INTRODUCTION

Urachal carcinoma (UC) is a rare urological tumor, accounting for 0.17–0.34% of all bladder carcinomas. The incidence of bladder carcinoma is 6.7/100,000 people, whereas the incidence of all types of cancer is 237.4/100,000 people. Therefore, brain metastases from UC are extremely rare in neurosurgical practice, and only seven cases have been reported in the literature. Established risk factors include male sex, white race, cigarette smoking, and exposure to certain chemicals. The most common clinical presentation is asymptomatic hematuria. To assess for bladder cancer, patients can be evaluated by cystoscopy, renal function testing, and upper urinary tract imaging. Transurethral resection of the bladder allows for definitive diagnosis. Nonmuscle-invasive disease is treated with transurethral resection, while muscle-invasive disease is typically treated with radical cystectomy and neoadjuvant chemotherapy. Survival rates vary for different tumor stages, with the five-year survival rate ranging from 96% for carcinoma in situ to 5% for distant metastasis. UC is believed to originate from an urachal remnant that undergoes intestinal metaplasia or it might be derived from progenitor cells of the cloaca left behind during embryologic development. Histologically, the majority of UCs are adenocarcinomas arising from the bladder. Here, we present such a case of a cerebellar metastasis from UC of the bladder.
**CASE REPORT**

A 77-year-old female had been diagnosed with UC and had undergone total cystectomy 2 years prior to presentation with dizziness for which she was referred to our department. Magnetic resonance imaging revealed a 35-mm sized lesion in the left cerebellum. This lesion showed low intensity on T1-weighted images, high intensity with perifocal edema on T2-weighted images, and avid heterogeneous enhancement on T1-weighted images after gadolinium administration [Figure 1]. The differential diagnosis included metastasis, glioma, and abscess. Surgery was performed using a standard midline suboccipital approach with the patient in the prone position under general anesthesia. The tumor was totally removed and postoperative course was uneventful. The excised specimen was pathologically diagnosed as mucinous adenocarcinoma consisting mainly of proliferating signet ring cells, which was identical to the features seen in the primary urachal adenocarcinoma comprised a mixture of signet ring cells, mucus-producing cells, and duct-forming cells [Figure 2]. This tumor was pathologically, therefore, diagnosed as cerebellar metastasis of the UC. Recurrence was observed at the site of tumor removal 6 months after surgery, and subsequently, gamma knife radiotherapy was performed to remove the lesion.

**DISCUSSION**

UC is a rare variant of bladder carcinoma which differs from normal bladder carcinoma in several ways: Instead of arising from the urothelium, it is believed to originate from an urachal remnant that underwent intestinal metaplasia or from progenitor cells of the cloaca left behind during embryologic development. This theory is reflected in the pathohistological differences observed between these two cancer types. UC normally exhibits features of adenocarcinoma and often contains ductal cells, mucus-producing cells, and signet ring cells, mimicking features of gastrointestinal cancers. In this case, pathology was typical of UC developing a metastasis to the brain.

Generally speaking, UC is treated in a way comparable to normal bladder carcinoma. Surgery is the treatment of choice, whereas chemotherapy and radiotherapy can be chosen for advanced cases. In cases with infiltration of adjacent organs or with distant or lymph node metastasis, poor prognosis is expected; the 5-year survival rate is believed to be 20% or less. The treatment modalities include surgery, chemotherapy, radiotherapy, and a combination of these.

As of now, seven cases of brain metastasis from UC have been reported. Five of these are published in English or Japanese and include detailed discussions regarding brain metastasis [Table 1]. Except in one reported patient, tumor removal was performed in all cases. Adjuvant radiotherapy was provided in four of the cases, one of which employed gamma knife treatment. Chemotherapy for brain lesions was not performed for any of these cases because it was not thought to cure brain metastasis. Prognosis was poor, with the survival time ranging from 1 to 18 months. Hirshman et al. reported that the cumulative volume of metastasis to the CNS is considered a predictive factor associated with treatment response and survival rate in patients with CNS metastasis. The cases in the current report tended to follow this theory. Among them, only one patient died of the brain lesion, whereas the others died due to progression of the primary lesion or other metastases.

According to the five cases and our own case, the efficacy of radiotherapy remains unclear. Whole brain radiotherapy was provided in one case for multiple lesions, but there was no effect on tumor size. Postoperative conventional local radiotherapy was performed in two cases; these cases showed no local recurrence. Two cases, including our case, did not involve postoperative conventional local radiotherapy; both of the cases showed local recurrence 3 and 6 months after surgery. Gamma knife was used for treating multiple lesions in one case; later, the lesions disappeared on contrast-enhanced magnetic resonance imaging. We also...
administered gamma knife radiotherapy to cure the local recurrence in the cerebellum at the site of tumor removal. We are currently observing the patients to evaluate the treatment effects. It seems that local radiation including conventional local radiotherapy and gamma knife is more effective, whereas routine whole brain radiation does not seem to be the best choice for this carcinoma, considering the later complications.

**CONCLUSION**

We reported a rare case of brain metastasis from urachal carcinoma. According to the five previously reported similar cases and our case, tumor removal is the treatment of choice. Routine whole brain radiotherapy is not the best treatment choice; however, local radiotherapy including conventional local radiotherapy or gamma knife radiotherapy could be implemented instead.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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