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Case Report

Mucinous Ovarian Adenocarcinoma cloaked as an intestinal Tumor-Case Report

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Abstract

Ovarian mucinous neoplasia are extremely heterogeneous tumors in morphology and clinical presentation, being a setback for the appropriate diagnosis. Cross-reactions and immunohistochemical ambiguity are adversities that limit accuracy, leading to the need for greater resourcefulness in the management of these patients. Because it is a neoplasm with a high probability of becoming invasive, surgical excision is usually the most assertive alternative.

Keywords: Adenocarcinoma; Mucinous; Intestine; Ovary

Introduction

Mucinous ovarian tumors are among the most challenging for pathologists to interpret, primarily because a significant portion of their presentations are imprecise, and they can even exhibit concurrent other subtypes [5]. As it commonly presents asymptomatically, approximately 75% of women have the disease in an advanced state at the time of diagnosis, resulting in a high mortality rate [3].

The literature also reports that it is possible for a significant number of mucinous carcinomas, primarily of intestinal and pancreatic origin, to spread to the ovaries and produce cystic masses that, both macroscopically and microscopically, mimic the genesis of primary ovarian tumors [5]. It is crucial to search for other tumor sites when this neoplasm is present.

Approximately 90% of malignant ovarian tumors are of epithelial origin, which, in turn, can be histologically differentiated into six types: serous (75%), mucinous (20%), endometrioid (2%), clear cell (2%), Brenner, and undifferentiated (1%) [18]. Due to their histological glandular differentiation similar to gastrointestinal tissue, mucinous ovarian adenocarcinomas tend to be referred to as invasive mucinous carcinomas of the intestinal type. However, it should not be interpreted as a primary intestinal origin site.

Due to its rarer nature, the prevalence and incidence vary depending on the type of study, but it is observed that this neoplasm mainly affects women in their 4th and 5th decades of life [3-5], with an incidence of 3% [10]. In an epidemiological study conducted using the Surveillance, Epidemiology, and End Results (SEER) database, it was revealed that out of a total of 40,571 women with epithelial ovarian neoplasms, only 4,811 (11.9%) had the mucinous subtype [17].

The objective of this work is to report a case of primary mucinous ovarian adenocarcinoma with immunohistochemical expression in only 2% of cases.

Case Report

A 41-year-old female patient was referred from a primary care clinic to the gastroenterology outpatient clinic with a complaint of severe, daily abdominal pain for about 3 months. The patient also

reported a progressive increase in abdominal size over the past 4 months, as well as nausea, frequent vomiting, and a change in bowel habits. She denied weight loss or urinary symptoms. As for her medical history, she reported having arrhythmia, hypothyroidism, dyslipidemia, depressive-anxiety disorder, and panic disorder. Her surgical history included a cesarean section 18 years ago and a decompressive laminectomy 10 years ago. She was on continuous medication, taking atenolol, simvastatin, fluoxetine, lithium, valproic acid, and chlorpromazine. On the day of the clinical examination, she brought a computed tomography (CT) scan from 3 months earlier, which showed a large mass formation in the lower abdomen measuring 164 x 225 x 193mm with septation and mild peripheral contrast enhancement. There were no cleavage planes with the mesentery, which could suggest a mesenteric cyst. On general physical examination, she appeared to be in good overall condition, well-hydrated, with a normal complexion, and without jaundice. However, on abdominal examination, her abdomen was distended and tense due to the lesion, which was palpable in all quadrants of the abdomen. There was also tenderness upon palpation, but no rebound tenderness.

A median transumbilical longitudinal exploratory laparotomy was indicated for the removal of the tumor. During the intraoperative procedure, ascitic fluid was observed (collected and sent for oncotic cytology examination). An extensive mass lesion with well-defined borders and of ovarian origin was identified on the left side, but with adhesions to the omentum, small intestine, and ovary. Left oophorectomy and salpingectomy were performed in conjunction with the excision of the tumor, which weighed approximately 10 kilograms. An inventory of the cavity was conducted, and the colonic and small bowel loops were assessed, showing no abnormalities.

In the immediate postoperative period, the patient was in the Intensive Care Unit (ICU) and was doing well, with only slight complaints of operative wound pain. There was a decrease in bowel sounds, and a moderate degree of pallor (+/4+). Considering the extent of the surgery, it was decided to provide analgesia through a peridural catheter with a solution of 2ml (1mg of morphine + 19mg of 1% lidocaine without a vasoconstrictor) + 3ml of 0.9% saline, without any complications. On the 2nd day post-surgery, the operative wound appeared to be in good condition, intact, and without active discharge of secretions. There were no signs of inflammation, and the patient was allowed to start a clear liquid diet (water, tea, and gelatin) and was encouraged to start walking. By

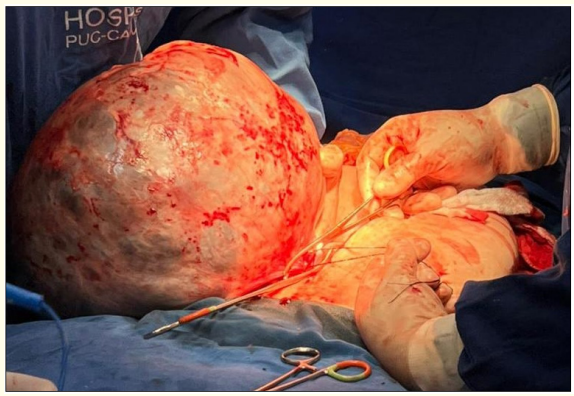

Figure 1

Figure 2

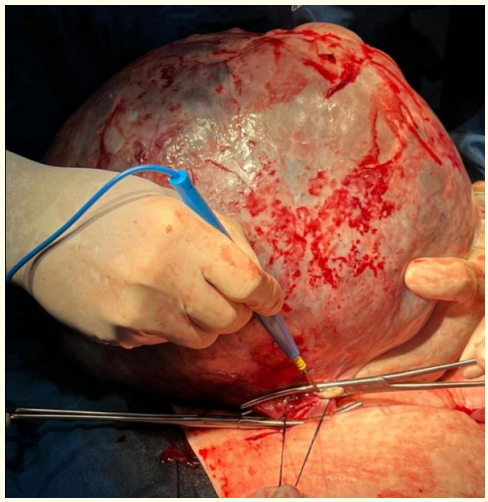

Figure 3

the 4th day post-surgery, the patient was in good general condition, without nausea or vomiting, and had good tolerance for a soft diet. She was then transferred to the regular hospital ward. On the 6th day post-surgery, the patient continued to progress well, had good tolerance for a regular diet, and was discharged on the 8th day of the postoperative period.

The anatomopathological examination revealed that the lesion had a weight of 10,340g and dimensions of 30.0 x 29.0 x 16.0cm with a smooth and intact external surface and a internal solid aspect. The histological examination indicated that it was a tubulopapillary adenocarcinoma lined by stratified epithelium with rounded, hyperchromatic nuclei and prominent nucleoli. There was a loss of basal polarity associated with a multilocular and solid component. However, the positive staining for immunohistochemical markers such as RE (estrogen receptor), CK20, RP (progesterone receptor), CK7, Ki67, and CEA suggested that the neoplasm might have its primary site in the gastrointestinal tract.

With this information in hand, the patient underwent colonoscopy and endoscopy in search of the primary site, but without success, leading to the conclusion that it was a primary mucinous ovarian adenocarcinoma with expression in only 2% of cases. Furthermore, in a new immunohistochemical study, the neoplasm exhibited positivity for CDX2, which further supported the ovarian primary site.

Discussion

Mucinous ovarian tumors can be classified as benign (-oma), borderline (pseudo-), or malignant (-carcinoma) and can also be categorized as invasive or non-invasive [10]. Invasive mucinous carcinomas are very uncommon primary ovarian tumors, provided that cases of metastasis or pseudomyxomas of intestinal origin are rigorously excluded. In retrospective studies, within the total number of mucinous ovarian tumors, researchers have found rates as low as 2.4-3%, constituting primary invasive tumors confined to the ovary at the time of diagnosis [5,6,9].

In terms of their morphological composition, mucinous adenocarcinomas tend to present with solid areas and firm nodules, but only 4% are predominantly or entirely solid, while the majority (75%) have a cystic or semi-cystic appearance [5]. This data highlights the uniqueness and heterogeneity of the neoplasm type in the presented case and its significance in academic discourse. Furthermore, the solid appearance also raised suspicions of an intestinal tumor during clinical investigation. Regarding its morphology, the average size of mucinous ovarian carcinomas is approximately 16cm [4], whereas our neoplasm measures twice that length in its longest dimension.

Metastasis to pelvic lymph nodes is rare, and the recurrence of the neoplasm does not appear to be associated with pelvic lymphadenectomy [4]. In cases of metastasis, the first sites of implantation are typically the peritoneum, omentum, and liver. In contrast, colorectal adenocarcinoma is known for having one of its sites of metastasis being the ovary [5].

Despite the diagnostic challenges, another piece of data supporting the ovarian primary site is that primary mucinous ovarian tumors tend to be larger (16-20cm) and unilateral, whereas metastatic tumors tend to be smaller (3-4cm) and bilateral [6]. However, metastases originating from the pancreas are an exception, forming larger, cystic masses. Based on these observations from retrospective studies analyzing medical records and pathological reports, a simple algorithm using size and laterality can be highly useful in determining the likelihood that a mucinous carcinoma in the ovary is primary or metastatic. The algorithm designates unilateral tumors > 10 cm as primary and categorizes all other tumors (including all bilateral tumors and unilateral tumors < 10cm) as metastatic [6].

Metastatic colorectal carcinoma can be very histologically similar to primary mucinous ovarian adenocarcinoma, precisely because it can simulate its morphology [9]. Distinguishing these two entities can be quite challenging from the pathologist's perspective. In terms of immunohistochemical markers and diagnosis, the literature reports that CK20 is negative in 90-98% of cases of primary mucinous ovarian adenocarcinoma [8,9]. In contrast, the case reported in this work exhibits expression of both CK20 and CK7, which can complicate the diagnosis. It is at this point that pathology, in collaboration with clinical findings, should intervene to search for other possible primary sites. Given that the intestinal site is one of the most common and major possibilities for the origin of metastases, endoscopy and colonoscopy were requested, and they returned without any clear signs of primary sites, ultimately confirming the diagnosis of primary mucinous ovarian adenocarcinoma [10].

Furthermore, another finding supporting the diagnosis is the positivity of the immunohistochemical markers Cd x 2 along with CK7. According to the literature, Cd x 2 is a highly sensitive and

reliable marker for metastatic colorectal carcinoma to the ovary, which is more specific than CK20. Using a limited panel consisting of Cd x 2 and CK7 can help distinguish metastatic colonic carcinomas in the ovaries, which typically present with Cd x 2 positive and CK7 negative staining, from mucinous adenocarcinomas, which exhibit positivity for both Cd x 2 and CK7 [9].

Conclusion

Primary mucinous ovarian adenocarcinoma stands out as a clinical entity not only due to its rarity and low incidence but also because it has historically presented a challenge in achieving an accurate diagnosis. It is a cancer for which the only potentially curative and favorable prognosis approach is surgery. The patient's medical history combined with the findings from imaging studies were crucial factors in achieving an accurate diagnosis and led to the appropriate procedure. Therefore, it is a disease that demands multi and interdisciplinary care, involving surgical, radiological, and pathological teams to provide comprehensive patient care.

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