

# Splenic metastasis of mucinous appendix adenocarcinoma: Case report

## Abstract

Mucinous appendix tumors are rare, even more a metastatic disease including splenic invasion. We report a case of 69-year old lady diagnosed with CT scan and confirmed intraoperatively with an intraabdominal spread disease, including splenic metastasis without involvement of the ovaries of mucinous appendix adenocarcinoma. Cytoreductive surgery was done with a complement chemotherapy treatment and 3-year follow-up with no recurrence. Most of these cases are incidentally discovered and the histopathologic report is very important to plan the treatment, which is frequently surgical.

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## Introduction

Tumors that produces mucous of the appendix are rare 1-2% of all appendectomy specimen and 0,1% of frequency of appendiceal adenocarcinoma.<sup>1</sup> The mean age at presentation is 60 years old and has no sex predilection.<sup>2</sup> Patients often can present rupture of the primary tumor with spread of mucin and tumor cells resulting in a mucinous ascites known as pseudomyxoma peritonei (PMP),<sup>3</sup> extraperitoneal metastasis is rare that's why peritoneal disease may be considered a regional metastatic site. The diagnosis is made generally in the intraoperative time,<sup>4</sup> but is also seen with a computed tomography scan, the density of mucinous material is similar in density to water.<sup>5</sup>

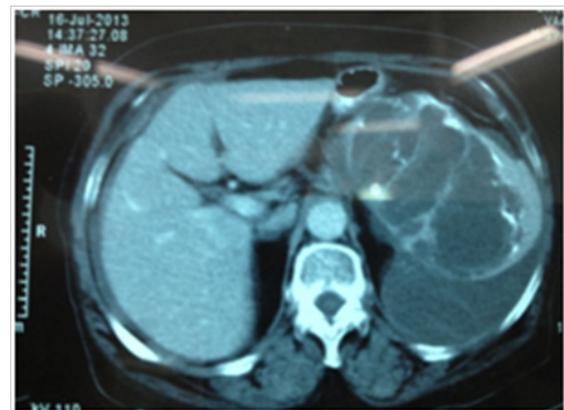
Surgical treatment depends on the dimensions and histology of the mucinous neoplasm as the clinical presentation.<sup>5-7</sup> Laparotomy is advised. All appendectomies must include a wide mesoappendix resection (that includes lymph nodes). For PMP Sugarbaker recommends aggressive peritonectomy with intraoperative hyperthermic intraperitoneal chemotherapy (HIPEC) and postoperative systemic chemotherapy.<sup>8</sup> The 3-5 year survival is 90-95% in patients with only mucinous appendix adenocarcinoma and 10-40% in patients with peritoneal disease. Metastases to the ovaries are frequent, and splenic metastasis were never described in literature.

## Case report

A previously healthy white woman 69-years-old, native of Bragança Paulista, a small district of São Paulo-Brazil, presented to an Emergency Hospital with complaints of abdominal pain; she performed an abdominal ultrasonography giving the diagnosis of cholelithiasis and spleen's cysts. A cholecystectomy was the treatment proposed, but in the intraoperative time they've seen a mucinous secretion and a nodular ovarian's lesion, they only collected samples of the effusion and a biopsy of the ovary; the results of the cytology and pathology were edema and vessels with amorphous basophilic material with no neoplastic cells.

After 15 months, the patient was presented to Heliópolis Hospital in the state of São Paulo-Brazil, with the complaints of abdominal pain and increase of the size of the abdomen; the physical examination showed an abdominal mass of 20 cm underneath the costal ridge. All tumor markers were negative. Abdominal and chest CT scans

showed, splenomegaly with hypoechogenic nodular lesions (Figure 1) and thickened appendix. A laparotomy was indicated and the intraoperative findings were mucin that was collected, thickened appendix, mucocele, splenomegaly with several nodules inside, the report of the frozen biopsy of the appendectomy reveals a Mucinous Neoplasm, so a Debulking Surgery (cytoreduction) (splenectomy, appendectomy, lymphadenectomy, cholecystectomy, bilateral oophorus salpingectomy) (Figures 2-4) because the PMP. The histopathologic report was: Mucinous appendix adenocarcinoma of high grade invading the spleen and the gallbladder's serosal surface (G2 high grade mucinous, moderately differentiated; the ovaries were not involved. Peritonectomy and HIPEC were not performed. She continued the treatment with IV chemotherapy (capecitabine and oxaliplatin (CAPOX)), after 3-years of follow up, there was no evidence of recurrence of the disease.



**Figure 1** Axial contrast-enhanced CT Image reveal hypoechogenic splenic nodular lesions.

## Discussion

This type of tumor is very rare, it presents itself as acute appendicitis commonly, abdominal pain and sometimes is asymptomatic. A surgical evaluation should be recommended for all appendiceal mucoceles more than 2cm in diameter. Metastasis sites are common in the ovaries, splenic metastasis are very rare, and were not described in literature precisely. Sugarbaker surgery is a procedure with a highly

risk of comorbidities, that's why it was not performed in this case. Is important a multidisciplinary team including a radiologist, surgeon, pathologist, internist, etc, for an early diagnosis for optimal results.



**Figure 2** Surgical resection: Gallbladder, spleen, bilateral oophorus salpingectomy.



**Figure 3** Mucinous lesions inside the spleen.



**Figure 4** Appendectomy.

## Conclusion

Tumors confined to the appendiceal mucosa are cured by simple excision, the others are associated with dissemination of the disease. Cytoreductive Surgery and HIPEC are considerate as better treatment to advanced disease.

## Conflicts of interest

None.

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