

## Clinical Features of Urachal Carcinoma in Japan: Review of 157 Patients

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**Summary.** Carcinoma of the urachus is not as uncommon as previously considered. 157 cases diagnosed to have urachal carcinoma collected from the Japanese literature were reviewed and the findings were compared with those reported in the English literature. The incidence of the disease ranges from 0.55 to 1.2% of bladder tumours in Japan in contrast with 0.07 to 0.70% in the Western countries. Males accounted for 72% of the patients. The highest age incidence occurred between fifth and sixth decades. The commonest presenting symptom was haematuria (71%). Adenocarcinoma accounted for 88% of the tumours most being mucous producing. Various treatment modalities were used, however, and the prognosis remained uniformly poor. An analysis of 66 patients with known outcome revealed an overall 5-year survival rate of 6%. The authors conclude that the disease tends to have a relatively higher incidence in Japan (Far East). When comparing the parameters of sex, age, presenting symptom, histopathology, treatment and prognosis, urachal carcinoma appears to have the same characteristics in the Far East as in Western countries.

**Key words:** Urachal carcinoma, Clinical feature in Japan, Comparison with Western countries.

### Introduction

Carcinoma of the urachus arises from the embryonic rests of glandular cells that remain after the process of urachal involution. The most common malignancy of the urachus is adenocarcinoma. There are, at present, 154 cases of urachal carcinoma reported in the English literature [4, 6, 10, 12–14, 18–20, 23, 25]. We herein review 157 cases of urachal carcinoma collected from the Japanese literature and compare our findings with those reported in the Western countries.

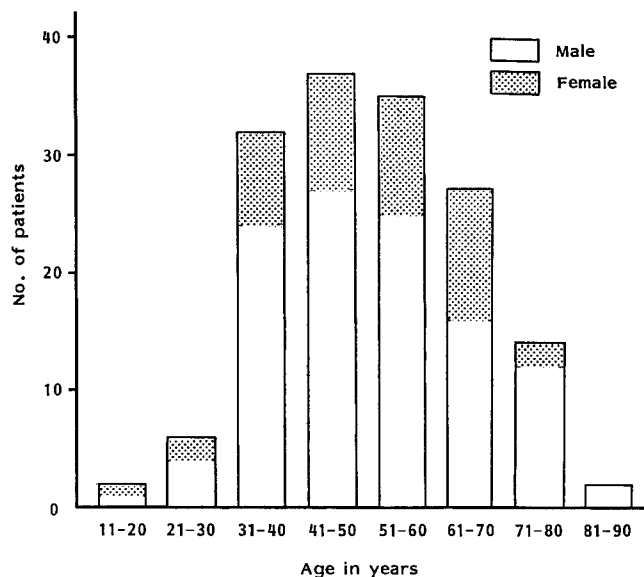


Fig. 1. Sex and age distribution of 155 patients

### Review of the Literature

From 1932 to 1981 (49 years) Japanese literature related to the urachal carcinoma was reviewed. Excluding duplications and ambiguous reports, 157 cases appeared to be fit for this study. Data were collected with reference to the incidence, sex, age, presenting symptom, histopathology, treatment and prognosis. Only 66 patients had sufficient data to permit analysis of survival.

### Sex and Age Incidence

Males accounted for about 72% of the patients, whereas 28% were females; the ratio being 2.6:1. The ages ranged from 19 to 82 years with a mean age of 51.6 years in males and 51.2 years in females. The highest age incidence occurred between the fifth and sixth decades (46.5%). One-fourth of the patients (25.8%) were less than 40 years old (Fig. 1). The youngest patient was a 19 year-old female and the oldest an 82 year-old male. The sex and age of two patients are not known.

**Table 1.** Presenting symptoms of patients

Symptoms <sup>a</sup>	No. of patients (%)
Haematuria	112 (71)
Suprapubic mass	18 (11)
Dysuria, Frequency	16 (10)
Lower abdominal pain	9 ( 6)
Mucous in urine	6 ( 4)
Necrotic debris in urine	2 ( 1)
Umbilical discharge	1 ( 1)
Unknown	14 ( 9)

<sup>a</sup> Some patients had more than one symptom

**Table 2.** Histopathology of tumours

	No. of patients (%)
Adenocarcinoma	138 ( 88)
Transitional cell carcinoma	5 ( 3)
Squamous cell carcinoma	3 ( 2)
Anaplastic carcinoma	6 ( 4)
Mixed type	5 ( 3)
Total	157 (100)

**Table 3.** Treatment of patients

	No. of patients (%)
Partial Cystectomy ± Radiation and/or Chemotherapy	89 ( 57)
En bloc Resection ± Radiation and/or Chemotherapy	33 ( 21)
Total Cystectomy ± Radiation	8 ( 5)
Radiation + Chemotherapy	4 ( 2)
Radiation	3 ( 2)
Chemotherapy	1 ( 1)
Unknown	19 ( 12)
Total	157 (100)

**Table 4.** Crude survival data of 66 patients with adenocarcinoma of the urachus

Years	No. of patients alive	percentage of patients alive
1	28	42.4
2	15	22.7
3	9	13.6
4	5	7.6
5	4	6.0
6	4	6.0
7	3	4.5
8	2	3.0
9	1	1.5
10	1	1.5

## Clinical Features

The clinical manifestations of urachal carcinoma frequently resemble those of bladder tumours (Table 1). The most common symptom was haematuria (71%), followed in order of frequency by suprapubic mass (11%), dysuria and frequency (10%) and lower abdominal pain (6%). The presence of mucous material in urine was uncommon. Some patients presented with more than one symptom. The symptomatology is not known in 14 patients.

## Histopathology

Histological evaluation of the tumours showed that the majority, i.e. 138 (88%), were adenocarcinomas. Of these, 88 (64%) were mucous producing. This status is not known in 26 patients. Other histological varieties of the tumour were noted with less frequency (Table 2).

## Treatment and Prognosis

Various modes of therapy were used in this series of patients (Table 3). Eighty-nine patients were treated by partial cystectomy and 33 patients had en bloc segmental resection. The latter entails removal of the tumour and contiguous bladder wall with transversalis fascia and peritoneum along the urachal tract with or without the umbilicus. Only in 8 patients was more aggressive surgery, the (total cystectomy and urinary diversion) used.

In 29 patients radiation with or without chemotherapy was employed as an adjunct to surgery. The most common chemotherapeutic agent used was 5-fluorouracil. In 19 patients, treatment is not known and the remainder was dealt with by radiation and/or chemotherapy alone. The outcome was known in 66 of the 157 patients, which provided the basis for analysis of survival. The crude survival of these patients is shown in Table 4.

In spite of different treatment trials, the prognosis was uniformly poor. The overall 3- and 5-year survivals were 13.6% and 6.0% respectively. The average survival of the 66 patients was 17 months.

## Discussion

Adenocarcinoma of the bladder can be classified into 3 categories, depending on its origin, as primary, metastatic and urachal adenocarcinomas.

Primary adenocarcinoma of the bladder may occur at any site in the bladder, but is frequently located in the base or lateral walls [16]. It may be caused by irritation of the urothelium associated with infection, trauma and obstruction [1, 22]. Wheeler and Hill [22] suggested the following criteria for diagnosis of this lesion: 1. location in the base or lateral walls of the bladder, 2. presence of cystitis cystica or glandularis and, 3. presence of a transition from non-malignant urothelium to adenocarcinoma.

Metastatic adenocarcinoma of the bladder usually originates from a primary lesion in the colon, prostate or female genital tract.

Urachal adenocarcinoma originates from the epithelial mucosa of the urachal remnant. It is located in the dome or anterior bladder wall. The criteria for the diagnosis of this disease entail [16, 22]: 1. location in the dome or

**Table 5.** Incidence rates of primary adenocarcinoma and urachal carcinoma as a percentage of all bladder tumours in several populations

Country	Reference	Source of data	Total bladder tumors	No. of patients (incidence)	
				Primary adenocarcinoma	Urachal carcinoma
United States <sup>a</sup>					
Iowa	Jacobo et al. [9]	Bladder Tumor Registry	2,628	20 (0.76)	6 (0.23)
Seattle	Jones et al. [11]	Virginia Mason Medical Center	648	10 (1.54)	.....
England					
Bristol	Miller et al. [15]	Bladder Tumor Registry	1,437	.....	1 (0.07)
Sheffield	Thomas et al. [21]	United Sheffield Hospitals	5,300	25 (0.47)	18 (0.34)
Sweden	Öhman et al. [18]	Cancer Registry of Sweden	6,000	50 (0.83)	10 (0.17)
West Germany					
Mainz	Jakse et al. [10]	University of Mainz	715	13 (1.8)	5 (0.70)
Hong Kong	Yu and Leong [25]	Queen Mary Hospital	290	.....	1 (0.34)
Japan	Ichikawa [8]	38 University Hospitals	1,018	40 (3.93)	12 (1.18)
Shikoku	Present Study	Cancer Center	365	6 (1.64)	2 (0.55)

<sup>a</sup> Also Massachusetts, Cornil et al. [5]: Urachal carcinoma comprises 0.01% of all malignant diseases

anterior wall of the bladder, 2. absence of cystitis cystica and glandularis, 3. presence of a suprapubic mass, 4. invasion into muscular or deeper layers, 5. presence of urachal remnant, 6. sharp demarcation between tumor and surface epithelium, 7. deep ramification into the bladder wall with extension to the space of Retzius, abdominal wall or umbilicus.

A small number of adenocarcinomas of the bladder are of urachal origin. Recent data from the Cancer Center of Shikoku Island in Japan shows that of 365 cases of bladder tumour, there were 6 cases of primary and 2 cases of urachal adenocarcinoma, comprising, respectively, 1.64% and 0.55% of the bladder tumours in this district. In Table 5 the incidence rates of primary adenocarcinoma of the bladder and urachal carcinoma derived from the available literature are summarized. It can be seen that the incidence of urachal carcinoma ranges from 0.07% to 0.70% of the bladder tumours in Western countries in contrast with 0.55% to 1.2% of the bladder tumours in Japan. This reveals that the disease tends to have a relatively higher frequency in Japan than in Western countries.

In our series, 72% of the patients were male subjects which is comparable to the 80% incidence reported in Western literature [23, 17]. The ages ranged from 19 to 82 years old. Cornil et al. [5] reported the youngest patient, a 15 year-old girl, and Bandler et al. [21] the oldest, an 83 year-old male. The highest incidence was between the fifth and sixth decades which corresponds to that reported in Western literature [17, 20]. One-fourth of our cases (25.8%) were less than 40 years old. This is suggestive of an increased frequency of the disease in the younger age group.

Clinical features of urachal carcinoma frequently resemble those of tumours of the bladder, i.e. haematuria, dysuria and frequency (Table 1).

In our series 71% of the patients had haematuria. Suprapubic mass, dysuria and frequency and abdominal pain were noted with less frequency. The passage of mucous in the urine has been considered pathognomonic for adenocarcinoma of the urachus [24], but, was observed rarely in this series. Beck et al. [3] reviewing 78 cases of urachal carcinoma from the English literature also found the majority of patients (78%) to have haematuria, while the other symptoms were infrequent. Rubell and Porges [20] reported a case of urachal carcinoma in which the presenting symptom was vaginal bleeding.

Histological distribution of the tumours in our series showed the majority (88%) to be adenocarcinomas mostly of the mucous producing type. This is consistent with that of 80 to 90% reported in Western countries [6]. The diagnosis of urachal carcinoma is usually suspected at cystoscopic examination whenever a lesion at the bladder dome is encountered. Subsequent tumour biopsy may show adenocarcinoma; however, in most cases the site of the tumour origin is established only at operation. Radiographic examination, i.e. IVP and cystogram may show deformity of or a filling defect in the bladder. Ultrasonography and computerized axial tomography have been reported to be worthwhile in the preoperative establishment of the site of the tumour origin in urachal carcinoma [14, 7]. The treatment of urachal carcinoma is always surgery. En bloc removal of the tumour with contiguous bladder wall, transversalis fascia and peritoneum along the urachal tract is reported to be the best choice of treatment with a 5-year survival rate of 25% [23].

In our series various modes of therapy were used (Table 3); however, the prognosis remained uniformly poor. In 66 of the 157 patients, an overall 5-year survival of 6% was obtained (Table 4). Rubell and Porges [20] reviewing

142 cases of urachal carcinoma from the English literature stated that the 5-year survival varied from 6.5% to 16%, but even in the best series only 6% were tumour-free at the end of 5 years.

In conclusion, carcinoma of the urachus appears to have a relatively higher incidence in Japan (Far East) than in Western countries. When comparing the parameters of age, sex, presenting symptom, histopathology, treatment and prognosis, the same characteristics in the Far East were found as in the Western countries.

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