Case Report

High-Grade Appendicular Mucinous Neoplasia (jelly belly): A Rare Case Report

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Introduction: Primary neoplasms of the appendix are present in less than 2% of surgical appendectomy specimens. The major categories of primary neoplasms include epithelial tumors, mesenchymal tumors and lymphomas. Mucinous neoplasms of the appendix are a complex, diverse group of epithelial neoplasms often causing cystic dilation of the appendix due to accumulation of gelatinous material, morphologically referred to as mucoceles. A case of High grade Mucinous Neoplasia of appendix with Pseudomyxoma peritonei is presented with a discussion of the histologic and radiologic features as well as the surgical management.

Material and Methods: This is a case report of Patient with Appendicular mucinous neoplasia with pseudomyxoma peritonei diagnosed in an elderly with it's surgical management.

Discussion: Appendiceal mucinous neoplasms are a heterogeneous group of neoplasms ranging from simple mucoceles to complex pseudomyxoma peritonei. Mucoceles are subset that consists of gradual cystic dilation of the vermiform appendix due to slow accumulation of mucoid contents. It is uncommonly seen, occurring in between 0.2% and 0.4% of appendectomies and 0.3 %and 0.7% of all appendiceal pathology. The classification of mucinous neoplasms of the appendix is controversial and different terminologies have been used to describe these lesions. Recent efforts to build a consensus naming system have led to the development of a classification system that includes LAMN, HAMN, and mucinous adenocarcinomas.

Conclusion: In conclusion, Classification and taxonomy of mucinous neoplasms is complex and controversial. Distribution of mucin and the presence of nodal and visceral metastases beyond the peritoneum help to differentiate low-grade and high-grade PMPs. PMP with simultaneous appendix and ovarian neoplasm should be treated as a primary appendiceal tumor. Clear communication between the radiologist, pathologist and surgeon is important for optimal patient manageme.

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Introduction

Less than 2% of surgical appendectomy specimens had primary appendix neoplasms. Mesenchymal tumors, lymphomas, and epithelial tumors are the three main types of primary neoplasms. The appendix's mucoceles, or buildup of gelatinous material, are the morphological term for a complex and diverse collection of epithelial neoplasms known as mucinous neoplasms. These tumors frequently cause cystic dilatation of the appendix. Mucoceles were first identified by Rokitansky in 1842. They are frequently incidentally found in people who are asymptomatic, both clinically and radiologically. Nonetheless, it's critical to understand that the mucocele may have a variety of underlying causes, from benign retention cysts to malignant adenocarcinomas, and that rupture of the mucocele may give rise to the terrifying pseudomyxoma peritonei (PMP) consequence.

A case of High-grade Mucinous Neoplasia of an appendix with Pseudomyxoma peritonei is presented with a discussion of the histologic and radiologic features as well as the surgical management.

Material And Methods

This is a case report of patient with Appendicular mucinous neoplasia with pseudomyxoma peritonei diagnosed in the elderly with its surgical management.

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Case Details

Our Patient 62-year-old Male, came to the emergency surgery department with complaints of pain in the abdomen for 3 days, sudden onset, progressive in nature and non-radiating, associated with fever, 1 episode 1 day back. The patient also complained of loss of mission in right eye since 3 days. There was no h/o vomiting, diarrhea, constipation. The patient is a k/c/o Hypertension since 7 years on medication. H/o CVA 7 years back associated with left hemiplegia. On Examination. GC of the patient was Average, thin built and patient was vitally stable. On Per Abdomen examination, the abdomen was soft, with diffuse generalized tenderness present no local signs of inflammation present. Per rectal Examination was not significant. On USG, there was e/o aperistaltic, blind-ended, non-compressible cystic lesion seen in RIF region measuring 4.7x3.5cm which seems to be communicating with thick Organised septate collection measuring 17.2x7cm extending uphill left lumbar region. Overall s/o Mucocele of appendix with Organised collection. CECT whole abdomen was done, which suggested cystic lesion measuring 3.2x3.1x6.1cm in RIF extending from the base of the caecum and communicating with a loculated collection of size 13.1x7.3x19.3cm in the left lower peritoneal cavity extending from left lumbar to left pelvic region s/o mucocele of an appendix (another name- jelly belly). The patient was planned for a Diagnostic laparoscopy. Intraoperatively fragile appendicular mass of size 10x 8 cm appreciated with diffuse thick collection in the peritoneal cavity. The decision was made to convert to Exploratory laparotomy. Intraoperatively 10x8 cm whitish appendicular mass localized at IC junction along with 300ml of mucinous fluid was drained. Postoperatively, the patient is vitally stable, accepting oral feeds and has a healthy midline wound. The Patient was Discharged to be followed up in Cancer Hospital for further chemotherapy.

Histopathology Report

High-grade Appendiceal Mucinous Neoplasia. Mucinous material is seen dissecting in the wall of the appendix.

Discussion

The group of neoplasms known as appendiceal mucinous neoplasms is diverse and can include both simple mucoceles and sophisticated pseudomyxoma peritonei. Mucoceles are a subgroup of vermiform appendix that gradually dilate cystically as a result of a steady buildup of mucoid contents. It occurs in 0.2 to 0.4% of appendectomies and 0.3 to 0.7% of all appendiceal pathologies, making it an unusual occurrence. A variety of kinds of diseases can cause an appendiceal mucinous neoplasm. These include neoplasms, hyperplastic mucoceles (caused by appendix and caecal mucosa hyperplasia), and simple mucoceles (obstruction caused by degenerating epithelium). Up to 25% of cases have simple or hyperplastic mucoceles; the remaining cases have malignant aetiologies.

Both CT and ultrasound are useful imaging modalities that are frequently used in emergencies to detect AMN. A mass with tiny echo spots and/or concentric, echogenic layers—also referred to as "onion skin"—are typical ultrasonography results. Characteristic findings on the CT scan are a luminal diameter of more than 1.3 cm, mural calcification, and low attenuation cystic dilatation of the appendix. There is disagreement over how to classify mucinous neoplasms of the appendix, and various terms have been used to refer to these lesions. LAMN, HAMN, and mucinous adenocarcinomas are included in the classification scheme that was developed as a result of recent attempts to create a consensus nomenclature system.

Whereas LAMN and HAMN relate to non-invasive lesions with variable degrees of cytologic atypia, the term "mucinous adenocarcinoma" is often reserved for mucinous tumors with characteristics of infiltration into the appendiceal wall. The word "HAMN" was coined in 2015 as a result of an international agreement by the Peritoneal Surface Oncology Group. Before, this condition was known as "cystadenocarcinoma" or "non-invasive mucinous adenocarcinoma," which was considered imprecise and incongruous with other terms for appendiceal neoplasms. Under a microscope, this lesion exhibits pushing-border invasion into the wall without any infiltration, like abnormalities in the appendiceal wall seen in LAMN. On the other hand, cytologic atypia in the epithelium is of a higher grade than in LAMN. Mutational studies and morphologic evidence suggest that the histogenesis of LAMN and HAMN is similar and that HAMN most likely develops from LAMN. Both entities exhibit elevated frequencies of KRAS and GNAS mutations, while HAMNs frequently carry extra ATM and TP53 mutations. To make sure that there is no centre of invasive cancer or HAMN, it is crucial to submit the complete appendix material for microscopic inspection in cases of LAMN. Given that mucinous neoplasms are known to be the most common cause of PMP, a potentially

fatal illness characterized by the deposition of pools of mucin in the peritoneum, including HAMN in the differential diagnosis for abdominal cystic masses, is crucial. Patients with PMP may get surgical hyperthermic intraperitoneal chemotherapy in addition to significant debulking. The patient is more vulnerable to PMP if a mucinous tumor ruptures during surgery and spreads into the peritoneum. To assess for lymph node metastasis, extensive mesoappendiceal excision is advised for non-invasive appendiceal mucinous neoplasms. Furthermore, a right hemicolectomy ought to be taken into account in cases of positive margins, tumors of 2 cm or more, or tumors with high-grade histology.

If there is lymph node-positive, substantial resection may also be necessary for invasive mucinous adenocarcinoma. Additionally, new studies have shown that folinic acid, fluorouracil, and oxaliplatin combination therapy (FOLFOX) neoadjuvant chemotherapy may be useful in treating metastatic disease or acute myeloid rupture when combined with serial debulking and hyperthermic intraperitoneal chemotherapy therapy.

Conclusion

In summary, mucinous neoplasm taxonomy and classification are difficult and contentious. Adenomas are neoplasms that are limited to the appendix's mucosa; LAMNs or adenocarcinomas are neoplasms that expand outside of the appendix. Findings from imaging, particularly MDCT, that are concerning include the existence of a soft tissue mass, uneven thickening of the wall, and consequences including rupture. It is critical to identify extra-appendiceal mucin in order to properly stage and prognosticate. If the diameter of an appendiceal mucocele is more than 2 cm, surgery should be advised. The ability to distinguish between low-grade and high-grade PMPs is aided by the distribution of mucin and the existence of nodal and visceral metastases outside of the peritoneum. PMP should be treated as a primary appendiceal tumor when it coexists with an ovarian malignancy and an appendix. For the best possible patient care, the radiologist, pathologist, and surgeon must communicate clearly.

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