CASE REPORT

Primary adenocarcinoma of the rete testis with elevation of serum carcinoembryonic antigen: a case report

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Abstract Primary adenocarcinoma of the rete testis, which arises from the collecting systems of the testis, is an extremely rare tumor with a poor prognosis. A case of primary adenocarcinoma of the rete testis with elevation of serum carcinoembryonic antigen (CEA) levels is reported. A 59-year-old man presented with left scrotal swelling. Ultrasonography showed a left hydrocele and swelling of the left testis with calcification. Computed tomography showed para-aortic lymph node swelling and multiple lung metastases. Germ cell tumor markers were within the normal range. Radical orchiectomy was performed, and histological examination showed adenocarcinoma. The tumor cells were immunohistochemically positive for CEA. There was no evidence of other primary carcinomas. The tumor was finally diagnosed as a primary adenocarcinoma of the rete testis. The patient had bone metastases 8 months after orchiectomy with increased serum CEA levels. CEA might be a marker for primary adenocarcinoma of the rete testis.

Keywords Adenocarcinoma · Rete testis · Hydrocele · Carcinoembryonic antigen · Prognosis

Introduction

Primary adenocarcinoma of the rete testis is a rare malignant tumor with a poor prognosis. This neoplasm was first

reported by Feek and Hunter in 1945 [1]. Seventy-two cases have been reported to the best of our knowledge, but standard treatment has not been yet established. This is a case report of adenocarcinoma of the rete testis with elevation of serum carcinoembryonic antigen (CEA) levels, and the clinical features and outcomes of adenocarcinoma of the rete testis are summarized based on this case and previous reports.

Case report

A 59-year-old man presented with swelling of the left scrotum. He noticed induration of the left testicle without scrotal pain or low back pain a few months earlier. He had no history of genitourinary infection or testicular trauma. Ultrasonography and computed tomography (CT) showed a left hydrocele and calcification of the left testis (Fig. 1). CT also showed para-aortic lymph node swelling and multiple lung metastases. Serum β -human chorionic gonadotropin, α -fetoprotein, lactate dehydrogenase, and interleukin-2 receptor were negative. First, radical orchiectomy including hydrocele was performed for a primary testicular germ cell tumor. The testicular tumor measured 5.5×3.0×2.5 cm. There was a solitary hard nodule (diameter 1.8 cm) in the left spermatic cord. Histological examination of the testis showed adenocarcinoma developed from the rete testis and atypical cells in the epithelium of the rete testis (Fig. 2a). The tumor cells were immunohistochemically positive for CEA (Fig. 2b). Serum CEA (24.1 ng/ml) and carbohydrate antigen 19-9 (CA19-9) (187.1 U/ml) levels were elevated. Positron emission tomography-CT, gastrointestinal endoscopy, and colonoscopy were performed, and there was no evidence of other primary carcinomas. The tumor was finally diagnosed as a primary adenocarcinoma of the

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Fig. 1 Abdominal computed tomography shows left testicular swelling. The tumor has calcification and is surrounded by a hydrocele in the left scrotum

rete testis. The patient refused any additional therapy. He had bone metastases 8 months after radical orchiectomy and received palliative care 14 months after surgery. Both CEA and CA19-9 levels increased with disease progression (Fig. 3).

Discussion

Primary adenocarcinoma of the rete testis is a rare malignant tumor that arises from the collecting systems of the testis. It has a poor prognosis and, with 3- and 5-year disease-free survival rates of 49 and 13%, respectively [2]. Patients with this tumor have no specific clinical symptoms except testicular swelling, and tumor markers of testicular germ cell tumors are negative. The diagnosis of this tumor is based on verifying that the adenocarcinoma developed from the rete testis and excluding other primary adenocarcinomas, such as digestive system cancer [3, 4].

Fig. 3 Change of serum carcinoembryonic antigen (CEA) levels after radical orchiectomy. CEA levels increase rapidly after bone metastases

To analyze the prognosis and management of adenocarcinoma of the rete testis, 72 cases published between 1945 and 2015 were collected using PubMed. Cases without a description of the follow-up period were excluded. Finally, 51 cases including the present one were reviewed. Demographic and clinical data were evaluated. Overall survival rates were estimated by the Kaplan–Meier method, and the log-rank test was used to compare data.

The median age was 57 years (range, 17–91 years). Adenocarcinoma of the rete testis occurred at an older age compared to testicular germ cell tumors [5]. The most common symptom of adenocarcinoma of the rete testis was a scrotal mass, found in 49 (96%) of the documented cases. Twenty-four cases (47%) had a history of hydrocele. Twenty-four (47%) and 27 (53%) patients had tumors on the left and

Fig. 2 **a** Histological findings of the testicular tumor show adenocarcinoma that developed from the rete testis. Atypical cells are found in the epithelium of the rete testis. **b** Immunostaining with specific

carcinoembryonic antigen (CEA) antibody shows that the tumor cells in the testis are positive for CEA

right sides, respectively; there were no cases of bilateral tumors. Sixteen cases (31%) had metastases at presentation. In the remaining 35 cases, 18 developed metastases after radical orchiectomy during the median follow-up of 12 months. Of 34 cases with metastases, the metastases occurred in the lung (19 cases, 56%), retroperitoneal lymph nodes (16 cases, 47%), skin (7 cases, 21%), liver (5 cases, 15%), and bone (4 cases, 12%).

In the present case, the serum CEA level was elevated, and it increased rapidly after bone metastases. Serum CEA levels were evaluated in 7 case reports [2, 6–10]; 4 of them had elevated serum CEA levels. Three of these cases had metastases at presentation [2, 8], and one had local recurrence, retroperitoneal lymph node swelling, and multiple lung metastases 13 months after orchiectomy [9] (Table 1). Three patients with normal serum CEA levels were alive during the observation period [6, 7, 10]. In four cases with immunostaining for CEA, the tumor cells were positive for CEA [2, 6, 8]. Thus, CEA might be a biomarker that reflects the progression of adenocarcinoma of the rete testis.

The present case also showed elevation of serum CA19-9 levels, which increased progressively with disease progression. Okada et al. reported that the serum CA19-9 level was within normal limits, but the tumors were strongly positive for CA19-9 immunostaining [6]. However, there have been few reports about the association of adenocarcinoma of the rete testis with CA19-9. Further cases are necessary to clarify the meaning of CA19-9 as a biomarker.

Radical orchiectomy was performed in all patients. Thirty patients (59%) underwent further treatments combined with radiation therapy (29%), chemotherapy (29%), and retroperitoneal lymph node dissection (RPLND) (18%). Twenty-one cases (41%) died during the median follow-up of 11 months (range, 1 to 84 months). The overall survival rates at 2, 3, and 5 years were 55%, 50%, and 25%, respectively, findings similar to the previous report in 1995 [2]. Several chemotherapy regimens were used for the treatment of metastatic adenocarcinoma of the rete testis, such as multi-agent chemotherapy consisting of bleomycin, etoposide, and cisplatin [5]; or paclitaxel, ifosfamide, and cisplatin [9]. Unfortunately, adenocarcinoma of the rete testis responded poorly to both chemotherapy and radiotherapy. Most patients with metastases died within 2 years after radical orchiectomy.

In conclusion, a case of adenocarcinoma of the rete testis with elevation of serum CEA levels was reported. Adenocarcinoma of the rete testis is an aggressive malignant tumor. It is difficult to detect in its early stages because it tends to occur at an older age than testicular germ cell tumors, and because it is often accompanied by a hydrocele. A patient who has a testicular tumor with a hydrocele and negative germ cell tumor markers should be suspected

Table 1 Patients with adenocarcinoma of the rete testis, who received serum CEA evaluation

Case	References (year)	Age (year)	Tumor side	Clinical symptoms	Serum marker			Immunostain- ing CEA	Metastases at presentation	Follow-up	Outcome (M)
					AFP	hCG	CEA				
1	Sanchez (1995)	74	Lt	Scrotal mass Hip pain Lymphangitis	–	–	+	+	Bone	8	Dead
2	Okada (1997)	60	Lt	Scrotal mass	–	–	–	+	None	60	Alive
3	Hagiuda (2010)	80	Lt	Scrotal mass	–	–	–	NA	Lymph node Lung	8	Alive
4	Lee (2013)	54	Rt	Flank pain	–	–	+	+	Lymph node Lung	8	Dead
5	Chovanec (2014)	47	Lt	Scrotal mass	–	–	+	NA	None	51	Dead
6	Ghosh (2015)	32	Rt	Scrotal mass	–	–	–	NA	None	6	Alive
7	Present	59	Lt	Scrotal pain Scrotal mass	–	–	+	+	Lymph node Lung	14	Alive

CEA carcinoembryonic antigen, AFP α -fetoprotein, hCG human chorionic gonadotropin, NA not available

of having adenocarcinoma of the rete testis; CEA might be a marker for this rare tumor.

Compliance with ethical standards

Conflict of interest Authors declare that they have no conflict of interest.

Research involving human participants and/or animals The study was approved by the Ethics Committee of Tohoku University Hospital.

Informed consent Informed consent was obtained from the patient included in the study.

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