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CASE REPORT

High Grade Appendiceal Mucinous Neoplasma Reviewing Primary Tube Malignancy : A Case Report

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Abstract

Appendiceal mucinous neoplasms (AMNs) are rare tumors accounting for less than 1% of all cancers. The clinical presentations of AMN vary significantly and the diagnosis of most patients is during surgery for suspected appendicitis, peritonitis or gynecological cancer. There were many case reports AMN mimicking an ovarian cancer. In this case, we reported a 64-year postmenopause woman with diagnosed peritonum carcinomatosis dd/ ovarian cancer and it actually AMN in RSUP dr. M. Djamil Padang, West Sumatera. The diagnosis is estimated by ultrasonography (USG), computed tomography (CT), magnetic resonance imaging (MRI) and diagnostic laparoscopy followed by histopathologic verification. The evaluation of the prognosis of appendix mucinous adenocarcinoma mainly depends on whether the tumour is in an advanced stage, the degree of malignancy, and whether an AMN is formed

Keywords:

INTRODUCTION

Appendiceal mucinous neoplasms (AMNs) are rare tumors that account for less than 1% of all cancers and are occasionally discovered incidentally, during follow-up or at the time of surgery for other causes, and are often diagnosed at a late stage. significantly, and is asymptomatic in 25% of cases. 3 An acute appendicitis-like presentation with right lower quadrant pain secondary to distension of the appendix by mucins is the most common clinical presentation in the early stages of disease. Advanced disease presents with an increase in abdominal circumference due to accumulation of mucinous ascites in the peritoneum. Other clinical presentations for this stage include chronic abdominal pain, weight loss, anemia, infertility, and new-onset umbilical or inguinal hernias.

The diagnosis of most patients is intraoperative for suspected appendicitis, peritonitis or gynecological cancer. 4,5 Preoperative diagnosis by ultrasonography (USG), computed tomography (CT), magnetic resonance imaging (MRI) and diagnostic laparoscopy followed by histopathological verification. In gynecology, appendix neoplasm (AN) is frequently misdiagnosed as a gynecological tumor. 6 Here, we report a case of AMN that mimics primary cancer of the fallopian tube.

CASE REPORT

A 64 year old postmenopausal woman (gravida 6, para 6), was referred from Pariaman Hospital because she was suspected of having ovarian cancer with a history of enlarged abdomen since 1 year ago and getting bigger since 3 months ago. This complaint is followed by abdominal pain. There was no vaginal bleeding, bleeding after intercourse, dyspareunia, leukorrhea, or history of trauma. There was no history of hypertension, diabetes, heart, lung, liver and kidney disease. There is no history of previous surgery. On physical examination, he was hemodynamically stable and abdominal distention accompanied by abdominal pain. From normal genital examination. Abdominal ultrasonography shows large amounts of free fluid in the adnexa to subhepatic with a fibrin component in between (Fig.1).



Figure 1: Ultrasound shows large amounts of free fluid in the adnexa

Laboratory examination results obtained total protein (8.9 g/dL), globulin (5.3 g/dL) and creatinine (1.3 mg/dL) patients were higher than the normal range, thrombocytosis (611,000/mm³), and albumin (3.6 g/dL) were lower than the normal range. Laboratory examination for tumor markers CA 19.9 was normal (5.19 /mL) and CA-125 was normal (14.10 /mL). Computed tomography shows the presence of a number large amount of free fluid of inhomogeneous density and several hypodense lesions in the pelvic cavity (Fig. 2).



Figure 2: CT scan showing large amounts of free fluid and multiple lesions in the pelvic cavity

From the initial examination the patient was diagnosed with suspected primary peritoneal carcinomatosis and a differential diagnosis with ovarian cancer. The patient underwent exploratory laparotomy, intraoperatively found mucinous gelatinous fluid that filled the abdominopelvic cavity with multiple miliary lesions throughout the peritoneum, colon, ileum with the largest main mass seen in both fallopian tubes which were tubular enlarged and fragile. Both ovaries of normal shape and size with surface metastatic lesions. The appendix also appears enlarged and fragile. Clinical impression Advanced stage of primary tubal cancer with peritoneal carcinoma it was decided to debulk.

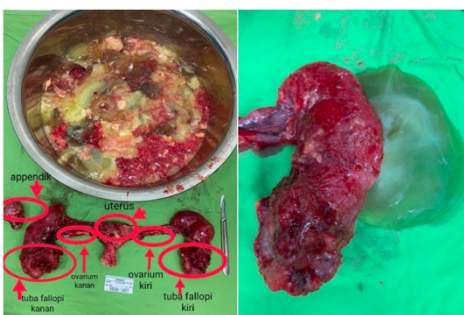


Figure 3. Surgically resected specimen

Postoperative pathology is as follows: (uterus, adnexa and appendix)



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High-grade mucinous neoplasm of the appendix with ovarian and uterine infiltration, which was not consistent with the initial intraoperative suspicion. The patient was given adjuvant chemotherapy adjuvant therapy.

DISCUSSION

Appendix neoplasms are included in the group of neoplasms with the lowest incidence rate among all gastrointestinal tumors. In all specimens after appendectomy, the tumor accounted for about 0.7 to 1.4%, of which the incidence of mucinous adenocarcinoma of the appendix accounts for 0.01% ~ 0.08%.⁷ In terms of demographics, the current patient is a 64 year old female. The peak age of onset is between 50 and 60 years, however, it can occur at any age.^{3,7,8}

Most patients are women; its occurrence may be related to long-term inflammation and infiltration of the appendix. Also, patients with familial adenomatous polyposis and KRAS,GNAS,TP53 mutations have an increased risk of developing mucinous adenocarcinoma of the appendix. KRAS mutations were distributed among low- and high-grade pseudomyxoma peritonei (PMP). It has been observed in 70% of patients with appendiceal adenomas.⁹⁻¹¹

Appendix tumor clinical manifestations are usually acute appendicitis, abdominal masses and intestinal obstruction.¹² There are many case reports that describe AMN that resembles ovarian tumors. Symptoms that may arise from AMN range from lower abdominal or flank pain, fever, nausea, and vomiting to an asymptomatic presentation.^{8,13} Invasive appendix adenocarcinoma represents only 4-6% of appendix cancer and primary diagnosis is difficult due to non-specific clinical presentation.¹² In our case, symptoms such as abdominal pain and abdominal distention were reported on admission.

Tumor markers of mucinous adenocarcinoma of the appendix can increase, such as CA 125, CA 199 and CEA. Preoperative tumor markers were more consistent with the TNM staging.⁷ However, in this case the preoperative tumor marker examination was normal. Imaging examination is an important detection method in the diagnosis of mucinous appendix adenocarcinoma. Colonoscopy can be used to perform a biopsy of the diseased part, and the diagnosis can be confirmed by pathological examination. MRI and PET-CT combine the metabolic state of the lesion with its structure

anatomy, which is useful for increasing tumor detection rates and guiding the development of diagnosis and treatment plans.⁷ Late diagnosis of a mucinous variant can lead to peritoneal perforation and spread leading to mucinous ascites associated with Pseudomyxoma peritonei. This type of manifestation is usually treated by peritoneal resection and intraoperative peritoneal administration of mitomycin followed by intravenous administration of 5-fluoracil for 4-5 days.

It is recommended that women with menopausal mucinous appendix adenocarcinoma undergo menopause before the tumor is removed along with the ovary, which can prevent metastases and increase survival rates.⁷ The prognostic evaluation of appendix mucinous

adenocarcinoma mainly depends on whether the tumor is at an advanced stage, the degree of malignancy, and whether AMN is formed. . Spread of mucus beyond the right lower quadrant is an independent factor contributing to poor disease prognosis. Formation of a peritoneal pseudomyxoma and morphological evidence of suspected infiltration around the abdominal organs all imply a poor prognosis. If the patient has mild clinical symptoms and there is no obvious infiltration or spread of the surrounding tissue, complete removal of the appendix tissue can significantly prolong life.

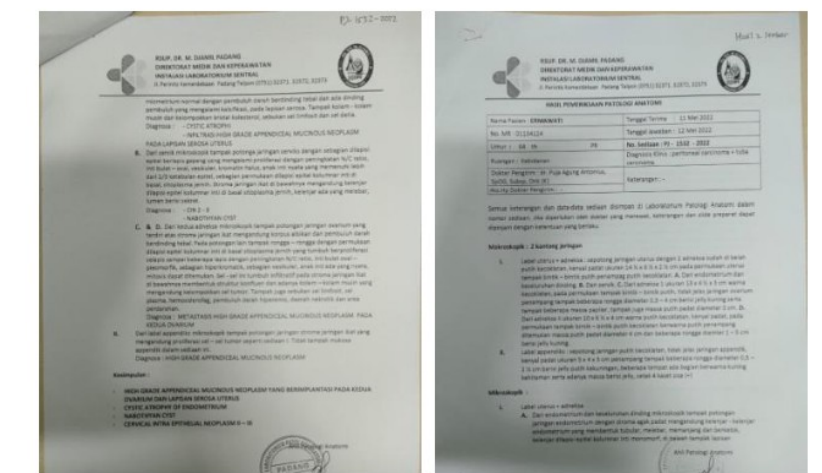


Figure 4. Anatomical Pathology Examination Results

CONCLUSION

AMN in combination with PMP is a rare occurrence, which can mimic tubo-ovarian cancer. Differential diagnosis with tumors of the digestive organs should be considered for patients especially those with malignant mucinous masses in the internal genital organs.

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