

Incidental Microscopic Finding Of Acellular Mucin In An Inguinal Hernia Sac As A Warning Sign Of Pseudomyxoma Peritonei

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Citation

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Abstract

Here we describe, an incidental microscopic finding of acellular mucin embedded within the wall of a grossly unremarkable hernia sac specimen in a male patient presenting clinically with an inguinal hernia.

Due to its acellular nature and routine associated clinical history, the presence of microscopic mucin within the hernia sac may be easily dismissed. As the following case illustrates, the finding of acellular mucinous material in a hernia sac specimen can be associated with mucin-producing abdominal malignancy and may be an initial presentation of pseudomyxoma peritonei.

INTRODUCTION

Inguinal hernia is a very common surgical condition occurring in all age groups and both sexes. Microscopic examination of the hernia sac specimen is a part of daily routine in pathology practice. In most instances, there are no significant gross or microscopic findings associated with hernia sac. However, on occasion, one can encounter benign lesions such as mesothelial hyperplasia, embryonal remnants, endometriosis or spermatic cords. Rarely, malignant mesothelioma or metastatic carcinoma may be present.

A particularly problematic finding in an inguinal hernia is presence of acellular mucinous material that can be often interpreted as myxoid and degenerative changes, a finding of no clinical significance.

Here we describe, an incidental finding of microscopic foci of acellular mucin within a grossly unremarkable hernia wall in a 48 year old male patient who presented clinically with a typical inguinal hernia.

Due to an insignificant clinical history and its acellular nature, the presence of microscopic mucin within the hernia sac can be overlooked. The finding of acellular mucinous material can be associated with malignancy within abdominal cavity and can represent a first sign of pseudomyxoma peritonei.

CASE PRESENTATION

The patient is an otherwise-healthy 48-year-old man undergoing herniorrhaphy for an indirect right inguinal hernia. A grossly unremarkable hernia sac and cord lipoma were identified and resected. No free mucinous material was visible grossly within the hernia sac. The specimen was consisted of a cord lipoma measuring 8.0 x 4.0 x 1.5 cm associated with a membranous pink hernia sac measuring 6.5 x 4.5 x 0.8 cm. No significant clinical history other than "right inguinal hernia" was available at the time of microscopic examination. The gross hernia specimen had a smooth and shiny surface without nodularity, induration, hemorrhage, or mucinous change. A single routine section of the cord lipoma was submitted for microscopic examination disclosing mature benign adipose tissue. The routine microscopic hernia sac section showed a mesothelial-lined fibrovascular membrane with diffuse deposits of mucinous/myxoid material present within and expanding slit-like spaces. Non-specific findings between the collections of mucinous material included fibrosis, endothelial proliferation, and chronic inflammation (Fig 1.). There were no intact epithelial layer or free-floating epithelial cells, benign or malignant, associated with the mucinous material. The first impression of the pathologist was that the abnormal findings represented a non-neoplastic mesenchymal proliferative reaction with myxoid degeneration. However, due to the somewhat unusual morphologic features, the entire specimen was submitted for

microscopic examination revealing occasional larger pools and cyst-like collections of mucinous material (Fig 2.) staining with PAS and mucicarmine. No epithelium was identified in association with the mucin in any of the sections examined. These findings raised suspicion for the presence of an occult mucin-producing appendiceal tumor, either adenoma or carcinoma. A follow-up CT scan showed extensive peritoneal “caking,” and subsequent cytoreduction surgery with omentectomy and appendectomy was undertaken. An appendiceal mucinous tumor and extensive peritoneal carcinomatosis (Fig 3.) were noted intraoperatively. The appendectomy specimen microscopically showed large pools of mucin extending into and distorting the bowel wall with a relatively small amount of mildly atypical columnar epithelium with apical cytoplasmic mucin. The findings are consistent with low grade mucinous carcinoma peritonei (MCP-L) [1]. Perforation was not apparent either grossly or microscopically; however the entire omentum was involved with a similar process containing malignant mucinous epithelium. The patient was treated with chemotherapy (intraperitoneal 5FU and intravenous MMC) and did well for 1 year until new pelvic disease reoccurred.

Figure 3

Figure 1: Diffuse deposits of mucinous/myxoid material present within and expanding slit-like spaces within the wall of the hernia sac. Between the collections of mucinous material fibrosis, endothelial proliferation, and chronic inflammatory cells are seen. No epithelial lining is present. H&E (400X)

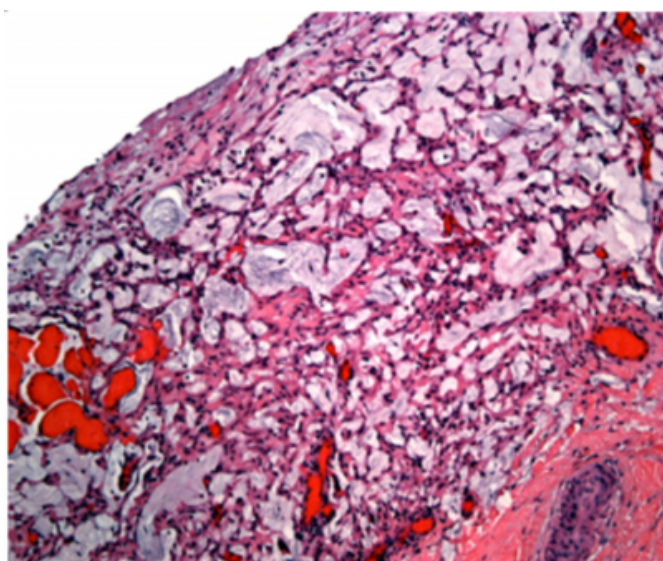
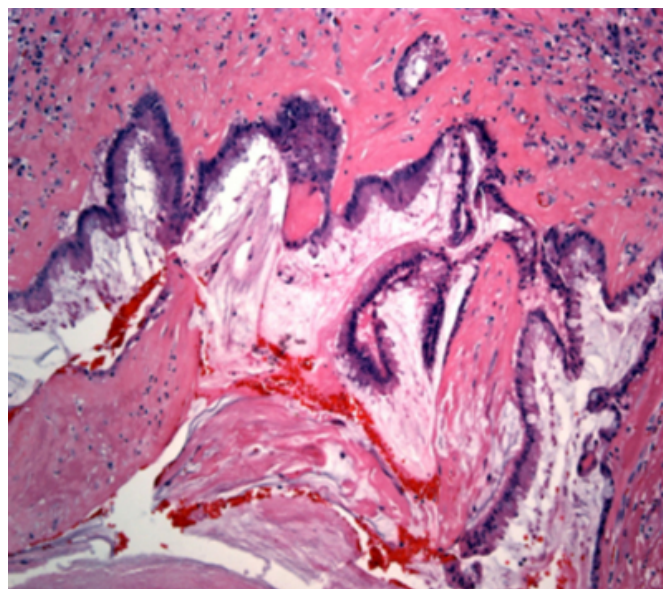


Figure 2

Figure 2: Additional sampling revealed larger cyst-like collections of mucinous material with no epithelium associated with the mucin in any of the sections examined. H&E (400X)



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DISCUSSION

Pseudomyxoma peritonei (PMP) has been a subject of confusion and varied interpretation for most oncologists and many pathologists. Three different microscopic patterns have been described in pseudomyxoma peritonei: the first and most common is disseminated peritoneal adenomucinosis (DPAM) which consists of abundant extracellular mucin, with relatively scant mucinous epithelium with limited or no cytological atypia. The second pattern designated as peritoneal mucinous carcinomatosis- (PMCA) contains extracellular mucin with abundant markedly atypical malignant mucinous epithelium. In third pattern malignant cells associated with extracellular mucin are of low to intermediate grade and are classified as PMCA-I. Recently, Bradley et al. proposed a new and simplified morphological classification of pseudomyxoma peritonei of appendiceal origin in which all of the above lesions are categorized as either low or high grade mucinous carcinoma peritonei (MCP-L and MCP-H) [1]. Most often these patients present with progressively increasing abdominal distention, acute appendicitis, or in many cases PMP is detected incidentally at laparotomy [2]. In male patients, PMP can be associated with or even present as an inguinal hernia. Esquivel et al noted as many as 25% of male patients with PMP initially presented with inguinal hernia [3]. Despite the

significant association of PMP with inguinal hernia, hernia sac, as a surgical pathology specimen, rarely harbors abnormal findings. Kassan et al evaluated the value of routine pathologic examination in hernia sacs. In their study of 1,020 specimens, only 3 cases contained unexpected pathology including Hodgkin's lymphoma, liposarcoma and atypical lipoma [4].

Since PMP is a rare clinical entity, the incidence of hernial sac mucin as a disease manifestation is unknown. However, about a dozen of case reports describe gross presence of gelatinous and mucinous material within hernia sacs, which led to further investigation and discovery of PMP, most frequently in male patients with primary appendiceal tumors [5,6,7,8,9,10,11]. The presence of gross mucin is an established alarm signal for both surgeons and pathologists. Cases in which malignant cells are present within mucin are typically associated with a known mucinous abdominal tumor and a gross finding of overt mucinous material [1]. Cases likely to be missed are those such as ours, where no gross abnormalities are noted during surgery or on pathologic gross exam such as presence of mucin or a mass within hernial tissue. As far as we are aware, this is the first grossly normal hernia sac with completely acellular mucous proven to be associated with an appendiceal mucinous neoplasm and PMP.

The presence of acellular, microscopic foci, of mucin may be dismissed or may be interpreted as benign non-neoplastic mesenchymal proliferation with myxoid stroma [5]. Along these lines, a possible differential diagnostic consideration should include "pseudosarcomatous myofibroblastic proliferation," which can be seen as an incidental surgical or pathological finding at hernia repair. However, typically such changes are grossly visible, and the myxoid areas are populated by spindle cells contrasting with the striking acellular nature of the process in the current case [5]. We microscopically identified small pools of acellular basophilic material lined by what appeared to be flattened mesothelial cells within the hernia sac wall. This prompted additional sectioning and staining with mucicarmine and PAS to confirm the epithelial origin of the acellular material. Even after careful examination of the entire hernia sac including multiple levels no epithelial tissue was identified.

CONCLUSION

Cases like this underscore the importance of the observation

that small basophilic lakes of acellular, seemingly myxoid material within hernia sacs should be taken seriously. Their presence should prompt the use of confirmatory stains for epithelial mucin and complete submission and careful examination of the entire hernia specimen, including multiple levels. Even in the absence of neoplastic epithelium, a verbal communication with the surgeon and recommendation for further imaging or laparoscopic examination in search of abdominal mucinous tumor may be justified and necessary.

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