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RESEARCH ARTICLE

PSEUDOMYXOMA PERITONEI SECONDARY TO A VERY RARE OVARIAN CANCER: PRIMARY INFILTRATIVE INTESTINAL TYPE MUCINOUS ADENOCARCINOMA OF THE OVARY WITH INTESTINAL METASTASIS (A CASE REPORT)

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Abstract

Pseudomyxoma peritonei (PMP) syndrome is a disease that typically occurs from ruptured appendiceal mucocele neoplasms. PMP syndrome may arise from malignant transformation of a primary ovarian cancer. In this case study, we present the evaluation and treatment of a 48-year-old patient diagnosed with a left ovarian mass and significant symptomatic abdominopelvic mucinous ascites. Peritoneal cytology indicated paucicellularmucin. The final diagnosis confirmed the presence of diffuse peritoneal adenomucinosis. The treatment approach involved a cytoreductive surgery and chemotherapy. Our findings contribute to the existing body of literature endorsing the use of this intensive treatment approach, typically reserved for advanced abdominal malignancies. We applied this strategy to a patient presenting with a rare clinical entity, further supporting its efficacy in such cases.

Introduction:-

The association of 02 rare entities in a single case makes it very interesting and a source of information for subsequent studies. PMP is a rare disease with an estimated incidence of 0.2 per 100,000 inhabitants per year. The age at the time of diagnosis varies from 20 to 80 years. The term pseudomyxoma peritonei (PMP) also called "jelly belly cancer" corresponds to a clinical-radiological situation defined by the accumulation of mucin, localized or diffuse, in the abdominal and/or pelvic cavity, related to the presence of a mucinous neoplasia located in the vast majority of cases in the appendix, much more rarely in the urachus, pancreas and ovary (teratoma). Mucinous ovarian cancer (MOC) is a rare subtype of epithelial ovarian carcinoma (EOC), around 80% of mucinous carcinomas of the ovary are metastatic, it is agreed that diagnosing primary MOC requires careful pathological assessment as it is histologically very similar to other mucinous carcinomas, especially colorectal carcinomas (CRC). We report a case of a woman (48 years old) diagnosed and treated at the obstetrics and gynecology department of the Hassan II University Hospital in Fez.

Case Report:

This is the case of a 48-year-old married lady, G1P1, referred for management of abdominal distention of 6 months duration, associated with weight loss estimated at 5kg, having benefited from a mini laparotomy at the provincial central hospital in el HAJEB in the month of February 2023 as part of the exploration of her pathology with

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cytology and peritoneal biopsy in favor of low-grade peritoneal mucinous neoplasm (peritoneal pseudomyxoma grade I of WHO 2020). Patient was then referred to our hospital for further treatment. Clinical examination: reveals a stable conscious patient in good general condition with a BMI of 27 and presence of a median sub umbilical scar, very distended abdomen. A vaginal examination revealed; peri-orifical redness, on a firm cervix, without any bleeding.

Thoraco-abdomino-pelvic CT scan showed a peritoneal pseudomyxoma of great abundance, Tumorous ovaries (increased in size with a cystic component difficult to measure on the scanner) probably related to a borderline serous cystadenoma: to be compared with the data of a pelvic MRI with an Absence of visualization of the appendix.

Pelvic MRI objectified a typical uterine myoma of 15 mm postero-fundic lateralized to the right (type 3), Intra-cavitary polyp of 7 mm, Pauci-follicular right ovary of 29x20 mm, Absence of visualization of the left ovary, Absence of visualization of the appendix. Ascites of great abundance septateencapsulating compatible with pseudomyxoma peritonei.

Tumor markers from 5th November 2023: CA125 rising from 24 to 162 U/ml and, CA19.9 at 5 U/ml.

Given the patient's context, a laparotomy: hysterectomy + bilateral adnexectomy + appendectomy + multiple biopsies + cytological analysis was indicated.

Surgical exploration revealed a gelatinous ascites of great abundance (figure A) estimated at 5 liters, a septate mass measuring 25 cm in diameter (figuresB, C) at the expense of the left ovary adhering to the sigmoid colon with liquid flowing into the Douglas fold, presence of another mass of the same appearance in the distal end of the appendix (figure D).

Surgery consisted of: release of adhesions, removal of the appendix + hysterectomy + bilateral adnexectomy + multiple biopsies. The aftermath of the operation posed no immediate compliations. The anatomopathological study was in favor of mucinous adenocarcinoma of the infiltrating intestinal type.

Discussion:-

The term pseudomyxoma peritonei (PMP) corresponds to an anatomo-clinical entity without direct indication of its origin, characterized by an effusion of variable abundance, of viscous or mucinous appearance, in the peritoneal cavity associated or not with epithelial cells, [1]. The origin is appendicular in at least 90% of cases, secondary to a mucinous tumor of the appendix (LAMN) ruptured into the free peritoneum [2], Other causes of PMP can be an ovarian tumor (teratoma)[3], in our case it’s due to a highly malignant tumor: infiltrative intestinal type mucinous adenocarcinoma.

Intestinal type mucinous adenocarcinoma is an uncommon type of primary ovarian tumor that accounts for approximately 2.4% of ovarian carcinomas [4]. The pattern of invasion of the ovarian stroma can be classified into two groups: expansive and infiltrative invasion; however, a mixture of the two patterns is commonly found. Studies show that the infiltrative pattern occurs in approximately 44–63% of cases and is related to increased disease aggressiveness and mortality [5-6]. In the present report, primary intestinal-type mucinous adenocarcinoma of the ovary was diagnosed on the basis of its characteristics as a large, multilocular, and unilateral tumor [5-7-8], and on the histological and immunohistochemical study of the surgical specimen, positive for CK7, CK20 markers, which indicated the primary location of the tumor [5-7-9-10] after having cleared the digestive tube by colonoscopy and endoscopy which was without abnormalities. There are 02 particularities in this case, firstly the association of two very rare entities for which we have not found support in the literature. Secondly the difficulty of making a radiological diagnosis either by CT or MRI which makes the Surgical management essential in the early stage and in metastatic disease. Chemotherapy is usually administered for stage II MOC and beyond. Prognosis is better in early on in the disease, but worse in the advanced stages [5-6]. Our patient was diagnosed with primary ovarian cancer with intestinal metastasis classified IIIc by FIGO and referred for chemotherapy.

Conclusion:-

According to our case the PMP can be due to rare invasive tumors of the ovary and not only the teratoma. The dilemma here is to know how to make a diagnosis of (MOC) correctly based on radiology something which was difficult in our case given that the absence of visualization of the ovary, the appendix and the digestive tumor on the MRI. However, the anatomopathological and histological study of the surgical specimen is the gold standard for accurate diagnosis and determination of primary tumor location and its classification.

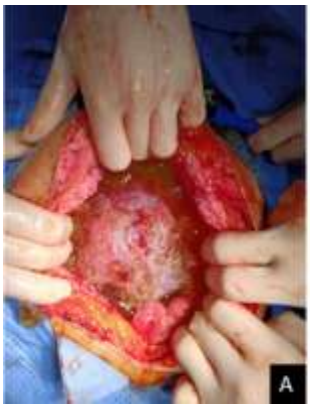


Figure A

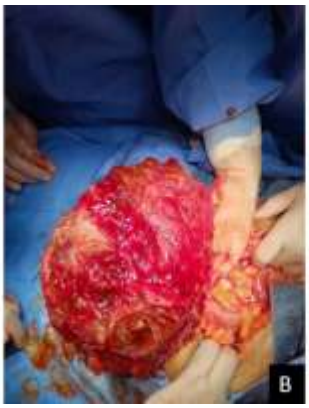


Figure B

Figure C

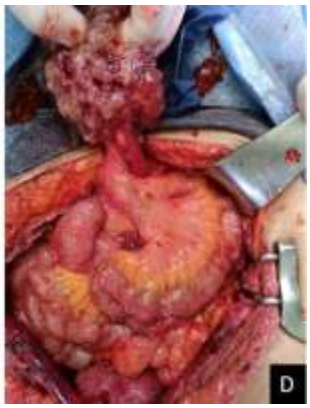


Figure D

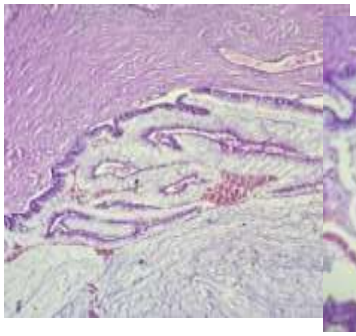


Figure 1

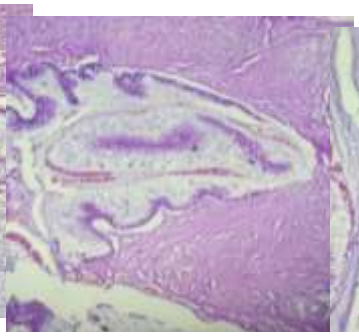


Figure 2

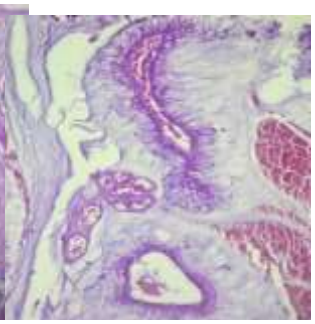
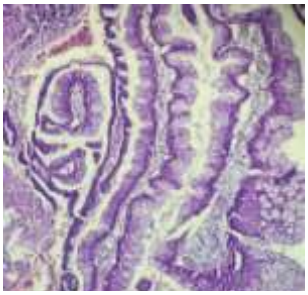


Figure 3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Authors' contributions

The authors declare no conflict of interest.

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