kurman RJ, Shmookler BM, Wu L, Sugarbaker PH. Patients with pseudomyxoma peritonei associated with disseminated peritoneal adenomucinosis have a significantly more favorable prognosis than patients with peritoneal mucinous carcinomatosis. *Cancer.* 2001; 92(1): 85-91.
13. Burch JM, Moore EE, Moore FA, Franciose R. The abdominal compartment syndrome. *Surg Clin North Am.* 1996; 76(4): 833-842.
14. Meldrum DR, Moore FA, Moore EE, Franciose RJ, Sauaia A, Burch JM. Prospective characterization and selective management of the abdominal compartment syndrome. *Am J Surg.* 1997; 174(6): 667-673.
15. Schein M, Ivatury R. Intra-abdominal hypertension and the abdominal compartment syndrome. *Br J Surg.* 1998; 85(8): 1027-1028.
16. Saggi BH, Sugeran HJ, Ivatury RR, Bloomfield GL. Abdominal compartment syndrome. *J Trauma.* 1998; 45(3): 597-609.
17. Bailey J, Shapiro MJ. Abdominal compartment syndrome. *Crit Care.* 2000; 4(1): 23-29.
18. de Rezende Pereira JC, Trugilho JC, Sarmat AA. Mucocele of the appendix. *Surgery.* 2004; 136(5): 1096-1097.
19. Hinson FL, Ambrose NS. Pseudomyxoma peritonei. *Br J Surg.* 1998; 85(10): 1332-1339.
20. Mann WJ, Jr., Wagner J, Chumas J, Chalas E. The management of pseudomyxoma peritonei. *Cancer.* 1990; 66(7): 1636–1640.
21. Elias D, Laurent S, Antoun S, Duvillard P, Durceux M, Pocard M, Lasser P. Pseudomyxoma peritonei treated with complete resection and immediate intraperitoneal chemotherapy. *Gastroenterol Clin Biol.* 2003; 27(4): 407-412.
22. Elias D, Sabourin JC. Les pseudomyxomes péritonéaux. Mise au point. *J Chir (Paris).* 1999; 136(6): 341-347.
23. Seidman JD, Elsayed AM, Sobin LH, Tavassoli FA. Association of mucinous tumors of the ovary and appendix. A clinicopathologic study of 25 cases. *Am J Surg Pathol.* 1993; 17(1): 22-34.
24. Sherer DM, Abulafia O, Eliakim R. Pseudomyxoma peritonei: a review of current literature. *Gynecol Obstet Invest.* 2001; 51(2): 73–80.

25. Shimoyama S, Kuramoto S, Kawahara M, Yamasaki K, Endo H, Murakami T, et al. A rare case of pseudomyxoma peritonei presenting an unusual inguinal hernia and splenic metastasis. *J Gastroenterol Hepatol.* 2001; 16(7): 825–829.
26. Ronnett BM, Shmookler BM, Diener-West M, Sugarbaker PH, Kurman RJ. Immunohistochemical evidence supporting the appendiceal origin of pseudomyxoma peritonei in women. *Int J Gynecol Pathol.* 1997; 16(1): 1-9.
27. Lersch C, Frimberger E, Ott R, Classen M. Gray-scale sonographic findings in a patient with pseudomyxoma peritonei. *J Clin Ultrasound.* 2001; 29(3): 186–191.
28. Esquivel J, Sugarbaker PH. Pseudomyxoma peritonei in a hernia sac: analysis of 20 patients in whom mucoid fluid was found during a hernia repair. *Eur J Surg Oncol.* 2001; 27(1): 54–58.
29. Ronnett BM, Zahn CM, Kurman RJ, Kass ME, Sugarbaker PH, Shmookler BM. Disseminated peritoneal adenomucinosis and peritoneal mucinous carcinomatosis. A clinicopathologic analysis of 109 cases with emphasis on distinguishing pathologic features, site of origin, prognosis, and relationship to “pseudomyxoma peritonei”. *Am J Surg Pathol.* 1995; 19(12): 1390–1408.