Case Report: Intussusping mucocele appendix [version 1; peer review: awaiting peer review]

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Abstract
Background: A case study of a presentation of a mucocele appendix, a rare condition accounting for 0.2% of appendicectomies. The case and operative management are discussed along with the possible progression to pseudomyxoma peritoneii and its differing management.
Case: A 15-year-old girl had two presentations with atypical Right Iliac Fossa pain over 2 months. This was investigated with ultrasound and CT which revealed a calcified, intussuspecting mucocele of the appendix. This was surgically resected with partial Right Hemicolectomy. The patient was discharged on day 3 with no complications.
Discussion: The presentation, malignant potential, investigation and management of the mucocele appendix are discussed. The rare presentation of a mucocele appendix necessitates care to eliminate the risk of pseudomyxoma peritoneii. The operative management should minimise disturbance of the peritoneum in this presentation. In this case, due to an intersussepting nature a limited Right Hemicolectomy had to be performed. This is compared to the literature.

Keywords
Mucocele, Appendix, Pseudomyxoma peritoneii

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**Introduction**

Mucinous or mucocele appendix is a rare but clinically significant presentation, presenting as 0.2–0.3% of appendicectomies. While uncommon, it has potential to cause great morbidity if not recognised. It may be detected incidentally or in the setting of appendicitis. The landmark study by Higa et al. advocated describing mucocele appendix as a spectrum encompassing both benign and malignant cystadenoma, as both have potential to progress to pseudomyxoma peritoneii. In 25–50% of presentations the condition is not suspected prior to operation. There may however be prior suspicion if the nature of the presenting complaint is chronic from intussception. Classically 50% of cases will also present with a palpable mass in the right iliac fossa (RIF). In the case presented below, there was suspicion raised due to the chronicity of the pain, and negative markers on investigation of infection or inflammation. A ‘volcano’ sign may be seen at the appendiceal orifice at colonoscopy.

The condition may also present late with development of pseudomyxoma peritoneii. This condition, also known as ‘jelly belly’ has a poor prognosis, with a 5-year survival of 25%. It typically presents with nonspecific abdominal pain and painless increasing abdominal girth. On CT, intraperitoneal mucus is seen encasing small bowel and having the same density as ascites. There will be a calcified cyst and frequently a perforated appendix. The suspicion or operative findings of pseudomyxoma can drastically change the management and prognosis of the patient. Clinical and intraoperative suspicion allows for this potent pathology to be managed safely.

**Case**

A timeline of the case is shown in Table 1.

The patient, a 15-year-old-girl, had been experiencing 2 months of intermittent right iliac fossa pain. She presented to Toowoomba Hospital, a public facility in regional SE Queensland. It was an atypical presentation, with normal inflammatory markers, βHCG and white cell count. Initial ultrasonography eliminated gynaecological pathology but failed to visualise the appendix. The patient was initially observed overnight and discharged before presenting the next day with ongoing pain. A repeat ultrasound showed an ‘aparistatic’ segment of distal ileum (Figure 1). This was followed up with CT that revealed a calcified mucocele appendix which was intussuspected into the caecum (Figure 2).

The patient was taken to theatre for resection (Figure 3 and Figure 4). The plan was for laparoscopic resection, with low threshold for open resection due to the risk of pseudomyxoma peritoneii. The caecum was able to be safely mobilised during the case, however due to the intussuspected mucocele in situ it was not felt safe to do a limited ileocolic resection with a linear stapler without risk of spillage. The decision was made to proceed to a lap-assisted limited ileocolic resection. The specimen was safely removed and sent for histopathology (Figure 5).

**Post-operative care and follow-up**

The patient recovered quickly from the surgery, being discharged on day three post op. She was not given post-operative antibiotics, only prophylactic 1g cefazolin intraoperatively. She required only paracetamol and ibuprofen as required for pain relief. She was seen in the outpatient clinic at 2 weeks for

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**Table 1. Timeline of the case presented in this case report.**

<table>
<thead>
<tr>
<th>Day/Time</th>
<th>Event</th>
<th>Investigations/Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Initial presentation</td>
<td>Presented with RLQ pain and nausea</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Normal FBC and e/LFT</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Observed overnight</td>
</tr>
<tr>
<td>1</td>
<td>Discharged home</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Represented with ongoing pain</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>USS – non-peristaltic mass RLQ</td>
</tr>
<tr>
<td></td>
<td></td>
<td>CT – Mucocele appendix</td>
</tr>
<tr>
<td>2</td>
<td>Taken to theatre</td>
<td>Laparoscopic ileocolic resection</td>
</tr>
<tr>
<td>3–4</td>
<td>Post-operative recovery</td>
<td>Diet upgraded to full, mobilised, discharged day 3 post-operative</td>
</tr>
<tr>
<td>16</td>
<td>Chart review</td>
<td>Phone review, histology checked</td>
</tr>
</tbody>
</table>

**Figure 1. Ultrasound showing an ‘aparistatic’ segment of distal ileum.**

**Figure 2. CT scan showing mucocele intusscepting into caecum.**
operative procedure. This avoided putting the patient at risk of rupture of the mucocele itself and inadvertent iatrogenic seeding of the peritoneum. Classically it has been advocated that a mucocele appendix be removed via an open approach to facilitate dissection of the caecum and minimise risk of rupture. In this case, it was possible to mobilise the caecum to facilitate a limited Right Hemicolectomy.

The presentation of this case allowed for the early detection and operative planning of the mucocele resection. In the event of finding an unexpected mucocele appendix, there is controversy as to whether an initial right hemicolectomy should be completed. A study by Fernado et al. recommend a tylectomy if there is a base larger than 2 centimetres. If the base was less than this an appendicectomy with mesoappendiceal resection could be undertaken. Frozen section, if available, could allow the elimination and confirm whether a further colectomy would be required on table for oncological resection. If this were not available they recommend returning for further right hemicolectomy if necessary. In this case however, due to an intersuscepting mucocele, it was not safe or possible to perform a typhlectomy with a linear stapler without risking spillage of mucus. The caecum was safely mobilised laparoscopically, and the specimen was safely delivered intact.

In the event of finding a ruptured mucocele with free mucus the question arises whether to perform an immediate Right Hemicolectomy or proceed to a more limited appendicectomy, with the option for more major surgery later. A study by Gonzalez-Mareno and Sugarbaker found that there was increased risk of seeding of the retroperitoneum with an initial Right Hemicolectomy. In their prospective study of 501 patients treated with proven peritoneal seeding requiring peritoneal cytoreductive surgery and intraperitoneal chemotherapy, it was found that the initial surgery itself was more predictive of mortality and morbidity than nodal status.

The rare nature of this presentation makes the clinical suspicion of this condition prior to operation difficult. The morbidity and mortality of pseudomyxoma peritoneii, risk of progression either in benign or malignant mucocles and the young presenting age of this patient, emphasise a careful approach, avoiding handling of the mucocele itself and minimal disruption to the peritoneum. In this case the chronic nature of the pain on the history, lack of inflammatory signs on the bloodwork and detection on imaging facilitated this approach.

Consent
Informed written consent from the patient’s guardian was obtained for publication of this case.

Data availability
All data underlying the results are available as part of the article and no additional source data are required.

Grant information
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References

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