Primary urachal leiomyosarcoma: A rare case report and literature review

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ABSTRACT

Primary urachal leiomyosarcoma is a rare entity with unclear prognosis and treatment strategy. We report a case of urachal leiomyosarcoma presenting, and treated with open urachal tumor resection and partial cystectomy. Pathological diagnosis showed low-grade leiomyosarcoma. At 53-months follow-up, no recurrence or metastasis was found. The purpose of this case is to raise awareness of the disease.

1. Introduction

Leiomyosarcoma is a malignant tumor characterized by the differentiation of smooth muscle cells and can occur in various tissues containing smooth muscle cells. However, Urachal leiomyosarcoma is extremely rare. Since 1973, only 5 cases urachal leiomyosarcoma have been reported (Table 1). We report a case of urachal leiomyosarcoma with symptoms of dysuria and frequent urination.

2. Case presentation

A 40-year-old man presented to us for an admission for dysuria, frequent urination, and urgency without a significant past medical history. Physical examination revealed no significant positive clinically signs. Urinalysis revealed microscopic hematuria. Urogenital ultrasonography examination revealed a hypoechoic mass above the bladder (Fig. 1A). Therefore, an abdomen computer tomography was administered and showed a mass above the bladder with mild to moderate inhomogeneous enhancement (Fig. 1B,C,D). Urachal carcinoma was the primary consideration. Histopathology after fine needle aspiration showed necrotic area with more neutrophil infiltration, and a few loosely arranged spindle cells around the area. Due to too little living tissue, the diagnosis could not be confirmed. Considering the tumor is larger, open surgical resection was performed. During the operation, the tumor infiltrated the bladder wall, so urachal tumor resection and partial cystectomy were performed. Postoperative histopathology showed smooth muscle tumor with negative margins, local necrosis, and mitotic figures were easily seen (Fig. 2). Immunohistochemical studies were performed revealing tumor cell immunopositivity to SMA and Desmin, B-Catenin. The cells were immunonegative of EMA, S-100, ALK, CD117, DOG-1, and Myo D1. The Ki-67 proliferation index was noted as 5%. Finally, low-grade leiomyosarcoma in urachal was diagnosed. Further adjuvant therapy was not suggested by the multidisciplinary team. At 53-months follow-up, no recurrence or metastasis was found.

3. Discussion

Urachal leiomyosarcoma is a very rare tumor. It can occur in children and adults, mostly in men. It may originate from the smooth muscle tissue of the outer layer of the completely atretic urachal. It often reaches large measurements when found, because leiomyosarcoma grows in the Retzius space.large leiomyosarcoma infiltrates and compresses the surrounding tissue, causing abdominal pain, hematuria, bladder irritation, and dysuria. In addition, Kim1 and Saied2 reported cases of ruptured urachal leiomyosarcoma with bleeding. However, its clinical features are non-specific.

Surgery is the mainstay of treatment for urachal leiomyosarcoma. Radical resection with negative margins is the recommended surgical goal. Specifically, the surgical range consist of the complete urachal, the peritoneal cuff, the bladder with 2 cm margins and the perivesical fat.3 Adjuvant chemotherapy and radiotherapy for urachal leiomyosarcoma is controversial due to rarity. Kim1 reported a 12-year-old boy with spontaneously ruptured urachal Myxoid leiomyosarcoma. He received radiation therapy and alternating adjuvant chemotherapy (Vincristine, doxorubicin and cyclophosphamide regimen, vincristine, dactinomycin and cyclophosphamide regimen). Unfortunately, no long-term follow-up data are available. Moreover, neoadjuvant chemotherapy for bladder leiomyosarcoma may provide reference for

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neoadjuvant regimens are doxorubicin, ifosfamide, cisplatin, Adriamycin, and vincristine. The prognosis of urachal leiomyosarcoma is difficult to assess due to lack of data. From the existing literature, the low-grade leiomyosarcoma is the most common type, the prognosis is good, and further adjuvant therapy was not suggested.

4. Conclusion

Urachal leiomyosarcoma is a rare tumor that is considered to be highly aggressive. As a result a radical surgical treatment is considered as necessary. Further adjuvant therapy is controversial. However, For a low-grade leiomyosarcoma, further adjuvant therapy was not suggested. Although the prognosis is good, cystoscopy and imaging should be regularly reviewed for diagnose of early recurrence and metastasis.

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| Author, Year | Sex | Age | Symptoms | Grade | Follow-up | Outcome |
|--------------|-----|-----|----------|-------|-----------|---------|
| Tara et al., 1973 | F | 54 | Dysuria, frequency | Unknown | 7 y | uneventful |
| Noyes et al., 1981 | M | 28 | Dysuria, urgency, frequency | Low LMS | 9 mo | uneventful |
| Kim et al., 2007 | M | 12 | Spontaneous rupture | Myxoid LMS | 6 mo | uneventful |
| Saied et al., 2012 | F | 21 | Traumatic rupture | Low LMS | 4 y | uneventful |
| Current report, 2022 | M | 40 | Dysuria, frequency | Low LMS | 53 mo | uneventful |

F, female; M, male; LMS, leiomyosarcoma.

Table 1

Reported cases of urachal leiomyosarcoma.

Fig. 1. Color Doppler ultrasonography showed a solid hypoechoic (4.5cmx3.7cm) above the bladder(A). Abdomen computed tomography (CT) showed an oval mass above the bladder (5.1cmx3.2cm) with mild to moderate inhomogeneous enhancement (B, C, D).

Fig. 2. H&E-stained section of the tumor. Microscopically demonstrated tumor necrosis(A). Microscopically showed irregular intersecting bundles of spindle cells, the majority of the nuclei showed atypia (B).
Declaration of competing interest

The authors declare no financial or relationships that may pose conflict of interest.

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