Ovarian metastasis from a urachal adenocarcinoma primary in a young female

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ABSTRACT

Metastasis to the ovary from a bladder primary is extremely rare with limited case reports. The most common primary sites arise from stomach, colon or renal origin and usually occur from 40 years of age onwards. We report a case of an 18-year-old woman with a right sided Krukenberg tumour from a bladder primary. This patient is much younger than any other case report of Krukenberg tumour and reiterates the need for early excision of urachal masses in all age groups.

1. Introduction
Krukenberg tumours are a type of metastatic tumour to the ovary with a characteristic “signet ring” appearance. They/also almost always arise from the gastrointestinal tract with 70% arising from a gastric carcinoma. Other primary sites include the colon, breast, appendix, biliary tract and cervix. However, those arising from the urinary tract are rare, and usually consist of a transitional cell carcinoma from a bladder primary or mucinous adenocarcinomas from the urachus.

2. Case presentation
An 18-year-old Maori female, gravida 0, presented to our regional hospital with acute on chronic pelvic pain, dysmenorrhea but no haematuria. A computed tomography (CT) abdominal scan showed an incidental 3.2cm mass at the superior aspect of the bladder in keeping with a urachal remnant (Fig. 1). It was discussed at a Urology-Oncology multi-disciplinary meeting (MDM) and recommended for surgical removal. Intra-operatively she appeared to have an aggressive mass of the urachal remnant that had grown significantly since the CT scan along with peritoneal deposits. Complete oncological resection was unable to be performed. Therefore, excision biopsy and partial cystectomy occurred to gain a histological diagnosis.

Histology of the urachal remnant and peritoneal deposits showed a mucinous (colloid) adenocarcinoma arising in a urachal remnant with serosal ulceration and lymphatic invasion. Immunohistochemical stains showed strong positivity for CK20 and CDX2 and weak positivity for CK7 (Fig. 2).

The patient then presented one week later with severe right iliac fossa pain. A repeat CT scan demonstrated a large mass in the right adnexa concerning for a urinoma or haematoma. She underwent a laparotomy and the mass was a macroscopically malignant right ovary. An oophorectomy was performed. Histology of the ovary showed a mucinous adenocarcinoma similar to the previous specimen in keeping with a Krukenberg tumour (Fig. 3).

Given her extensive and irresectable disease, the MDM unanimously agreed that radical cystectomy was not an option and the patient was referred to medical oncology for further management.

3. Discussion
The urachus is a remnant that connects the bladder and umbilicus where it aids in draining urine during the 1st trimester. This usually closes during the 12th week of gestation and leaves a small fibrous cord. Urachal adenocarcinomas are rare as they account for only 0.5–2.0% of all bladder tumours. Unlike in our case, these occur more commonly in males who account for approximately 70% of all cases. Furthermore it is seen predominately in white patients (94%). Patients with a urachal adenocarcinoma have a 5-year survival rate of 43% mostly due to delayed presentation. Although there have been many systems proposed to best stage the outcomes of patients with urachal tumours, the Tumour Node Metastasis (TNM) staging system is the main predictor of
outcomes after surgery. Other staging systems that have been proposed are the Sheldon, Mayo and Ontario staging systems.

To our knowledge, there are only five reported cases of metastatic adenocarcinoma ovarian tumours originating from the urachus, but this is the first case of a patient this young, with previous cases being at least 40-years-old. The most common risk factors for urachal adenocarcinoma is haematuria and age >55. There are two reported cases of 10-year recurrence-free survival from Krukenberg tumours. These required multiple wide resections and multiple courses of chemotherapy. However, generally the prognosis is poor with no consensus on an optimal treatment regimen. Logothetis et al., treated three patients with doxorubicin, mitomycin-C and 5-fluorouracil, but found all patients passed away within 12 months. Quilty reported a 28-month recurrence free-survival in a single case treated with mitomycin-C, 5-fluorouracil and mitoxantrone combined with localised radiation to the bladder and extra-vesical tumour with 55Gy in 20 fractions over 4 weeks.

Surgical intervention is critical in these patients to help with disease-free survival. The gold standard of surgical resection for localized primary urachal adenocarcinoma is excision of the urachal remnant, umbilicus, partial cystectomy and bilateral pelvic lymph node dissection. It is important to note that 7% of all urachal adenocarcinomas can occur in the umbilicus so therefore wide excisions are required.

Surveillance post resection has not been described before in terms of Krukenberg tumours from a urachal primary. While ultrasound scan (USS) is usually the initial investigation for workup, CT is an overall more sensitive investigation. There is no determined surveillance schedule but an intense schedule such as a CT scan in 3 months, 1-year, 2-year and 3-years post resection may be required due to the aggressive nature of urachal primaries. As most patients with Krukenberg tumours from a urachal primary are greater than 40 years of age, the risk of using CT radiation for follow up does not outweigh the importance of surveillance. Given the young age of the patient in this case report, an alternate surveillance schedule may be required. In this instance we

Fig. 1. Transverse CT section of the 3.2cm urachal remnant (red arrow) and bladder (blue arrow); courtesy of Hawkes Bay District Health Board, Hastings, New Zealand. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Fig. 2. Mucinous carcinoma with neoplastic glands and extracellular mucin. Bulk of tumour located in region of urachus. Immunohistochemistry demonstrated positivity for CDX2 and CK20 (not shown). H&E original magnification ×100; courtesy of Hawkes Bay District Health Board, Hastings, New Zealand.
recommend using MRI to reduce the burden of ionising radiation from CT.

4. Conclusion

Krukenberg tumours from a urachal primary are extremely rare with no general consensus on treatment and very poor outcomes. A multi-disciplinary approach is required involving surgeons, oncologists and radiotherapists. While there are no universal guidelines on management, early wide excision is the best initial management. However no follow up surveillance schedule has been studied or approved.

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Declaration of competing interest

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