

**URACHAL MALIGNANCY PRESENTING AS ACUTE GENERALIZED PERITONITIS:
A CASE REPORT AND REVIEW OF LITERATURE****Dr. Supriya¹, Dr. Vijay Verma^{*2} and Dr. Ravi Verma³**¹Department of General Surgery, Dr. RPGMC Tanda, Kangra, H.P, India.²Department of General Surgery, IGM, Shimla, H.P, India.³Department of Paediatrics, IGM, Shimla, H.P, India.***Corresponding Author: Dr. Vijay Verma**

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ABSTRACT

Acute generalised peritonitis (AGP) is a potentially life-threatening intra-abdominal pathology worldwide which partly manifests as an acute abdomen. AGP is the most frequent cause of acute abdomen. It has several causes, the most common being hollow viscus perforation. Another causes could be perforation of any tumour, post trauma, post surgery, diseases like tuberculosis and pyometra etc. Here we present a case of an elderly chronic smoker patient who presented to us with features of acute generalized peritonitis and was later found to be a case of tumour perforation arising from a very rare vestigial organ, called urachus.

KEYWORDS: AGP, urachus, vestigial organ, acute abdomen, perforation.**INTRODUCTION**

Acute generalised peritonitis (AGP) is a potentially life-threatening intra-abdominal pathology worldwide which partly manifests as an acute abdomen.^[1,2] AGP is the most frequent cause of acute abdomen^[3,4] requiring broad-spectrum antibiotics and source control with laparotomy or laparoscopic surgery for definitive cure,^[5,6] except in case of spontaneous peritonitis which is a medical emergency treated with antibiotics only.^[7] Furthermore, AGP is a major contributor to non-trauma deaths in all emergency settings, and the second leading cause of sepsis in critically ill patients.^[8,9] Its pathophysiology involves an inflammatory process of the peritoneum caused by an infectious or chemical agent which irritates the peritoneal cavity by either hematogenous spread of infection from another body part to the peritoneal cavity, or by perforation of an intra-abdominal hollow viscus with resultant spillage of bacteria into the peritoneal cavity.^[3,4,7,10] If not treated in time, it spreads to the systemic circulation leading to bacteraemia, septicemia, septic shock, multiorgan dysfunction and ultimately death.^[5]

Different from benign peritonitis, nodular peritoneal thickening is the hallmark of peritonitis induced by GI tract perforation due to primary or metastatic neoplastic bowel involvement with peritoneal spread. In fact, peritoneal carcinomatosis may be due to a primary abdominal tumour, such as gastric or colorectal or pancreatic or ovarian cancer^[11] or an extra abdominal tumour such as breast carcinoma, melanoma or lung cancer. Usually diffuse peritoneal involvement is

characterized by parietal and visceral nodular implants which cover and encase the small bowel loops, leading to obstruction, and occasionally to perforation.^[12]

Bladder carcinomas are usually diagnosed at an early stage as they most frequently present with hematuria. Other symptoms include frequent and painful urination and back pain.

The urachus is a vestigial structure that connects the bladder to the allantois during early embryonic development. After birth it becomes a fibrous cord known as the median umbilical ligament. If remnants of the allantois remain within the ligament, they may develop into cysts and epithelial neoplasms.^[13,14] Urachal carcinomas occur mostly in the bladder dome, comprising 22-35% of vesical adenocarcinomas^[15], and are generally treated by partial cystectomy with en bloc resection of the median umbilical ligament and umbilicus.^[16] These have a diffuse positivity for immunostains like 34BE12.^[17] Urachal carcinomas are rare and comprise of 0.35-0.7% of all bladder cancers.^[15] Perforation in these tumour masses may lead to peritonitis.

Here we present one such case of acute generalized peritonitis arising from urachus.

CASE REPORT

A 76 year old male patient presented to us with complaints of pain in whole of the abdomen for 3 days. Pain was severe in intensity, generalized, non radiating,

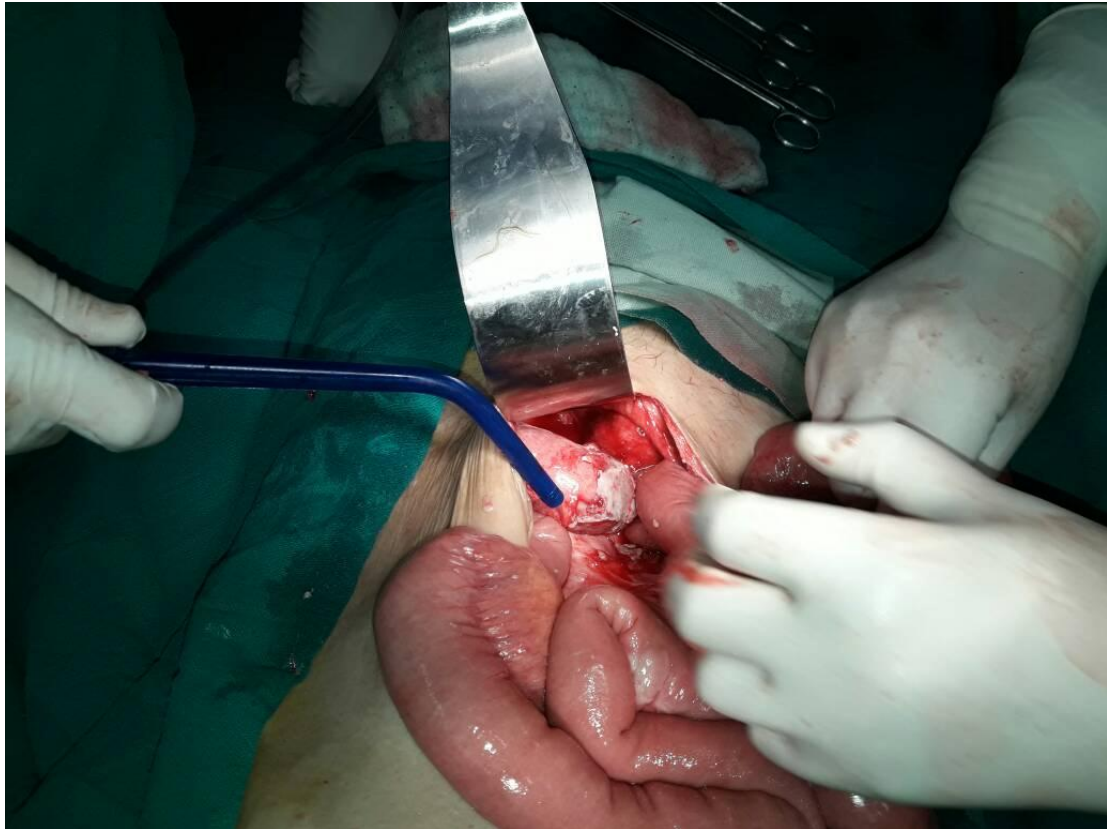
continuous and persisted throughout day. He also had complaints of vomiting which was single episode, non bilious, foul smelling and contained food particles. There were no associated aggravating or relieving factors. Patient was a chronic smoker with a smoking index of 600. He had no associated comorbidity. On per abdominal examination, patient was found to be having generalized tenderness all over the abdomen along with guarding, rigidity and rebound tenderness. Blood investigations were as follows: (Hb -6.5g/dl, TLC - 10800mcg/L, Urea-37mg/dl, Creat-1.8mg/dl, Na - 134mmol/L, K-5.02 mmol/L) His chest X ray revealed air under diaphragm. A diagnosis of acute generalized peritonitis was made and the patient was taken to operation theatre for peritoneal toiletting and further management.

Intraoperative findings

Midline laparotomy incision was given. About 500 ml of free fluid, mixed with pus flakes was present which was suctioned out carefully. Stomach was found to be distended and normal. There was no evidence of any perforation or growth in stomach. On gross examination

of duodenum, there was minimal scarring at D1. No perforation was found. Rest of duodenum was found to be normal. All solid organs were found to be grossly normal. Small gut loops were found to be jumbled up. On further proceed, we found a soft, tumour mass in pelvis, 4X4 cm in size, in superior aspect of the dome of urinary bladder. Jumbled small gut loops were attached to that mass. Careful dissection was done. Interloop pus flakes were present. The mass had necrotic central part with centrally placed opening from which a serous fluid was found to be coming out in drops. A small serosal tear in ileum was seen, which was sutured with absorbable sutures. Rest of the gut loops were normal including appendix. Multiple mesenteric lymph nodes were found to be enlarged. Biopsy of the lymph nodes was taken. Biopsy from the growth was also taken. Thorough peritoneal toiletting along with peritoneal drainage was done. A probable diagnosis of malignancy originating either from the urinary bladder or from urachus and its perforation was suspected and biopsy was awaited. The fluid coming out of the tumour mass was supposed to be urine leaking from the bladder.



**DISCUSSION**

Peritonitis is the acute or chronic peritoneal inflammation with characteristic local and general changes and severe dysfunction of vital organs. It

complicates approximately 0.8-2% of all clean surgeries and comprises of 20% of all inflammatory pathology of the abdominal cavity. Mortality rate of peritonitis rises to 70-80%. It is classified as follows:

Primary peritonitis	Spontaneous peritonitis of child Spontaneous peritonitis of adults Peritonitis in patients with CAPD (continuous ambulatory peritoneal dialysis) Tuberculous peritonitis
Secondary peritonitis	Non iatrogenic gastrointestinal perforation, postop peritonitis Post traumatic peritonitis(after blunt trauma abdomen, penetrating trauma)
Tertiary peritonitis	Peritonitis without pathogen Peritonitis with fungi Peritonitis with low grade pathogenic bacteria
Intra abdominal abscess	Intra abdominal abscess after primary, secondary or tertiary peritonitis

Acute generalized peritonitis has multiple causes. As the complication of surgical pathology, 50% of appendicitis, 16% of cholecystitis, 7% of gastric ulcer and cancer perforation, 6% of pancreatitis, 6% of mesenteric thrombosis and 2% of colon cancer progress to acute generalized peritonitis. 13% cases belong to postoperative peritonitis.

Signs and symptoms include painful abdomen, abdominal distension, fever and chills, passing few or no stools or gases, excessive fatigue, passage of less urine, nausea and vomiting, absent peristaltic sounds, thread like pulse, positive peritoneal signs(Blumberg's sign), rigidity, guarding, generalized tenderness and rebound tenderness.

The urachus is a vestigial structure that connects the bladder to the allantois during early embryonic development. After birth it becomes a fibrous cord known as the median umbilical ligament. If remnants of the allantois remain within the ligament, they may develop into cysts and epithelial neoplasms.

The urachus has intramucosal, intramuscular and supravescicular segments. It contains three distinct tissue layers: an epithelial canal lined by urothelium, submucosal connective tissues and an outer layer of smooth muscle. Urachal neoplasms can arise in any of these layers, and can be epithelial or mesenchymal. Similar to urothelium at other sites, the epithelium often demonstrates focal glandular metaplasia, and this provides a morphologic basis for the development of intestinal-type tumors.^[13,14] Urachal carcinomas occur mostly in the bladder dome, comprising 22-35% of vesical adenocarcinomas^[15], and are generally treated by partial cystectomy with en bloc resection of the median umbilical ligament and umbilicus.^[16] These have a diffuse positivity for immunostains like 34BE12.^[17] Urachal carcinomas are rare and comprise of 0.35-0.7% of all bladder cancers.^[15]

The criteria for a diagnosis of urachal carcinoma are somewhat controversial but most investigators agree with those set forth by Sheldon et al^[15] and Mostofi et al.^[18] These include (a) tumor in the dome of the bladder;

(b) absence of cystitis cystica and cystitis glandularis; (c) predominant invasion of the muscularis or deeper tissues with a sharp demarcation between the tumor and surface bladder urothelium which is free of glandular or polypoid proliferation; (d) presence of urachal remnants within the tumor; (e) extension of tumor into the bladder wall with involvement of the space of Retzius, anterior abdominal wall or umbilicus, and (f) no evidence of a primary neoplasm elsewhere. A few staging systems have recently been proposed^[19,20], but commonly followed is that proposed by Sheldon et al^[15]: pT1 – no invasion beyond the urachal mucosa; pT2 – invasion confined to the urachus; pT3 – local extension to the (a) bladder, (b) abdominal wall, (c) viscera other than the bladder, and pT4 – metastasis to (a) regional lymph nodes, (b) distant sites.

Points in support of our diagnosis is that the tumour was located in pelvic region, on the dome of urinary bladder with leak of fluid possibly urine from it. Also the patient is a chronic smoker and is a farmer by occupation who could be exposed to several chemicals, thus supporting our another probable diagnosis of bladder carcinoma. However, the biopsy is awaited.

CONCLUSION

It is important to inspect whole of the gut loops from stomach to entire colon including appendix while looking for any perforation to find out the cause of acute generalized peritonitis. It is equally important to inspect pelvic organs before making a diagnosis so that the exact diagnosis can be made and further management can be done accordingly.

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