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PRIMARY URACHAL ADENOCARCINOMA

Li Guanghui 李光辉　Hou Jianguo 候建国　Qian Songxi 钱松溪
Xia Guangbin 夏广彬　Ma Yongjiang 马永江

Department of Urology, Changhai Hospital, the Second Military Medical University, Shanghai 200433

Objective: To study the clinical characteristics, diagnosis and treatment of primary urachal adenocarcinoma. Methods: Six cases of histologically confirmed primary urachal adenocarcinoma were retrospectively reviewed. Results: The median age of this series was 55. Incidence duration was 4 to 7 months. Tumors were found in all patients by cystoscopy, CT and B-ultrasound. Urachal adenocarcinoma in 4 cases and bladder cancer in 2 cases were diagnosed before operation. The segmental resection of tissues around the mass was performed in 5 cases, all patients received chemotherapy or radiotherapy. Conclusion: It was difficult to confirmed diagnosis early, and differential diagnosis from bladder cancer was important. The wide surgical excision should be taken with adjuvant chemo-or radiotherapy.

Key words: Urachus, Neoplasm, Diagnosis, Treatment

Primary urachal adenocarcinoma is a rare neoplasm. The incidence, according to a large number of reports from different authors, is between 0.34 to 0.7 percent of bladder tumors.[1,2] This paucity of cases has obviously limited any unanimity of opinion regarding the clinical and pathologic features that are necessary to firmly establish the diagnosis of this rare malignancy, its usual natural history, and the optimum therapeutic approach. Lane[3] has suggested that this tumor has a rapidly progressive course and that only 6.5 to 16 percent of patients continue to live for five years. Others have also described the results. All this has thus prompted us to review our total experience with this malignancy.

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CLINICAL MATERIALS

The records of 1,785 patients with bladder cancer collected by Changhai Hospital in Shanghai during the 21-year period from 1975 through 1996 were reviewed. The records of 23 were related to urachal adenocarcinoma after the histologic material and clinical findings were carefully re-reviewed.

Among the clinical findings two patients were men and four were women. Ages ranged from thirty-one to seventy-seven (median age fifty-five years). Incidence duration was 4 to 7 months (median time 5.4 months). Symptoms and signs were painless gross hematuria in 6 cases, dysuria and frequency in 2 cases, a painful suprapubic mass in 5 cases (the diameter of the mass 3~5×4 cm), and lower abdomen pain in 4 cases. Tumors were found in 6 cases while the operation was performed by cystoscopy, CT and B-ultrasound. Urachal adenocarcinoma in 4 cases and bladder cancer in 2 cases were diagnosed before operation.

In each instance the tumor had been described as arising either in the midline of the vertex or in the posterior wall of the bladder. Most of the tumors were intramural, had deep ramifications in the bladder wall, and extended into the perivesical fat. Overlaid remnants of normal or focally ulcerative surface epithelium were devoid of polypoid proliferations. The tumors overlaid with cystitis cystica and cystitis glandularis were marked only in one patient, but transition from these areas of proliferating cystitis to

malignancy was absent. In 6 patients the tumors were of a colloid type; in 4 cases the tumors were of a colloid type; and in one instance each tumor consisted predominantly of either signet-ring cells or oat cells. In 2 cases signet-ring cells were admixed with colloid or colloid features. The tumors were well differentiated in 3 patients, moderately differentiated in one patient, and poorly differentiated in 2 patients. In none of the 6 tumors were urachal remnants identified.

RESULTS

Partial cystectomy and segmental resection of tissues around the mass were an integral part of the primary treatment of 5 patients.

Chemotherapy was used in 5 patients without appreciable benefit after operation. In 4 patients a combination program was tried which consisted of 5-fluorouracil, doxorubicin hydrochloride (Adriamycin) and mitomycin C (FAM); this was the primary therapy for one patient, then was a treatment of pelvic lymph node metastasis for 2 patients, and an adjuvant therapy following partial cystectomy for one patient. Two patients, one of whom had obtained a partial response initially to FAM, were treated for nodal metastasis with a combination of cisplatin, cyclophosphamide, and doxorubicin (CISCA).

Three patients died after operation as a direct result of their malignancy. After periods ranging from six months to three years (half a year, 2.5 years and 3 years respectively), the median interval from diagnosis to death was two years. The other 2 patients remain alive and have been free of disease for periods ranging from two to seven years. However, in one of the two living patients local recurrences developed so as to require further treatment, i.e., endoscopic resection two years later. In another patient a poorly differentiated adenocarcinoma developed in the lung, combined with a distant metastatic disease, white walls removed by wedge resection seven-years after partial cystectomy; she has been alive without disease for three years.

DISCUSSION

It has been suggested that malignancies classified as urachal tumors should meet the following criteria: (1) the tumor in the vertex of the bladder; (2) the absence of cystitis cystica and cystitis glandularis; (3) the invasion of muscle or deeper structures and either intact or ulcerated epitherlium; (4) the presence of urachal remnants; (5) the presence of a suprapubic mass; (6) a sharp demarcation between tumor and normal surface epithelium; and (7) the tumor growth in the bladder wall branching into the space of Retzius.[3] However, the data based on our findings and also on the results from other studies suggest that these criteria are too restrictive. Certainly not all urachal tumors need to arise at the vertex of the bladder, since Schubert, Pavkovic, and Bethke-Bedurftig[4] observed urachal remnants in the midline of the vertex in only 54 per cent of their patients, and identified such remnants in the posterior wall in 44 per cent. Since inflammation and cystitis cystica go hand in hand, one may occasionally encounter the area of cystitis cystica in the bladder with urachal carcinomas. The presence of proliferating cystitis should not exclude a diagnosis of urachal carcinoma unless a definite transition to malignancy is demonstrated. Although the presence of urachal resta in association with a malignancy might aid the pathologist in making the diagnosis, none was demonstrated in our patients, presumably due to the extent of the tumors.

The most important considerations in making a diagnosis of urachal carcinoma are: (1) the location of the tumor in the bladder wall; (2) the findings of a sharp demarcation between the tumor and the surface epithelium; and (3) the exclusion of a primary adenocarcinoma located elsewhere that has spread secondarily to the bladder. Although most urachal tumors have been mucin-secreting adenocarcinomas, signet-ring cell carcinomas and squamous cell carcinomas have also been described.[2,5]

The pathogenesis of urachal carcinomas remains to be poorly understood. Schubert, et al.,[4] who examined the 122 bladders obtained at autopsy, found intramural tubular urachal remnants in 32 percent. In one-third of these bladders the usual transitional epithelium lining the urachal remnants had been replaced by columnar cells, either as metaplasia or as superficial cells above the transitional cells. In no instance, however, was a communication demonstrated between the intramural remnant and the interior of the bladder. The authors reasoned that, even if the urachal canal reopened, the reflux of urine into the urachal remnants would remain impossible because of their tiny diameters and the presence of epithelium secretions that occluded their lumen. Consequently, it can explain the development of urachal carcinomas on the basis of urachal

carcinogens present within the urine. Therefore, carcinogenic factors seem to be different from those responsible for the adenocarcinomas of the renal pelvis, ureters, and bladder that must call for an operation.

It was interested in another difference that whereas adenocarcinomas arising in the renal pelvis occur predominantly in women and those developing in the bladder have a relatively high incidence in women when compared with transitional cell carcinomas, urachal adenocarcinomas occur most frequently in men, and similar findings have also been reported by other investigators.

Prognosis appears to be little influenced either by the histologic appearance of the tumor or by its grade. We could demonstrate no association between prognosis and (1) the cell type (colloid versus colonic); (2) the degree of tumor differentiation; or (3) the presence of signet-ring cells. In our group, only 2 cases are alive for over five years (20%).

Analysis of treatment failure reported in the literature reveals that postoperative failures occur locally in 47 to 51 percent of patients.[5,6] The main sites of distant metastasis, in order of decreasing frequency, are the lymph nodes, lung, and peritoneal cavity. Consequently, wide surgical excision, consisting of an en bloc removal of the umbilicus, ligamentum commune, and the involved portion of the urinary bladder, combined with bilateral pelvic lymph node dissections, appears mandatory if one wants to reduce the incidence of treatment failures.

Although we have used in several instances an integrated approach consisting of preoperative radiotherapy and partial cystectomy, the number of patients so treated is too small to determine its effect on the rate of survival. However, the results[7] suggest that partial cystoectomy can be used safely and without increased complications, whither in an integral program or in a salvage treatment for those who fail definitive radiotherapy. Radical cystoectomy should be reserved for patients whose adequate surgical margins are not feasible by segmental resection.

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