RARE INTRAABDOMIAL TUMOR

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ABSTRACT:

Pseudomyxoma peritonei (PMP) is a rare neoplastic, chronic and insufficient disease, which has the characteristics of the disseminated mucinous ascites and the presence of peritoneal implants. The making of precise and well-timed diagnosis is a difficult process because of the lack specific symptoms. The exact preoperative diagnosis 'PMP" can be prescribe after CT-imaging, which is sensitive with its 'mussel shell' symptom on the surface of the visceral organ, resulting from the compression of the vicouse mucinous ascites and the presence of fibrosis. We report a rare case of our practice, with anamnesis of abdominal swelling, lost of apetite, consumative syndrome – more than 10 kilos for two months. The difficulties in diagnostic process were overwhelmed. Despite of the quantity of the studies, directed to the treatment of PMP, this disease stay constantly enigmatic with prorogated clinical elapsing and the high level of recurrence in the complex treatment and have to be discussed the application of operative methods and chemotherapy.

Pseudomyxoma peritonei (PMP) is a rare neoplastic, chronic and insufficient disease, which has the characteristics of the disseminated mucinous ascites and the presence of peritoneal implants. [1]

There are two different biological types, which have prolonged elapse and tendancy to be aggressive:

- Disseminated peritoneal adenomatosis (DPA)-binign type
- Peritoneal mucinous carcinosis (PMC)-malignant type

PMP is female-like tumor and is associated with the presence of ovarial mucinous tumor. It is very rare met in men, because of the other type of origin – lessions of the appendix Other possible primary tumor sources could be lession in the rectum, colon, gall-bladder, pancreas, urachus, bladder, mammal gland, and lung, but they are very rare and are associated with peritoneal mucinous carcinosis (PMC) [2]

According Solkar et al. 2004, only 10% of the patients with PMP have 5.5 year survival rate after the diagnosing. [3, 4]

The making of precise and well-timed diagnosis is a difficult process because of the lack specific symptoms. The exact preoperative diagnosis 'PMP" can be prescribe after CT-imaging, which is sensitive with its 'mussel shell' symptom on the surface of the visceral organ, resulting from the compression of the vicouse mucinous ascites and the presence of fibrosis. Due to this image method there often can be found multiple cystic masses with lipid consistency in the peritoneum. They could have ferrules of calcificates with typical festooning of the liver outlines, the thickness of the omentum, and the differences in the compression of visceral organs and structures. The preoperative ultrasound examination frequent shows raw, sluggish ascites with septal adhesions and high levels of ultrasound consistency. The combination of clinical results and radiological signs are highly specifying for the diagnosis of PMP. In patients with occasional find, radiology can find abdominal calcifacation plates, ascites and unseparated soft tissue masses. In this cases the method of choice is CT. [5]

The accuracy and specify of MRI in the diagnostics is not clear yet.

The explorative diagnostic laparoscopy is indicated as gold standard in diagnosis-making and consequent morphological investigation.

A study of O'Connel J.T. et al. 2002 presents PMP as neoplastic disease from MUC-2 producing goblet cells. The extracellular accumulation of mucin is due to alteration of glucoprotein in the cells extremitas in the tumor's stroma. [6]

The most frequent operations are according to surgical treatment of PMP:

- Right Hemicolectomy- 88%
- Resection of the sigma- 65%
- Gastrectomy- 47%
- Colectomy- 6%
- Splenectomy- 79%
- Cholecystectomy- 76%
- Histerectomy and bilateral salpingo-ooforectomy 50%
- Omentectomy- 60%

(De Simone M, et al. 2006) [7]

The developing of treatment strategy in cases of PMP is a disputable question, yet. Complex therapy is applied, including citoreductive surgical procedure and segmentary resection the obdurate organ and omentectomy. Reoperation is necessary in the cases of recidives.

In a study of Sugarbaker et al. 1995, they proved that intraoperative chemotherapy with 5-fluorouracil and Mythomicyn C have minimal effect in the reduction of "tumor implants" of atypical epithelium and don't provoke apopthosis of these cells. [8]

Deraco et al. 2006 make resection of mainly engaged organ and intraoperituneal citoreductive chemotherapy with Mythomicyn C, warmed up to 42 ЬС. [9] The aim is to be removed the tumor "in toto" and to be destroyed the residual small tumor nodes, according to Smeenk et al. 2006. They notice that the conducting of combined therapy, despite of high levels of toxicity, enlarge the survival rate. [10]

Miner J.T. et al. 2005 defined that the prognosis of the outcome is connected with :

- Tumor differentiation (G) – morphologically confirmed.
- Preoperative extend of the tumor
- The resection of the targeted organ and citoreductive therapy. [11]

The most informative carcino markers are CEA and CA-19-9, which are increasing their levels in case of recidive. [12]

According to Sugarbaker et al. 1995, it is necessary to be applied chemotherapy with 5fluorouracil + Mythomicyn C with 'Second look" when are made big operations with grate extend of the surgery. The tumor markers have to be investigated in a 3 month period CT have to be made in a 6 month period. [8]

Deraco et al. 2003 classify the disease in four grade scale, according to followed criteria for mortality and morbidity:

- I. Grade – without complications
- II. Grade – small and light complications
- III. Grade – serious complications (demanding reoperation and intensive care)
- IV. Grade – in status that led to exitus letalis [13]

RESULTS:

We present a case of PMP, a male on 53 years, who enter in the Clinic of General and Operative Surgery with anamnesis of abdominal swelling, lost of appetite, consumative syndrome – more than 10 kilos for two months and laboratory data as presented: /TAB/

Hb	Hct	Leu	CYE	CEA	CA19-9	Protein	Albumin	Urea	Glucose
118	0,35	6,3	94	17,3	1257	75	36	3,8	4,3

By the applied abdominal ultrasonography we found a presence of moderate quantity of ascites, which fluctuated with ultrasonogenic structures and fixated intestinal stitches to the liver and diaphragma. We evacuate yellow ascites, which gelatinize as soon as it was get out of the body. The CT data showed us plenty of cystic lesions in the peritoneum, which were compressed the liver. The liver had a typical festoon image. The consistency of the ascites was about 20 HE. We found heavily ticked omentum majos and clinical view as PMP Through the exploration of the abdominal cavity in planned order, we found plenty brides between abdominal organs, yellow-like jelly ascites around 2 l (includes many light formation)

We found intact appendix, which was situated medial and retrperitoneal. The omentum was enveloped by the tumor formation with extends of 15/10/5 cm. (Fig. 1 - 7)

Omentectomia and lavage was made by serum physiologicum. At last, we put two drainages.

The early postoperative period was smooth, under the protection analgetics, cristaloid solutions, monosaccharids solutions and low-weight heparins.

Morphologiacally investigated materials were

introduced by multiple necrosis and confluated mucous tracts, separated by different in size septal brides with hyperfolicular vessels and lymphotic infiltrats. Morphological diagnosis: Pseudomyxoma peritonei.

The patient was directed for initiating of systematic chemotherapy with Mythomicyn C and 5-fluorouracil.

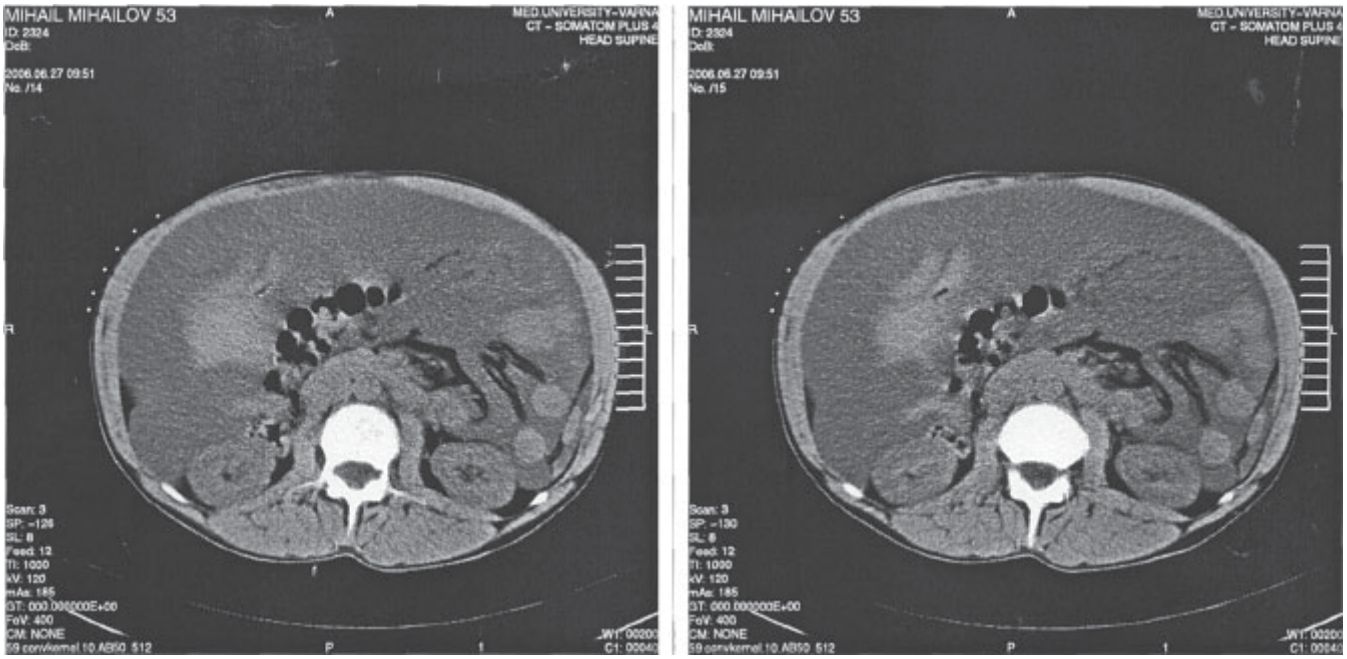

Fig. 1

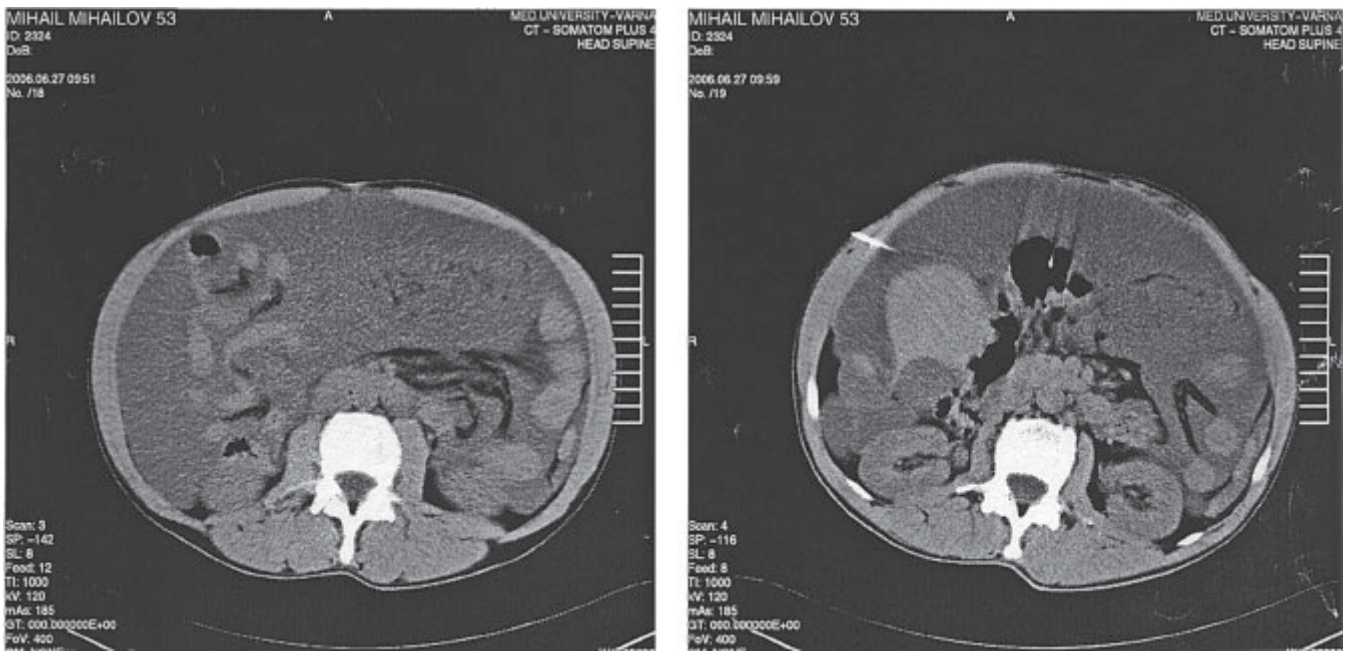

Fig. 2

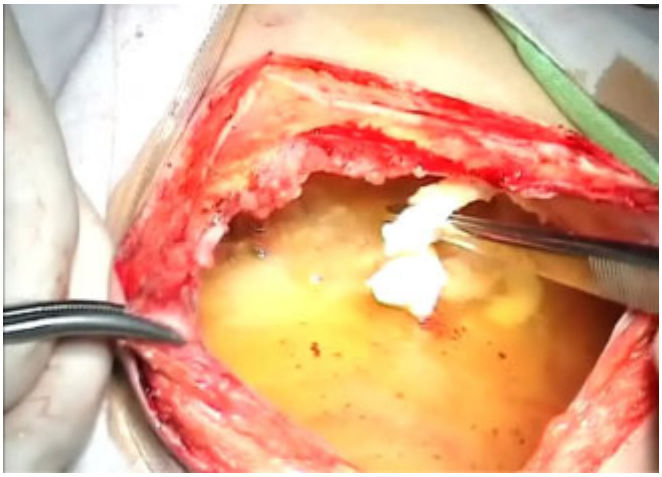

Fig. 3

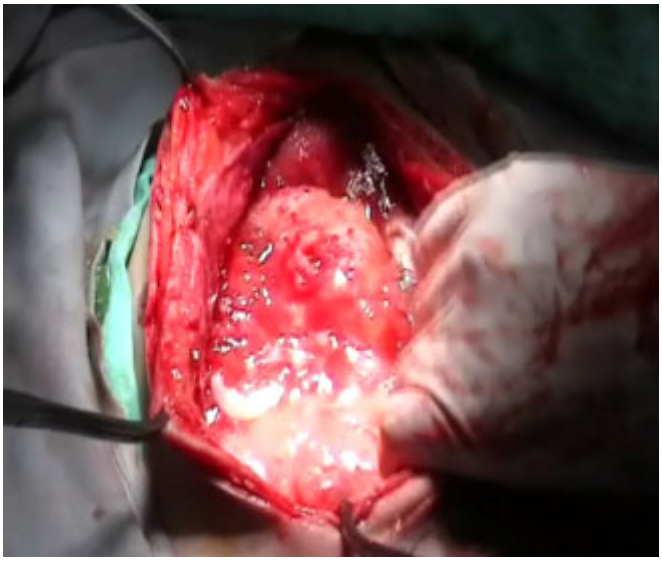

Fig. 4

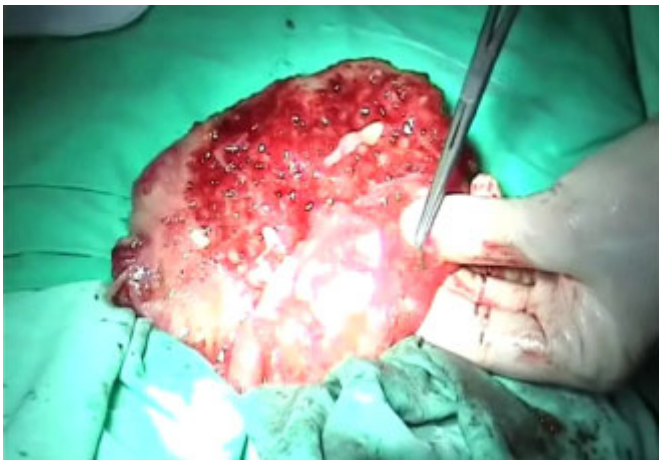

Fig. 5

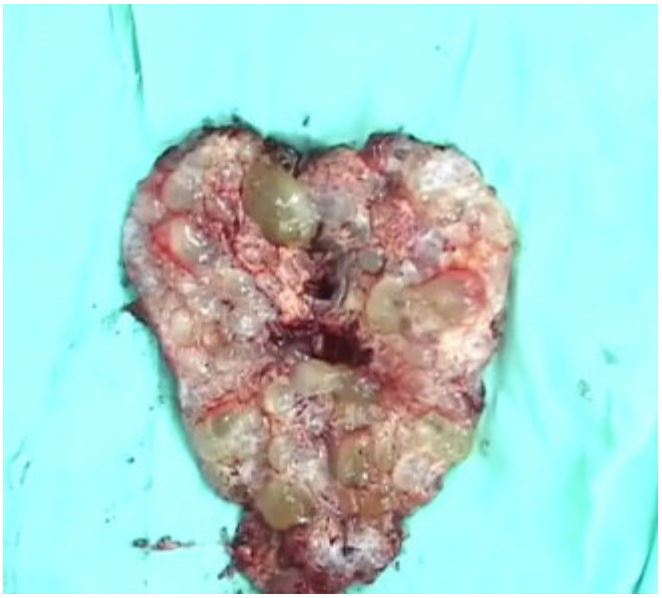

Fig. 6.

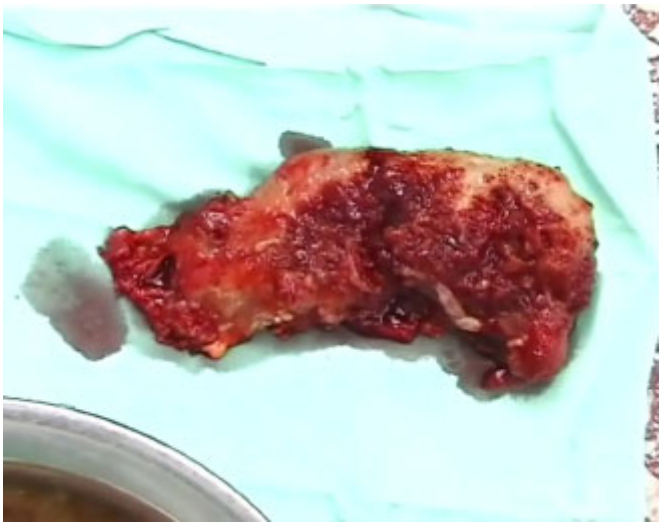

Fig. 7

CONCLUSION:

Despite of the quantity of the studies, directed to the treatment of PMP, this disease stay constantly enigmatic with prorogated clinical elapsing and the high level of recurrence in the complex treatment and have to be discussed the application of operative methods and chemotherapy.

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