Acta Scientific MEDICAL SCIENCES (ISSN: 2582-0931)

Volume 7 Issue 5 May 2023

Case Report

Peritoneal Adenomucinosis of Dual Ovarian and Appendicular Origin: Myth or Reality?

Ramzi Ghorbel, Amina Mnejja*, Nadia Ben Said, Sahbi Kebaili and Kais Chaabene

Department of Gynecology Obstetrics, Maternity of Sfax, Tunisia

***Corresponding Author:** Amina Mnejja, Department of Gynecology Obstetrics, Maternity of Sfax, Tunisia.

Received: February 07, 2023

Published: April 17, 2023

Abstract

Pseudomyxoma or peritoneal adenomucinosis is a rare condition.

Its incidence is estimated at 1 to 2 cases per year per million inhabitants, with a female predominance [1].

The origin of pseudomyxoma peritonei remains controversial.

Thanks to immunohistochemistry and molecular engineering, it is accepted that the origin is appendicular in about 90% of cases [2,3].

However, mature cystic teratomas associated with borderline ovarian mucinous tumours represent an exception to this rule [4].

Keywords: Pseudo Myxoma; Peritoneal Adenomucinosis; Ovary; Appendix; Surgery; Chemotherapy

Case Report

Patient AB, 52 years old, with a history of asthma under medical treatment, urogenital prolapse operated by promonto fixation, simple mucinous cyst of the right ovary operated with right adnexectomy, 5th gesture, 5th pare as well as 5 vaginal deliveries.

She consulted our emergency room for pain in the left iliac fossa.

On examination there was a tender mass in the left iliac fossa with multiple peritoneal carcinosis nodules.

Pelvic ultrasound revealed a 7 centimetres long solid cystic left ovarian mass with nodules of peritoneal carcinosis.

Pelvic magnetic resonance imaging revealed a well-limited mixed left ovarian latero uterine mass with two components, one with intermediate signal in T2, hypo signal in T1, discrete hyper signal in diffusion, enhancing in the periphery, measuring 94*70 millimetres in the sagittal plane and measuring 85*68 millimetres in the sagittal plane.

The second component, lateralized on the left, is T1 hypo signal, T2 frank hyper signal and diffusion hypo signal. It is the site of fine partitions in T2 hypo signal with micro punctate enhancement measuring 90*70 mm in the sagittal plane.

Thickening of the peritoneal layers associated with fat infiltration in the parieto-colonic gutters.

A small amount of effusion in the douglas and perihepatic pouch.

No iliac or lumbo-aortic adenomegaly.

Absence of bone lesions.

A suggestive aspect of left ovarian epithelial tumour of borderline type associated with peritoneal thickening.

The patient underwent an exploratory laparotomy of the left ovarian mass with peritoneal cytology and left adnexectomy without tumour invasion with epiploic biopsy and biopsy of the right and left parietal and colonic gutters and biopsy of the nodules of peritoneal carcinosis at the level of the douglas cul de sac.

Per operative discovery of a moderate amount of ascites, an enormous abdominal and pelvic mass developed at the expense of the left ovary, 18 centimetres long, mixed solid and cystic with the presence of exocystic vegetations.

Diffuse peritoneal carcinosis, particularly in the douglas cul de sac and the pre-vesical space.

Epiploic infiltration with an epiploic cake appearance.

Extemporaneous pathological examination: a borderline mucinous tumour of the left ovary.

In view of the extensive peritoneal carcinosis, the epiploic infiltration and the macroscopically suspicious appearance of malignancy, we decided to wait for the definitive histological result.

The definitive pathological examination:

Left adnexectomy specimen

- Borderline mucinous tumour of the left ovary with the presence of acellular mucus in the meso ovari and meso salpinx.
- Right and left parieto-colonial biopsy: no abnormality.
- Epiploic biopsy and a nodule of the douglas cul de sac: presence of acellular mucus (peritoneal adenomucinosis).
- Then the conduct was to do a fibroscopy oeso gastro duodenal and colonoscopy and total hysterectomy and appendectomy and omentectomy.

FOGD

- Moderate congestive antral gastropathy and sessile micropole of the greater antral curvature.
- An excisional biopsy done.

Histology

- Florida chronic antral gastritis with heicobacter pyolri without intestinal metaplasia or atrophy.
- Additional lesions of chronic gastritis.

Colonoscopy

- Presence of micro polyp of the recto sigmoidal junction of 5 millimeters in size.
- A biopsy was performed.

Histology

- Low grade (moderate) dysplastic tubular adenoma of the recto sigmoidal junction.
- The operative procedure was a total hysterectomy with appendectomy and omentectomy.

Histology

- A low grade mucinous appendiceal lesion with tumor surgical border.
- Low grade peritoneal mucinous carcinosis.
- Atrophic uterus.
- Endo cervical inflammatory polyp.
- After histological findings the course of action was a right colectomy and hyperthermic intraperitoneal chemotherapy.
- Simple postoperative course.

Discussion

Pseudomyxoma peritonei is rare, first described by R. Wyerth in 1884. He described the accumulation of extracellular mucin in the peritoneal cavity with the presence of an ovarian mucinous tumour [5]. Then in 1901, Frankel associated it with the presence of an appendicular mucocele [6].

The origin of pseudomyxoma peritonei in women is a controversial subject. It has been shown by immunohistochemical studies of CK7, CK20, and HAM-56 antigens that the origin of pseudomyxoma is most often appendicular and not ovarian [7].

This has been reinforced recently by molecular biology data concluding that the over-expression of the MUC-2 gene in pseudomyxoma is a consequence of the presence of gram-negative bacteria (from appendiceal perforation). This over-expression is correlated with germ density and a poor prognosis [2].

Mutations in the K-ras gene and loss of alleles at chromosomes 18q, 17p, 5q and 6q are found in pseudomyxoma peritoneum and are not found in true borderline ovarian tumours [3].

Ovarian mucinous tumours, considered to be the origin of pseudomyxomas, may give peritoneal tumour implants called peritoneal carcinosis, but not true pseudomyxoma. The only primary ovarian tumours capable of genuine pseudomyxomatous dissemination would be mature cystic teratomas, probably related to the existence of a gastrointestinal contingent in these embryonic tumours [4].

The association of peritoneal pseudomyxoma with ovarian mature cystic teratomas is found in 3 to 8% of cases [8].

Two recent studies, one of which concerned mucinous tumours of the ovary of the intestinal type, reported 3 cases of peritoneal pseudomyxoma associated with dermoid cyst of the ovary [9].

Two cases had a normal appendix, with no evidence of recurrence after 5 and 16 years.

The second study described three cases of mucinous tumours of the ovary with cystic teratomas [10]; the appendix was normal and the mucinous tumours were CK20 positive and CK7 negative. Peritoneal lesions were in favour of disseminated peritoneal adenomucinosis in two cases and intermediate in the third. No follow-up data.

In general, ovarian mucinous tumours are usually CK7 positive with variable expression of CK20, in contrast to those associated with mature teratomas, which are CK20+/CK7- [10].

The treatment of peritoneal pseudomyxoma is not yet standardised and no data in the literature allow clear conclusions to be drawn. Complete removal by cyto reduction surgery followed by intraperitoneal hyperthermic chemotherapy is considered the therapeutic «gold standard» [8].

No intraperitoneal recurrence has been reported for pseudomyxoma peritoneum of ovarian origin [9].

Adjuvant chemotherapy with 6 cycles of paclitaxel and carboplatin has been described in clinical cases [4,8]; however, no study has demonstrated the benefit of this chemotherapy [4].

Thus, our observation is a rare case of peritoneal pseudomyxoma resulting from a metastatic mucinous appendicular tumour with a secondary non-primary ovarian borderline mucinous tumour.

Conclusion

Peritoneal pseudomyxoma is of appendicular origin in the majority of cases but its ovarian origin is likely when the appendix is normal. The curative treatment remains cytoreduction followed by hyperthermic intraperitoneal chemotherapy (gold standard).

Bibliography

1. Smeenk RM., *et al*. "Appendiceal neoplasms and pseudomyxoma peritonei: a population based study". *EJSO* 34.2 (2008): 196-201.

2. O'Connell JT., *et al*. "Pseudomyxoma peritonei is a disease of MUC2-expressing goblet cells". *American Journal of Pathology* 161.2 (2002): 551-564.

3. Szych C., *et al*. "Molecular genetic evidence supporting the clonality and appendiceal origin of pseudomyxoma peritonei in women". *American Journal of Pathology* 154.6 (1999): 1849-1855.

4. Manmeet Saluja., *et al*. "Pseudomyxoma Peritonei arising from a mucinous borderline ovarian tumour: Case report and literature review". *Australian and New Zealand Journal of Obstetrics and Gynaecology* 50.4 (2010): 399-403.

5. Werth R. "Klinische und anatomische untersuchungen zur lehre von den bauchgeschwuelsten und der laparotomie". *Archives of Gynecology and Obstetrics* 24 (1884): 100-118.

6. Frankel E. "Uber das sogenannte pseudomyxoma peritonei". *Medizinische Wochenschrift* 48 (1901): 965-970.

7. Ronnett BM., *et al*. "Immunohistochemical evidence supporting the appendiceal origin of pseudomyxoma in women". *International Journal of Gynecological Pathology* 16.1 (1997): 1-9.

8. Hwang JH., *et al*. "Borderline-like mucinous tumor arising in mature cystic teratoma of the ovary associated with pseudomyxoma peritonei". *International Journal of Gynecological Pathology* 28.4 (2009): 376-380.

9. Lee KR and Scully RE. "Mucinous tumors of the ovary: a clinicopathologic study of 196 borderline tumors (of intestinal type) and carcinomas, including an evaluation of 11 cases with 'pseudomyxoma peritonei". *The American Journal of Surgical Pathology* 24.11 (2000): 1447-1464.

10. Ronnett BM and Seidman JD. "Mucinous tumors arising in ovarian mature cystic teratomas: relationship to the clinical syndrome of pseudomyxoma peritonei". *The American Journal of Surgical Pathology* 27.5 (2003): 650-657.