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Case Report

Mucinous carcinoma of the appendix: a case report

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ABSTRACT

Mucinous carcinoma of the appendix is a rare malignant tumor of the appendix. Incidence ranges about 0.01-0.2% in cases of malignancy in the gastrointestinal tract. Clinically, the symptoms appear to resemble acute appendicitis. The diagnosis was established at intraoperative time. Studies on this case are still few. A 69-year-old man complained of lower right abdominal pain since 6 days before going to Sanjiwani hospital. The patient also complained of fever, nausea, decreased appetite and constipation. Examination results temperature 37.6 ^{0}C, McBurney sign (+), rebound tenderness (+), localized defense on lower right abdomen. Ultrasound examination of the abdomen with an impression of the end tubular structure, a peristaltic diameter of 0.87 cm accompanied by a fluid collection, suspicious of appendicitis with perforation. Inflamed appendix and localized mucin were found at ileocaecal junction intraoperatively. The patient underwent a right hemicolectomy. Histopathological examination revealed a morphological picture suitable for mucinous carcinoma of the appendix. Mucinous carcinoma of the appendix is a rare malignant tumor of appendix. Patients can have the same symptoms as appendicitis. Ultrasound investigations can find a picture like appendicitis. Mucinous carcinoma of the appendix is more frequently found intraoperatively. Histopathological examination is necessary to determine a definitive diagnosis of mucinous carcinoma of the appendix. In high grade appendiceal mucinous neoplasm (HAMN) a right hemicolectomy can be performed and continued with adjuvant chemotherapy.

Keywords: Mucinous, Carcinoma, Appendix

INTRODUCTION

Mucinous carcinoma of the appendix is a malignant tumor of the appendix. Other tumors that can be found include neuroendocrine tumors, goblet/ex-goblet cells or composite carcinoids, lymphoma, adenocarcinoma, and lymphoid or mesenchymal sarcoma. Histologically, 65% of appendiceal tumors are of neuroendocrine origin, whereas adenocarcinoma (mucinous, signet ring or non-mucinous) accounts for approximately 20% of these tumors. Mucinous carcinoma of the appendix itself is found in 0.2-0.3% of appendectomy specimens.[1]

Mucinous carcinoma of the appendix is a rare malignancy. The incidence or prevalence ranges from 0.01-0.2% in cases of malignancy in the gastrointestinal tract.[2] This case is also only found in around 1.2 out of 100,000 people in the United States.[3] Cases of mucinous carcinoma of the appendix are also more often found at an average age of 60 years, and the incidence is more common in men.[3,4]

Clinically, symptoms that appear in mucinous carcinoma of the appendix can be found as acute appendicitis (with right lower abdominal pain, nausea, vomiting, fever, decreased appetite) or peritonitis (presence of widespread mucin in the peritoneal cavity), but some patients may show no symptoms at all at the time of diagnosis.[5,6]

The diagnosis of mucinous carcinoma of the appendix can occur on intraoperative findings during an appendectomy

procedure for appendicitis or other indications, and then the characteristic invasive glandular findings with high-grade cytologic atypia and extracellular mucin in more than 50% of the lesions.[7]

Cases of appendiceal carcinoma municus are still very rare, there is still little literature discussing this case, so the author is interested in discussing this case which was also found at Sanjiwani hospital, Bali.

CASE REPORT

A 69-year-old man was brought to the emergency room at Sanjiwani regional hospital with complaints of right lower abdominal pain. The patient had complaints since 6 days before entering the hospital. Complaints appear suddenly and seem to come and go. Complaints are felt to interfere with the patient's activities and sleep. Complaints of stomach pain are initially felt in the pit of the stomach, then spread and the pain persists in the lower right abdomen. The patient went to the surgical clinic and was advised to be hospitalized, but the patient's family refused. The patient said that the complaints had improved after being given medication, but then the complaints came back and got worse so the patient was taken back to Sanjiwani hospital. Other complaints felt by patients include fever, nausea, decreased appetite and not having defecated for 3 days. The patient is a smoker. The patient said he had no history of illness and was not taking routine medication.

The results of the general condition examination showed that the VAS scale was 7 out of 10, compos mentis consciousness, blood pressure 110/80 mmHg, pulse 88 times/minute, respiration rate 20 times/minute, temperature 37.6C, oxygen saturation 99% room air. Physical examination of the head, neck and thorax revealed no abnormalities. Abdominal physical examination did not show any scars. Auscultation revealed good bowel sounds, and percussion revealed tympany. Palpation revealed tenderness in the lower right abdomen (McBurney sign), rebound tenderness examination (+), defans localized in the lower right abdomen, masses, liver and spleen were not palpable. From the results of the history and physical examination, the patient was diagnosed with acute appendicitis, suspicious for perforation. The patient then underwent supporting laboratory tests and an ultrasound examination. From the results of laboratory examination of white blood cells, hemoglobin, and the percentage of neutrophils, eosinophils, basophils within normal limits. Ultrasound examination of the abdomen revealed the impression of an end tubular structure, aperistaltic with a diameter of 0.87 cm accompanied by fluid collection around it, suspicious for the appearance of appendicitis with perforation.

Based on the results of the history, physical and supporting examination, the patient was diagnosed with perforated appendicitis and an exploratory laparotomy was planned for appendicitis. During the exploratory laparotomy, the appendix was found to be inflamed and mucinous fluid was localized at the ileocaecal junction. From the findings obtained, it was decided to perform a right hemicolectomy.

After surgery, the patient was treated in the ICU with parenteral nutrition, antibiotics, analgesics and gastro-protectors, as well as monitoring the patient's hemodynamics. Once stable, the patient was transferred to a regular room, treated for 8 days and discharged in stable condition.

The results of the anatomical pathology show pieces of the resection margin, which consist of the mucosa, submucosa, muscularis and serosa layers. No tumor cells were seen. The appendix tissue appears, containing a tumor mass, consisting of a proliferation of anaplastic epithelial cells, round in shape, relatively monotonous, embedded in a large pool of extracellular mucin. There were 2 cut lymph node tissues that did not contain tumor cells. From the conclusion of the anatomical pathology, the morphological picture is suitable for mucinous carcinoma of the appendix. Next, the patient was planned to undergo adjuvant chemotherapy.

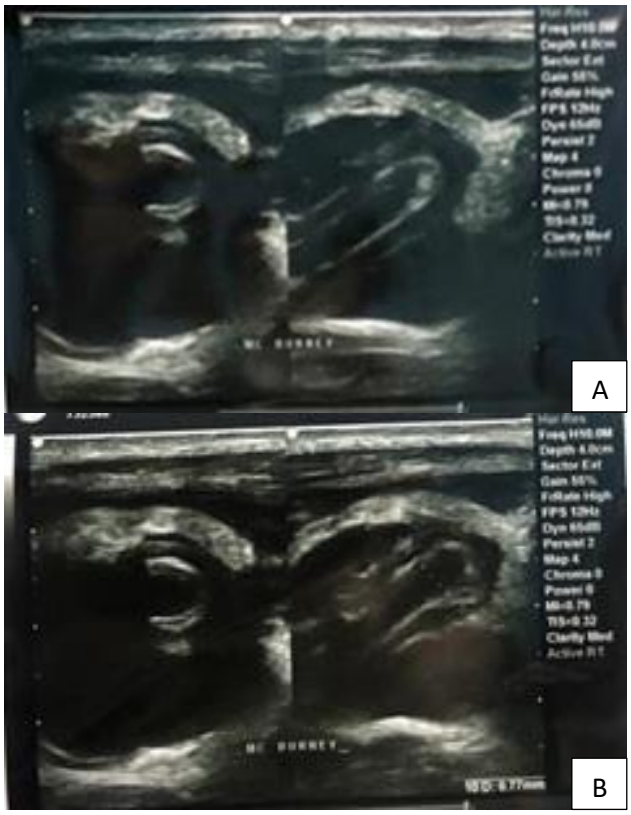

Figure 1 (A and B): Ultrasound of the Mc area of the abdomen, Burney.

DISCUSSION

Mucinous carcinoma of the appendix is a malignant tumor of the appendix. Histologically, 65% of appendiceal tumors are neuroendocrine in origin, whereas adenocarcinoma (mucinous, signet ring or non-mucinous) constitutes approximately 20% of these tumors. Mucinous

carcinoma of the appendix is found in 0.2-0.3% of appendectomy specimens.[1] Cases of mucinous carcinoma of the appendix are rare, but are the second most common malignancy of the appendix.[4]

Risk factors for malignancy in the appendix include smoking, a family history of appendix cancer, patients with a history of atrophic gastritis or pernicious anemia, old age or more than 60 years, and it occurs more often in women.[8] However, there are also studies which state that for malignancy of the appendix, mucinous carcinoma of the appendix, one of the risk factors is male gender.[3,4] From the studies above, risk factors can be found in this patient, namely the patient's age is 69 years or more than 60 years, the patient is also a smoker, and male gender which increases the risk of developing mucinous carcinoma of the appendix.

Mucinous carcinoma of the appendix is often asymptomatic or has nonspecific symptoms. Obstruction of the lumen of the appendix by malignant cells can cause inflammation of the appendix, venous stasis, and infection of the appendix. In mucinous carcinoma of the appendix, there may be cystic dilation of the appendix due to obstruction of the lumen of the appendix by mucoceles. This process is the basis of the most common clinical picture of this disease, acute appendicitis. The most common symptoms include acute or chronic right lower quadrant abdominal pain. As well as patients may present with intermittent colicky pain and gastrointestinal bleeding due to mucoceles intussusception, intestinal obstruction due to mass effect, genitourinary symptoms due to right ureteral obstruction, acute abdominal pain due to appendiceal rupture, or even sepsis. During an abdominal examination, a mass can also be felt.[7] In this case, the symptoms found by the patient were acute appendicitis, the patient experienced persistent pain in the lower right abdomen, nausea, vomiting and loss of appetite, and the patient had not defecated for 3 days, which could be one of the mass effects, namely intestinal obstruction.

Initial evaluation of the case can be found on ultrasonography (USG) examination with a cystic lesion in the right lower quadrant area with an internal appearance like an onion ring depicting multi-layered mucin. Defects in the appendix with search suggest rupture of the appendix. On CT scan or MRI findings, an appendix measuring more than 15 mm can be seen with thickening and irregular walls, which indicates neoplasia. The use of X-ray just doesn't give much results. Measurement of carcinoembryonic antigen (CEA), CA 19-9 and CA 125 levels can be of prognostic value. For a definitive diagnosis, a histology examination is necessary.[9,8]

In this case, the patient underwent an ultrasound examination and an impression of an end tubular structure, aperistaltic diameter, was obtained 0.87 cm accompanied by fluid collection around it is suspicious for appearance of appendicitis with perforation. The patient also underwent a complete blood laboratory examination, but the white blood cell and neutrophil percentages were within normal limits. This is not in accordance with the patient's suspicion of appendicitis where an increase in white blood cell levels can be found with a sensitivity of 91.81%.[10] Then the patient underwent a laparotomy, at which time an inflamed appendix and mucinous fluid were found localized at the ileocaecal junction, which led to malignancy. Then a histopathological examination was carried out and a tumor mass was found, consisting of a proliferation of anaplastic epithelial cells, round in shape, relatively monotonous, embedded in a large pool of extracellular mucin. So, this image establishes a definitive diagnosis as mucinous carcinoma of the appendix. This case shows that malignancy in the appendix is often found during an appendectomy procedure.[3]

Patients with mucinous carcinoma of the appendix often present with rupture of the primary tumor with spread of mucin and tumor cells throughout the peritoneal cavity and is known as pseudomyxoma peritonei (PMP) and is usually diagnosed as appendicitis, peritonitis, or suspected ovarian malignancy. The features of appendix carcinoma can be divided low grade appendiceal mucinous neoplasm (LAMN) and HAMN. In the case of LAMN, it is characterized by cells that are well differentiated, glandular, and produce mucin. Although classically described as “noninvasive” histologically, these cells are still suggestive of PMP and are considered malignant. In cases of HAMN, invasive adenocarcinoma cells can be found which may or may not have a signet ring cell component. high-grade cases show a much more aggressive clinical course than low-grade cases. Patients are more often symptomatic when they present with complaints such as unintentional weight loss, pain, bloating, and intestinal obstruction.[3] In this case, according to the study discussed, the patient can be included in the high level category with the histological findings and symptoms that arise in the patient, namely pain, bloating and constipation indicating the possibility of intestinal obstruction.

Treatment for appendiceal carcinoma can be determined based on the size of the tumor. It is important to visually inspect and palpate the intestines for possible multifocal abnormalities in the intestines. Simple appendectomy can be performed for tumors less than 1 cm in diameter because of the small possibility of lymph node involvement. Among patients with tumors 1 to 2 cm in diameter, right hemicolectomy may be considered for patients with positive margins or deep appendiceal mesoinvasion, higher proliferation rates, or the presence of angioinvasion.[11] For masses larger than 2 cm, right hemicolectomy is recommended. Because of concerns about increased metastatic potential, some studies also recommend right hemicolectomy regardless of tumor size in young patients; carcinoids at the base of the appendix; and/or histopathologic evidence of lymphatic invasion, lymph node involvement, spread to the mesoappendix, positive tumor resection margins, or cellular pleomorphism with a high mitotic index.[12]

In this case, when an exploratory laparotomy was performed, the appendix was found to be around 1 cm in size with an inflamed appendix and mucinous fluid localized at the ileocaecal junction. The findings obtained point to a higher proliferation rate, where the possibility of metastatic nodules occurring in 20% of all cases, so the operator decided to perform a right hemicolectomy, and sent the resection sample to the anatomical pathology laboratory.[3] In this case, after the results of the pathology examination came out, the patient was planned to undergo chemotherapy. Chemotherapy is one of the therapies for HAMN. Recently, The American society of peritoneal surface malignancy (APASMP) recommended the intraperitoneal chemotherapy method for patients with colorectal cancer and also in several centers for patients with appendiceal cancer. Intraperitoneal chemotherapy can be performed intraoperatively, or is called intraoperative hyperthermic intraperitoneal chemotherapy (HIPEC) or post intraperitoneal chemotherapy (EPIC).[13]

The overall prognosis for appendiceal carcinoid is excellent, but 5-year survival is just under 100%.[11] As for colon carcinomas in other locations survival varies with the stage of the disease, with 5-year survival rates estimated at 88% and 78% for local and regional malignancies, respectively, but only 25% for patients with distant (ovarian) metastases.[14] In a study conducted by Yantiss et al of 65 patients with mucinous neoplasms of the appendix, it was found that 96% of patients with extra-appendiceal mucin did not experience worsening (average 52 months), whereas 33% of patients with neoplastic proliferation outside the appendix.[15]

CONCLUSION

Mucinous carcinoma of the appendix is a malignant tumor of the appendix which is rarely found, only ranging from 0.01-0.2% of cases of malignancy in the gastrointestinal tract. This rare case was found at Sanjiwani hospital. Patients with mucinous carcinoma of the appendix can have appendicitis-like symptoms. Supporting ultrasound examination can reveal images such as appendicitis. Appendiceal carcinoma mucinosis is more often found intraoperatively. Then a histopathological examination was carried out to determine a definitive diagnosis of mucinous carcinoma of the appendix. The level of mucinous carcinoma of the appendix is a determinant in further management. In high grade appendiceal mucinous carcinoma or HAMN a right hemicolectomy can be performed and chemotherapy is recommended.

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