LETTER TO THE EDITOR / EDİTÖRE MEKTUP
DOI: 10.26650/IUITFD.1557577
İst Tıp Fak Derg 2024 / J Ist Faculty Med 2024

PSEUDOMYXOMA PERITONEI: A RARE ENTITY

PSÖDOMİKSOMA PERİTONEİ: NADİR BİR ANTİTE

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Cite this article as: Canbay Torun B. Pseudomyxoma peritonei: A rare entity. J Ist Faculty Med 2024;87(4):356-357. doi: 10.26650/IUITFD.1557577

Dear Editor,

I write to bring an attention to a rare disease, pseudomyxoma peritonei (PMP), which was first described by Werth in 1884, yet it remains an orphan disease with limited awareness (1). The incidence of PMP is one to two patients/per million in a year that might be due to misdiagnosis (2).

PMP typically arises from a ruptured mucinous tumour, usually originating from the appendix, stomach, gall bladder, small and large intestine, fallopian tubes, ovaries, pancreas and lung, and mucin accumulates inside the peritoneal cavity and is called 'jelly belly' (1). PMP is histopathologically classified into four subtypes, based on present of tumour cells, the amount of mucin and the aggressiveness level, which are low grade, high grade, high grade with signet ring cells and acellular mucin (2).

Despite being recognised for more than a century, PMP's management is still challenging due to misdiagnosis or delayed diagnosis. This delay usually causes significant progression of the disease and adversely affects patients' outcomes. Diagnosing PMP is still challenging due to it's indolent nature with nonspesific symptoms such as abdominal discomfort and distension at the time of the disease initiation. Imaging techniques may not distinguish mucinous material from other fluid collection and may cause difficulty in obtaining a definitive histopathological diagnosis (3).

The current standard treatment of PMP is cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC), which improves patient outcomes and should be performed in specialised centres (1, 4). PMP is frequently diagnosed as an unexpected finding during emergency surgeries such as acute appendicitis or gynaecologic surgeries or imaging for other conditions. As a result, general surgeons and gynaecologists will occasionally come across such cases and they will face challenges in both diagnosis and treatment due to its rarity, lack of awareness and limited guidance on management (1). When PMP is encountered unexpectedly during surgery, surgeons should focus on taking sufficient biopsies and removing the appendix, avoiding extensive resections. After recovery and diagnostic confirmation, specialised care should be sought for further management (1).

In a multicenter study, the outcome data of 2298 patients with PMP of appendiceal origin were analyzed, and the best results were obtained with proper cytoreduction. (5). Chua et al. showed that the five-year survival rate for patients who undergo incomplete (completeness of cytoreduction) (CCR2 or CCR3), where visible residual disease remains, is 24%; while it is 85% for those who achieve CCR0 and 80% for patients with a CCR1 resection (5). Delays in diagnosis or misdiagnosis and improper treatment management after diagnosis determine the completeness of cytoreduction and therefore the prognosis in PMP patients.

In conclusion, physicians', especially general surgeons and gynaecologists who perform intra-abdominal surger-

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Submitted/Başvuru: 28.09.2024 • **Revision Requested/Revizyon Talebi:** 09.10.2024 •
Last Revision Received/Son Revizyon: 09.10.2024 • **Accepted/Kabul:** 10.10.2024 • **Published Online/Online Yayın:** 14.10.2024

ies, awareness about PMP should be increased so that timely and appropriate treatment can be performed without missing diagnosis and referral to specialised centres will increase the survival of these patients.

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