

# Silent Hydronephrosis: Unveiling Primary Urinary Bladder Marginal Zone Lymphoma in the Background of Follicular Cystitis

Marie Tan, BA; Wesley Perez Cervantes, BS, MS; Chioma Alaka, BS, MS; Benjamin Feng, BS; Irma Palazzo, MD

Drexel University College of Medicine, Cooper Medical School of Rowan University, Crozer Chester Medical Center

## Background

- Lymphomas of the bladder can either by **primary lymphomas** (originate from the bladder) or **secondary lymphomas** (originates outside of the bladder)
- First described in 1885 by Eve and Chaffey
- Primary lymphomas is a type of extranodal non-hodgkin's lymphoma
- **Extremely rare:** 0.2% of extranodal lymphomas are located in the bladder
- **Common symptoms:** gross hematuria, concomitant urinary tract infection, dysuria, increased urinary frequency, abdominal or back pain
- **Rare complications:** hydronephrosis, vesico-vaginal fistula, involvement of the entire bladder
- Female > Male
- Most common type of primary lymphoma: **marginal zone lymphoma of the mucosa associated lymphoid tissue**
- Followed by diffuse large B cell lymphoma – may also originate from marginal zone lymphomas
- Most commonly preceded by cystitis, or follicular cystitis
- **Positively stain for CD20, CD19, FMC7**
- **Negatively stain for CD5, CD10, CD11c**

**This is a case of marginal zone lymphoma of the bladder that is in the background of follicular hyperplasia, with the only presentation being an incidental finding of hydronephrosis**

## Case Presentation

**78 year old Spanish speaking female with history of osteoarthritis, hypertension, GERD, hyperlipidemia, type 2 diabetes, and monoclonal gammopathy with negative bone marrow biopsy 3 years prior – no history of interstitial cystitis or smoking**

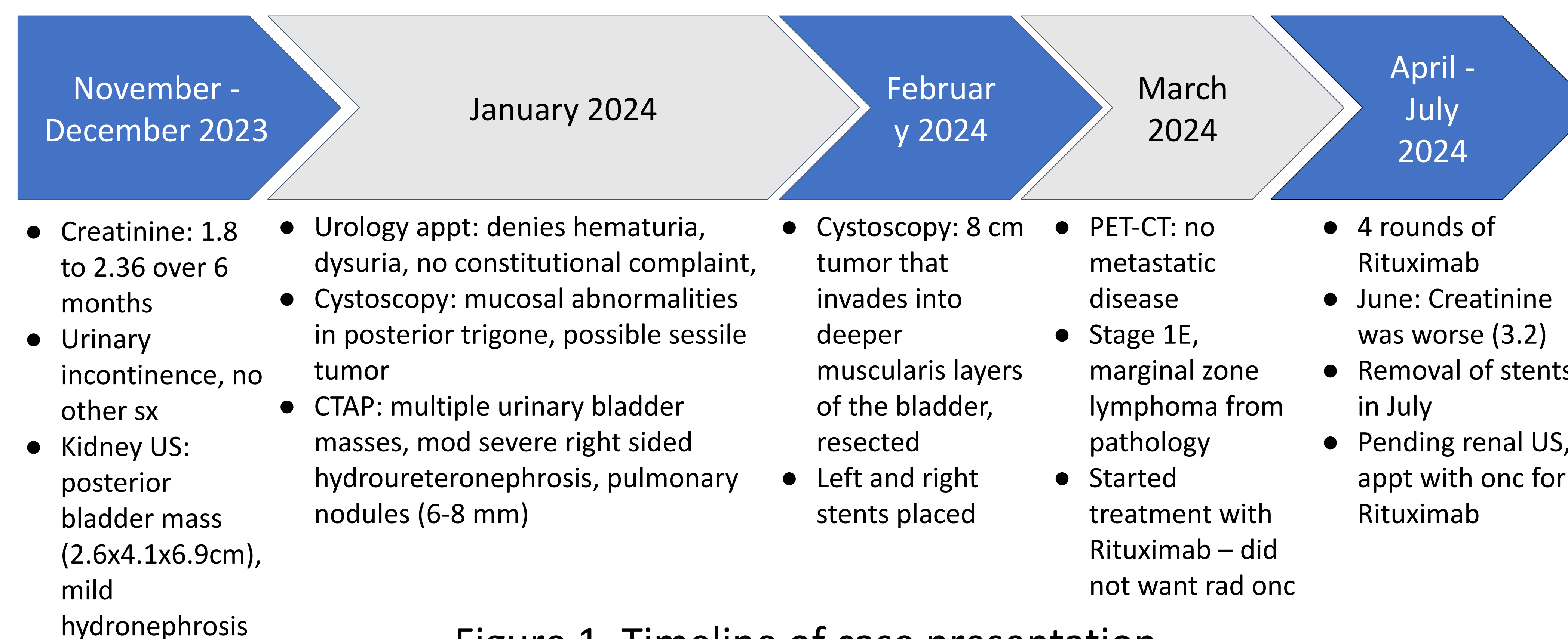


Figure 1. Timeline of case presentation

Figure 2. Renal ultrasound

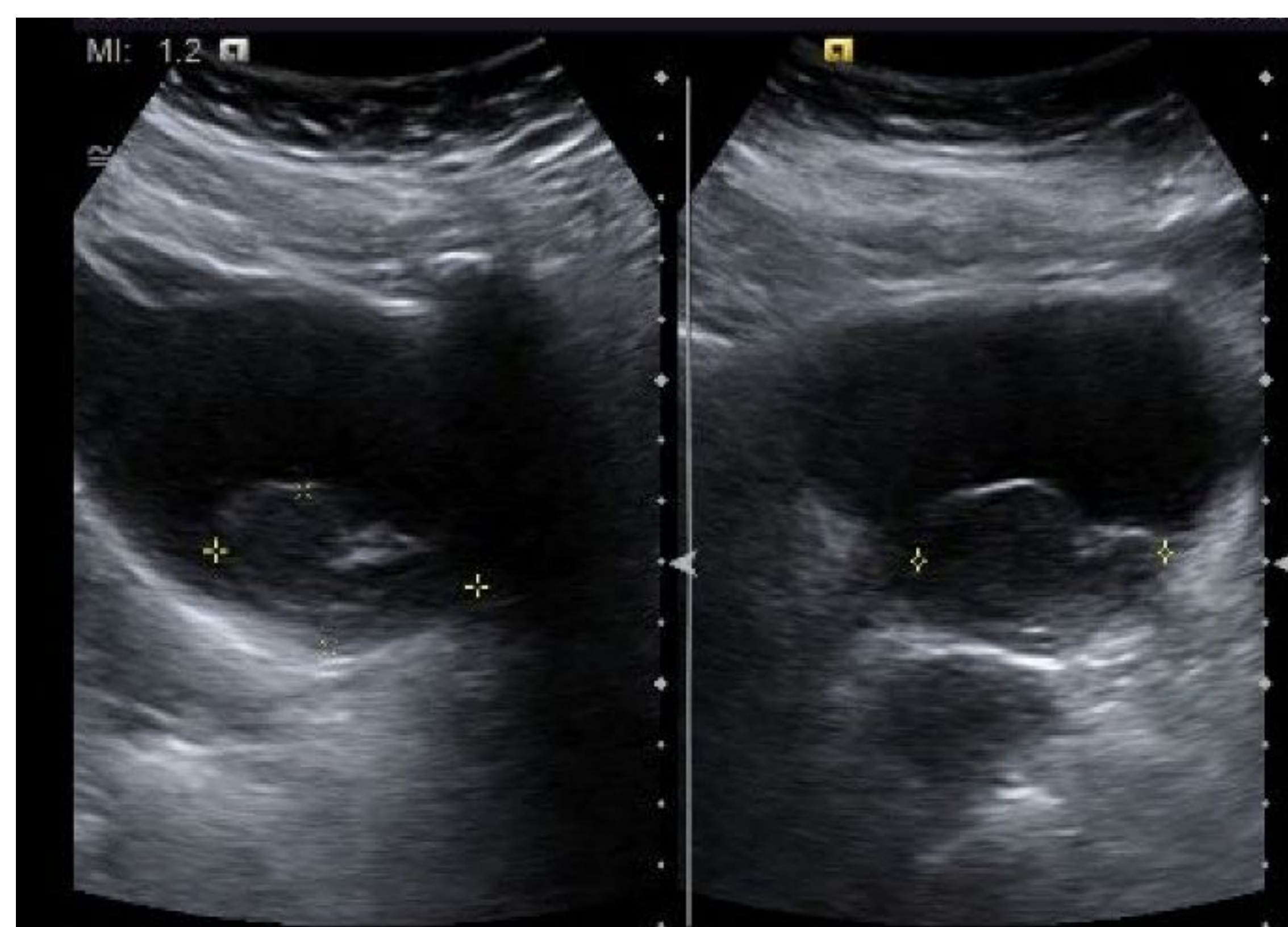


Table 1. Flow Cytometry report

CD20	Positive
PAX5	Positive
CD5	Negative
CD10	Negative
Cyclin D1	Negative
BCL6	Positive in areas that look like germinal centers
MYD66	Negative
CXCR4	Negative

## Discussion

- Hydronephrosis as the presenting symptom is extremely rare, usually a complication of long-standing, untreated bladder cancer
- In literature review: only one case - describes periureteral marginal zone lymphoma
- Patient did not have history of recurrent UTIs or interstitial cystitis
- Radiotherapy is commonly utilized in treatment – in this case, surgical resection and rituximab therapy was used
- Need to monitor patient's course

## Conclusion

This case highlights a rare complication of a rare disease: largely asymptomatic hydronephrosis in the setting of primary marginal zone lymphoma of the bladder. While radiotherapy is the preferred treatment, surgical resection and rituximab may also be used as therapy in the early stage.

## References

1. Venyo AKG. Lymphoma of the Urinary Bladder. *Adv Urol*. 2014;2014:327917. doi:10.1155/2014/327917
2. Eve FS. Two cases of sarcoma of the bladder. *Trans Pathol Soc Lond*. 1885;36:284.
3. Chaffey WC. Lymphosarcoma of the bladder. *Trans Pathol Soc Lond*. 1885;36:287.
4. Kempton CL, Kurtin PJ, Inwards DJ, Wollan P, Bostwick DG. Malignant lymphoma of the bladder: evidence from 36 cases that low-grade lymphoma of the MALT-type is the most common primary bladder lymphoma. *Am J Surg Pathol*. 1997;21(11):1324-1333. doi:10.1097/0000478-199711000-00007
5. Freeman C, Berg JW, Cutler SJ. Occurrence and prognosis of extranodal lymphomas. *Cancer*. 1972;29(1):252-260. doi:10.1002/1097-0142(197201)29:1<252::AID-CNCR2820290138>3.0.CO;2-#
6. Kawaguchi Y, Kimura S, Momozono K, Igawa T, Noguchi M. Malignant lymphoma of the bladder with bilateral hydronephrosis. *Rare Tumors*. 2019;11:203636151825165. doi:10.1177/203636151825165
7. Evans DA, Moore AT. The first case of vesico-vaginal fistula in a patient with primary lymphoma of the bladder - a case report. *J Med Case Reports*. 2007;1:105. doi:10.1186/1752-1947-1-105
8. Cohen DD, Lamarre C, Lamarre L, Fs FS. Primary low-grade B-cell lymphoma of the urinary bladder: case report and literature review. *Can J Urol*. 2002;9(6):1694-1697.
9. Sagara Ruwan Kumara MG, Thiraganama P, Sosai C, Abeygunasekera A. A case of follicular cystitis treated successfully with diethylcarbamazine. *Asian J Urol*. 2021;8(2):235-237. doi:10.1016/j.ajur.2020.03.003
10. Xu, Hewel MS; Chen, Zhonggen MS; Shen, Baixin MD; Wei, Zhongqing MD; Primary bladder mucosa-associated lymphoid tissue lymphoma: A case report and literature review. *Medicine* 99(28):e20825, July 10, 2020. | DOI: 10.1097/MD.00000000000020825
11. Jaudah Al-Maghrabi, Suzanne Kamel-Reid, Michael Jewett, Mary Gospodarowicz, Woodrow Wells, Diponkar Banerjee; Primary Low-Grade B-Cell Lymphoma of Mucosa-Associated Lymphoid Tissue Type Arising in the Urinary Bladder: Report of 4 Cases With Molecular Genetic Analysis. *Arch Pathol Lab Med* 1 March 2001; 125 (3): 332-336. doi: https://doi.org/10.5858/2001-125-0332-PLGBCL
12. Lyapichev KA, Nashkevich Y, Chernov Y, Chinenov D, Shipov E, Bessonov AA, Dabaja BS, Konoplev S. MALT Lymphoma of the Urinary Bladder Shows a Dramatic Female Predominance, Uneven Geographic Distribution, and Possible Infectious Etiology. *Res Rep Urol*. 2021 Feb 5;13:49-62. doi: 10.2147/RRU.S283366. PMID: 33575225; PMCID: PMC7873029
13. Vempati P, Knoll MA, Alqatani M, Strauchen J, Malone AK, Bakst RA. MALT Lymphoma of the Bladder: A Case Report and Review of the Literature. *Case Rep Hematol*. 2015;2015:343474. doi: 10.1155/2015/343474. Epub 2015 Aug 31. PMID: 26417464. PMCID: PMC4568333
14. Kakuta Y, Kato T, Saitoh J, Yazawa K, Hosomi M, Itoh K. [A case of primary mucosa-associated lymphoid tissue lymphoma of the bladder regressed after rituximab in combination with CHOP chemotherapy]. *Hinyokika Kyo*. 2006 Dec;52(12):951-4. Japanese. PMID: 17252980.
15. Zaharopoulos P. Cytologic manifestations of cystitis follicularis in urine specimens. *Diagn Cytopathol*. 2002 Oct;27(4):205-9. doi: 10.1002/dc.10166. PMID: 12357496.