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Management of mucinous urachal neoplasm presenting as pseudomyxoma peritonei

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ABSTRACT

Background. Mucinous neoplasms of the urachus are rare malignancies so that the physicians' index of suspicion for a timely and accurate diagnosis is low. Also, this disease may present with a wide variety of symptoms and signs.

Methods. Two patients with pseudomyxoma peritonei as the initial presentation of urachal mucinous adenocarcinoma were treated successfully. The medical literature regarding treatment options for this manifestation of the disease was searched.

Results. Two patients with large volume of pseudomyxoma peritonei originating in a mucinous urachal neoplasm were treated with cytoreductive surgery and perioperative intraperitoneal chemotherapy. Our first patient required two reoperations to palliate the accumulation of gross mucinous ascites. She died 11 years after diagnosis with progression of mucinous adenocarcinoma resulting in starvation. The second patient had ostomy closure with second look surgery at one year after definitive treatment; four small tumor nodules were seen and easily resected. The patient is currently without evidence of disease and has a normal quality of life. Seven prior manuscripts that report a single case of pseudomyxoma peritonei were reviewed to explore the full range of treatment options and survival for this rare condition.

Conclusion. Cytoreductive surgery combined with perioperative intraperitoneal chemotherapy may be a new treatment option for mucinous urachal neoplasms presenting with pseudomyxoma peritonei. Other management strategies such as systemic chemotherapy seemed to hold little promise for this group of patients.

Key words: *mucinous adenocarcinoma, mucosuria, intraperitoneal chemotherapy, cytoreductive surgery, carcinomatosis.*

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