



## **PRIMARY ADENOCARCINOMA OF RETE TESTIS; REPORT OF A RARE NEOPLASM**

### **Nadir bir Rete testis tümörü; primer adenokarsinom**

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#### **ABSTRACT**

Primary adenocarcinoma of rete testis is a very rare neoplasm of elderly males. Herein, a case of primary adenocarcinoma of rete testis in a 43 years old man is being reported who presented with a left sided scrotal mass. Scrotal ultrasonography suspected a malignant growth and a diagnosis of a poorly differentiated carcinoma was made on fine needle aspiration cytology. Radical orchiectomy was done. Histopathological examination revealed a primary papillary adenocarcinoma of rete testis. On follow up, he presented with metastasis in left inguinal lymph node. Block dissection of the inguinal nodes was done, and the patient has remained disease free for 1 year postoperatively.

**Key words:** Adenocarcinoma, rete testis, orchiectomy, and treatment.

#### **ÖZET**

Yaşlı erkeklerde primer rete testis adenokarsinomu nadiren karşılaşılan bir kanserdir. Burada, 43 yaşındaki bir erkek hastamızda sol testiste saptanan rete testis adenokarsinom olgusu sunuldu. Sol testisinde ultrasonografi ile kitle saptanan hastaya yapılan ince iğne aspirasyon biyopsisi sonucunda kötü diferansiye karsinoma tanısı konuldu. Radikal orşiektomi yapılan hastada histopatolojik olarak rete testisin primer adenokarsinomu tanısı kondu. Takip sırasında, sol kasık bölgesinde lenfatik metastaz saptanan hastaya bölgesel lenf nodu diseksiyonu yapıldı ve hasta bir yıldır hastaliksız olarak izlenmektedir.

**Anahtar kelimeler:** Adenokanser, rete testis, orşiektomi, tedavi.

#### **INTRODUCTION**

Primary adenocarcinoma of rete testis is a rare neoplasm seen in elderly males. Less than 65 cases have been reported so far (1,2,3). Rete testis is a testicular adnexal structure, located at the hilum of the testis with a complex tubular structure. This carcinoma needs to be distinguished from adenocarcinoma of the epididymis, malignant mesothelioma of tunica vaginalis and from a metastatic germ cell tumor deposit (4).

#### **Case**

A 53 year old man presented with a gradually enlarging swelling and dragging pain in the left

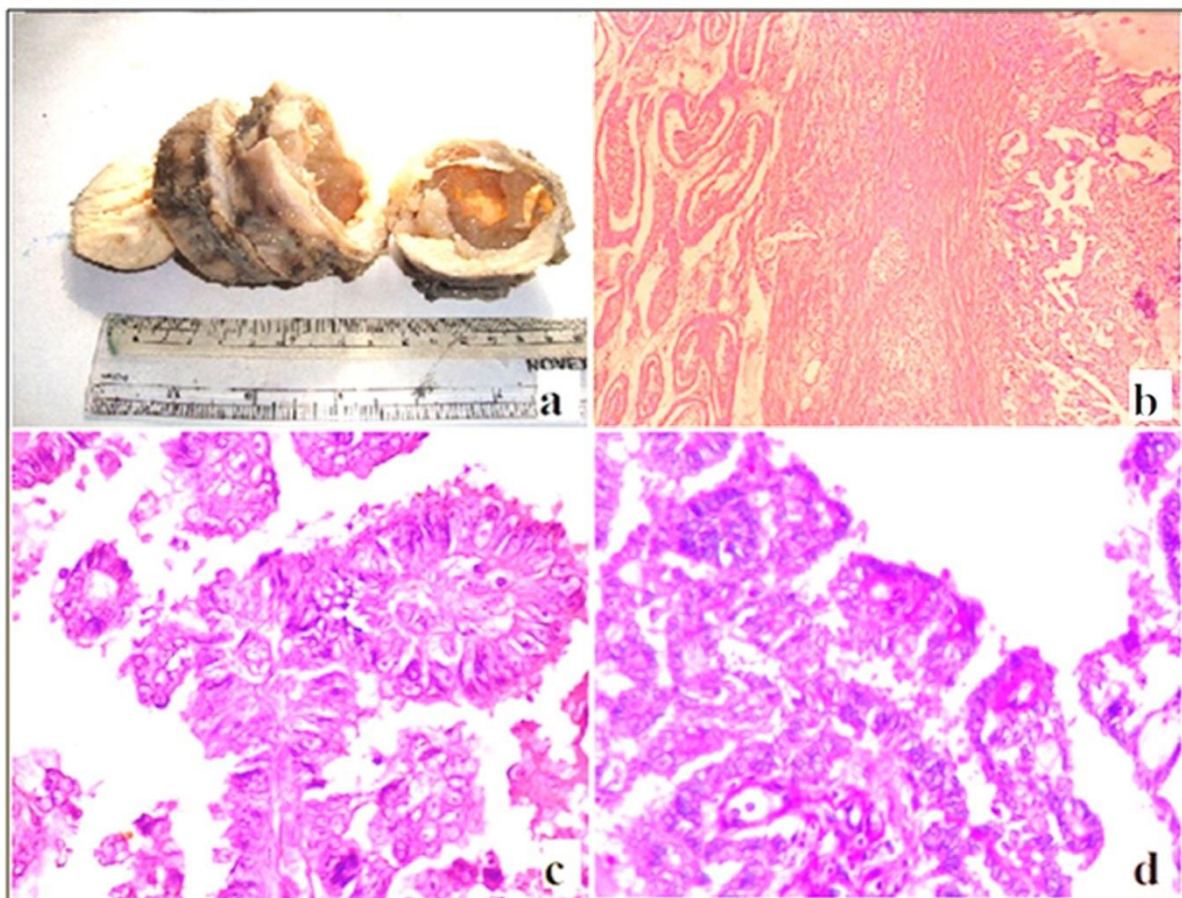
side of scrotum for last 5 months. On local examination, a hard mass (4.5x4 cm) with mild tenderness was palpable in central part of left side of the scrotum.

Scrotal ultrasonography (USG) showed a nodular soft tissue mass while USG of whole abdomen was unremarkable. Fine needle aspiration cytology (FNAC) of the mass revealed discretely arranged large polygonal pleomorphic cells and a provisional diagnosis of poorly differentiated carcinoma was given on cytologic smear examination. Left sided orchiectomy was done without retroperitoneal nodal dissection. Grossly, a left orchiectomy specimen was received (4.5x4 cm). On cut section, a well circumscribed, encapsulated, grey-white, solid, homoge-

nous tumor (4.5x4x2.5 cm) was seen (Figure 1a), occupying almost the whole of testis. No normal testicular and adnexal structure could be identified on gross. The adenocarcinomatous zone was visible just beside the normal rete epithelium (Figure 1b). Histologic sections from tumor showed a partially encapsulated tumor with polygonal cells arranged in tubules and papillary configuration (Figure 1c). The pleomorphic cells had moderate cytoplasm with vesicular nuclei (Figure 1d). Mitotic figures were low.

Based on morphology, a list of differential diagnoses like primary adenocarcinoma of rete testis, primary adenocarcinoma of epididymis, malignant mesothelioma of tunica vaginalis and metastatic germ cell tumor were included. Periodic acid Schiff (PAS)

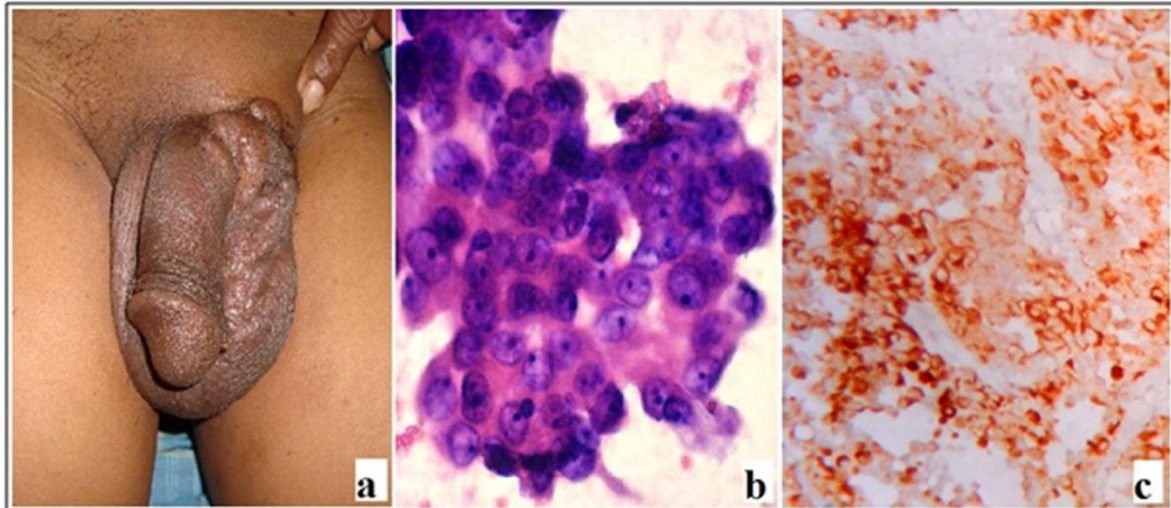
stain and immunohistochemistry (IHC) were suggested for confirmation. With IHC, the tumor cells were diffusely positive for pancytokeratin and carcinoembryonic antigen (CEA) (Figure 2). No seminiferous tubules were identified (Figure 1d). Result of periodic acid Schiff (PAS) stain was also positive (Figure 1d). The tumor cells were immunonegative for desmin, alpha fetoprotein (AFP), human chorionic gonadotrophin (hCG), placental alkaline phosphatase (PLAP). The possibilities of primary adenocarcinoma of epididymis (CEA -), malignant mesothelioma of tunica vaginalis (PAS -, CEA -) and metastatic germ cell tumor (AFP +, hCG +, PLAP +) were thus excluded. A final diagnosis of primary adenocarcinoma of rete testis was established.



**Figure 1:** Grossly, a left orchietomy specimen was 4.5x4 cm. On cut section, a well circumscribed, encapsulated, grey-white, solid, homogenous tumor (4.5x4x2.5 cm) was seen (a). The adenocarcinomatous zone showed just beside the normal rete epithelium (b). Histologic sections from tumor showed a partially encapsulated tumor with polygonal cells arranged in tubules and papillary configuration (c). The pleomorphic cells had moderate cytoplasm with vesicular nuclei (d). Mitotic figures were low.

Post-operative radiotherapy or adjuvant chemotherapy was not given as treatment protocol remains controversial due to inadequate study in small number of cases diagnosed so far. Eight months later

he presented with left inguinal lymphadenopathy (2x2 cm) and FNAC of the node proved metastasis (Figure 2). Block dissection of the inguinal nodes were done and one year follow up period was uneventful.



**Figure 2:** Left inguinal lymph node metastasis. Periodic acid Schiff (PAS) stain and immunohistochemistry (IHC) were suggested for confirmation. With IHC, the tumor cells were diffusely positive for pancytokeratin and carcinoembryonic antigen (CEA)

### DISCUSSION

Primary adenocarcinoma of rete testis is a rare neoplasm. It usually occurs in men over 60 years though the age range can vary from 17 to 91 years (2). Pathogenesis of this lesion remains unclear. Gruber et al found relationship with chemical exposure like lead, carbon dioxide (5). Our patient was relatively younger, aged 43 years without any definite history of chemical exposure. A painful scrotal mass is the most common presentation, another 25% cases present with a hydrocele (6, 7). Grossly, adenocarcinoma of rete testis is usually solid but may be cystic too (6, 7). Different histologic types are tubular, papillary, cribriform and solid. The tumor cells are columnar to cuboidal with moderate amount of eosinophilic or amphophilic cytoplasm. Nuclear stratification with moderate pleomorphism is noted. Mitotic activity is frequent. The stroma may be prominent, extensively desmoplastic or extensively hyalinised (4). Cases have been seen with adenomatous hyperplasia (1), spindle (metaplastic) component (8) and also focal sertoliform differentiation similar to that seen in benign cystadenoma of rete testis. Intratubular invasion of the testis can occur. Primary adenocarcinoma of rete testis should be differentiated from three main entities namely, primary adenocarcinoma of epididymis, malignant mesothelioma of tunica vaginalis and metastatic germ cell tumor. Primary adenocarcinoma of epididymis is recognised by tubular, tubulocystic or tubulopapillary growth lined by cuboidal or columnar, predominantly clear cells with invasion and necrosis and immunonegativity for CEA. Cells of malignant mesothelioma of tunica vaginalis are cuboidal with oval vesicular nuclei and eosinophilic cytoplasm and PAS negative and they also show immunonegativity for CEA. In most cases, distinction from metastatic germ cell tumor cannot be done by histology and those tumors show immunopositivity with AFP, hCG and

PLAP. All these features were absent in our case. Nochomovitz and Orenstein (9) laid down criteria for diagnosing primary adenocarcinoma of rete testis. They are : (i) absence of histologically similar tumor at other sites; (ii) tumor centered in the region of hilus; (iii) morphology incompatible with any other type of testicular or paratesticular tumor; (iv) a transition from unaffected rete testis to tumor; (v) a predominantly solid appearance with tubular, papillary and cribriform growth patterns. All these features were present in our case except the first point as the tumor was occupying the almost the whole of testis. Moreover, tumor cells in the present case were PAS positive and showed immunoreactivity for CEA and immunonegativity for desmin. However no adenomatous hyperplasia or any other component was seen in our case.

Present method of treatment is radical orchiectomy, which is relatively safe and complications are rare (10). Some authors claim that retroperitoneal lymph node dissection may increase longevity of the patient while others don't agree to it. Chemotherapy and radiotherapy don't have any role in this tumor as per some studies (2) while others have tried chemotherapy with some success (1).

In our case, tumor size was 4.5x4x2.5 cm and radical orchiectomy was done. Block dissection of ipsilateral inguinal lymph nodes was done after appearance of metastasis, but no adjuvant therapy was given.

The prognosis is controversial as the number of reported cases is very less in number. The outcome is usually poor, with approximately 13% survival at 5 years (4). But according to a study by Glazier et al, the tumor seems to have a varied prognosis with aggressive tumor being reported in some cases while in others survival was up to seven years (11).



In conclusion; primary adenocarcinoma of rete testis is an uncommon and aggressive neoplasm. It should be confirmed by defined diagnostic criteria. Cytological diagnosis is also crucial for early planning of management. Immunohistochemistry has a very important role to confirm the diagnosis. Though current recommendation is to treat these cases with radical surgery, more controlled clinical trial is needed to prove the utility of chemotherapeutic drugs in this tumor.

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