

Retroperitoneal mass a rare manifestation of mucinous adenocarcinoma of appendix; a case report

Mehdi Salehipour¹, Abdolreza Haghpanah¹, Jamshid Roozbeh¹, Anahita Dehghani¹, Armin Amirian¹, Nader Moin vaziri¹, and Sajad Kiani¹

¹Shiraz University of Medical Sciences

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Abstract

The retroperitoneal may be affected by variety of infectious, inflammations and tumors including benign or malignant ones. Although primary malignant tumors are the most prevalent tumors in this anatomic area, metastatic and invasive tumors rarely involve retroperitoneum. Gastrointestinal stromal tumors (GISTs) counted the most common tumors with invasion the retroperitoneum

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Running title: Appendiceal tumor leading to retroperitoneal mass

Mehdi Salehipour^{1,2}, Abdolreza Haghpanah^{1, 2}, Jamshid Roozbeh¹, Anahita Dehghani^{1*}, Armin Amirian³, Nader Moinvaziri⁴, Sajad Kiani²

1. Shiraz Nephro-Urology Research Center, Shiraz University of Medical Sciences, Shiraz, Iran
2. Endourology Ward, Urology Department, Shiraz University of Medical Sciences, Shiraz, Iran
3. Thoracic and vascular surgery Research center, Shiraz University of Medical Sciences, Shiraz, Iran
4. Laparoscopy Research center, Surgery Department, Shiraz University of Medical Sciences, Shiraz, Iran

*Corresponding author:

Anahita Dehghani, MD, Research fellow, Shiraz Nephro-Urology Research Center, Shiraz University of Medical Sciences, Shiraz, Iran

Postal address: 7193635899- Khalili Street, Mollasdra Avenue, Shiraz, Iran

E-mail address: Anahita.dehghani2997@gmail.com

Tel: +989177902507

Abstract

Background: The retroperitoneum (RP) may be affected by variety of infectious, inflammations and tumors including benign and malignant ones. Although primary malignant tumors are the most prevalent tumors in this anatomic area, metastatic and invasive tumors rarely involve retroperitoneum. Gastrointestinal stromal tumors (GISTs) counted the most common tumors with invasion the retroperitoneum, but to our knowledge it is time that a surgery team encountered the appendiceal tumor as a huge retroperitoneal mass.

Case presentation: A 68 years old man referred to emergency department with abdominal distension and weight loss. In his course of hospitalization, a huge right retroperitoneal mass was detected by CT scan and thereafter the patient underwent laparotomy, evacuation of massive mucinous tissue located in right retroperitoneum and right hemicolectomy due to appendiceal tumor. Histopathological exam showed 'mucinous appendiceal neoplasm'.

Conclusions: this is the first case study that appendiceal tumor had invaded visceral peritoneum into the retroperitoneum, so, an invasion of peritoneal tumor to retroperitoneum should be considered when an urologist approaches to retroperitoneal mass.

Keywords: Appendiceal tumor, Multiloculated mucinous collection, Retroperitoneal mass

Background

The retroperitoneum (RP) may be affected by variety of infectious, inflammations, rare benign tumors or malignant neoplasms that can be either primary or metastatic. Malignant tumors of the retroperitoneum happen four times more often than benign lesions.(1, 2)

Benign retroperitoneal tumors (RPT) are usually detected clinically and the most common one is neurogenic tumors (30%), followed by teratomas (15%).

Malignant RPTs are very rare tumor, count 0.1-0.2 % of all malignant tumors. Most of them are primary tumors, in them the most common type is retroperitoneal but metastatic tumors may involve retroperitoneum. (3, 4). Metastatic RPTs may originate from kidney , bladder or genitalia.(5)

Invasion of malignant tumors to the retroperitoneal is very rare and usually has been reported as an unusual presentation of gastrointestinal stromal tumors (GISTs).(6, 7)

On the other hand, appendiceal tumors are rare and usually manifest as appendicitis. Most are harmless and can be handled with appendectomy.(8) The WHO classifies three subtypes of

mucinous appendiceal neoplasms; mucinous adenoma, low-grade appendiceal mucinous neoplasm (LAMN), and mucinous adenocarcinoma (9)

In this case report we present a novel case of an appendix tumor which manifested with retroperitoneal mass by invasion from peritoneal cavity.

Case presentation

A 68-years-old male patient without any underlined disease, was admitted to the general surgery department with abdominal distention, weight loss and abdominal pain. During physical examination, significant distention and a palpable mass in the periumbilical area above the pelvic cavity with expansion to right side were identified. The abdomen was soft and smooth, with no tenderness or guarding. In the lab data, no leukocytosis (WBC: 7300), mild anemia (HB: 12.5 mg/dl) were found. Spiral computed tomography of the abdomen and pelvic region revealed a normal right kidney displaced by a large cystic lesion in retroperitoneum on the right side, with a multiloculated cystic appearance and is infiltrating part of the right psoas muscle.it extends inferiorly to the level of the right side of the lower abdomen and pelvic cavity and also has a pressure effect over the adjacent part of the urinary bladder. (figure1)

The patient underwent exploratory laparotomy. The appendix tumor which invaded the visceral peritoneum was removed, dissected and divided from the retroperitoneum. Right hemicolectomy was done from 10 cm before the ileocecal valve up to mid transverse colon. End to end ileocolic anastomosis was done. In addition, the multiloculated mucinous collection was drained, and the cavity walls were sent to pathology. Postoperative histopathological analyses revealed that the tumor size was 4*2.5*1 cm. and tumor type was 'Mucinous appendiceal neoplasm' However, no metastasis or lymph node involvement was detected. (figure2)

The patient had an uneventful postoperative course and was discharged 10 days after surgery and we refer him to an oncologist, oncologist has prescribed 8 courses of chemotherapy (FOLFOX regimen) every 2 weeks

for prophylaxis.

Discussion and Conclusions

In this study we present a unique case of mucinous appendiceal neoplasm that invaded from intraperitoneum to retro peritoneum. To our knowledge, this is the first case study that tumor had perforated (invaded) visceral peritoneum into the retroperitoneum.

Park et al reported a 32-year-old man with extra-GIST (EGIST) who referred to the hospital with dysuria and abdominal mass. Preoperative ultrasound and MRI revealed a lobulated mixed echoic mass in the retroperitoneum with heterogeneous enhancement. Patient underwent surgical excision of tumor and a low anterior resection because of invasion to the wall of the rectum.(7) In our case also no adhesion around tissue was found but tumor extends inferiorly to the right side of the lower abdomen and pelvic cavity.

Engin *et al.* reported case of GIST with mesenteric and retroperitoneal invasion, also entered the small bowel serosa, colonic submucosa and the left ureter which presented with abdominal bulging , weight loss , and hematuria (6) . It is inconsistent with our patient, but in general, it can be concluded that penetration of tumors to visceral peritoneum can be due to the weakness of this layer.

Mucinous appendiceal neoplasms are rare, but necessary to detect with imaging due to their malignant potential. Peritoneal seeding and pseudomyxoma peritonei can occur in both malignant and low-grade appendiceal neoplasms(9).

Regardless of their clinical seriousness, mucinous neoplasms may be challenging to diagnose since the symptoms are usually mild or nonspecific. They often present with stomach pain, weight loss, nausea, fatigue, a palpable mass, and acute appendicitis. Roma *et al* . report a late stage diagnose of apendiceal mucinous adenocarcinoma with rare presentation such as scrotal pain, abscess, and inguinal drainage (10) but our patient developed with typical symptoms like abdominal pain, weight loss and palpable mass without any appendicitis alarm.

In conclusion ,due to the non-specificity of the symptoms, when patient develops with abdominal distention , pain and retroperitoneal mass , we should consider invasion of intraperitoneal tumors such as appendiceal tumors while precise surgical management of these neoplasms may avoid delay in diagnosis and/or complications of peritoneal seeding.

List of abbreviations

RP : Retroperitoneum , GISTs :Gastrointestinal stromal tumors , EGIST : Extra Gastrointestinal stromal tumors ,

Declarations

Ethics approval and consent to participate

This study was approved as a case report by ethical committee of Shiraz University of Medical Sciences.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Availability of data and materials

The material and data of the case report is the authors' own original work obtained from this case, which has not been previously published elsewhere.

Competing interests

The authors have no conflicts of interest to declare.

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Author Contributions

All authors approved the final version, and accept accountability for all aspects of this manuscript.

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Authorship

The first four authors have made substantial contributions to conception and design, acquisition of data, analysis and interpretation of data;

Also been involved in drafting the manuscript, revising it critically for important intellectual content; and given final approval of the version to be published.

All authors agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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