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Case Report

# Adenocarcinoma of the Rete Testis: Interest of FDG PET/CT and Aggressive Surgical Management: A Case Report

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## Abstract

Primary adenocarcinoma of the rete testis is a rare testicular tumor, classically associated with a poor prognosis. Due to its rarity, therapeutic management is not yet clearly defined. We report a case of primary adenocarcinoma of the rete testis in a 42-year-old patient who initially presented a right testicular mass. He underwent a radical orchiectomy in 2003. Since then, regular follow-up by 2-deoxy-2(F18) Fluoro-D-Glucose (FDG) Positron Emission Tomography combined with Computed Tomography (PET/CT) revealed multi focal lymph node relapses for which multiple lymph node dissections (retroperitoneal, abdominal, cervical and mediastinal) were performed. Seventeen years after the initial diagnosis, the patient is alive and has not yet presented any visceral or bone metastasis. To our knowledge, this is the longest follow-up reported in the literature. FDG PET/CT follow-up associated with an aggressive surgical management of resectable masses should be considered given the lack of evidence of effective systemic treatments and radiotherapy.

**Keywords: Adenocarcinoma; Rete testis; Testicle tumor; FDG PET/CT**

## Abbreviations & Acronyms

FDG: 2-deoxy-2(F18) Fluoro-D-Glucose; PET/CT: Positron Emission Tomography combined with Computed Tomography; β-HCG: Beta Human Chorionic Gonadotropin; PSA: Prostatic Specific Antigen; CEA: Carcino Embriologic Antigen; CT: Computer Tomography; EMA: Epithelial Membrane Antigen; NSE: Neuron Specific Enolase; PLAP: Placental Alkaline Phosphatase; CA19-9: Carbohydrate Antigen 19-9; OCT3/4: Octamer-Binding Transcription factor ¾; SALL4: Sal-Like Protein 4; RPLND: Retroperitoneal Lymph Node Dissection

## Introduction

Adenocarcinoma of the rete testis is a rare testicular tumor that is part of non-germinal testicular tumors [1]. The prognosis is poor with a median survival time of 33 months and a disease-free survival of 49% at 3 years and 13% at 5 years [2-9]. Until now, approximately 70 cases have been described in the literature. Due to its rarity, diagnosis is often difficult and management is still poorly defined. Radical inguinal orchiectomy is the standard initial treatment. Management after orchiectomy remains controversial because chemotherapy and external radiotherapy have only shown a low response rate [2,4,9]. Some authors suggest to perform Retroperitoneal Lymph Node Dissection (RPLND) even in the absence of radiological metastasis due to the risk of occult metastasis [5]. We present a case of adenocarcinoma of the rete testis initially treated by radical orchiectomy and then followed regularly by FGD PET/CT. This exam detected on several occasions the presence of small lymph node metastases which were treated by multiple lymph node dissections. Seventeen years after the initial diagnosis, our patient is alive and has not yet presented any visceral or bone metastasis. This clinical situation illustrates the advantage of FDG PET/CT in the early detection of metastases. Based on ourcase, the literature and the low level of efficiency of systemic treatments and radiotherapy, aggressive surgical treatment of resectable metastases should be considered inappropriate candidates.

## Case Presentation

At the time of the diagnosis, the patient was 42 years old. For several months, he presented an indurated, non-mobile and painless mass in the right testicle. There was no associated local inflammatory sign. The scrotal ultrasound showed a poorly circumscribed hypervascularized formation of 18 mm in the upper pole of the right testis. The specific tumor markers, Beta Human Chorionic Gonadotropin (β-HCG) and α-fetoprotein, and the Prostatic Specific Antigen(PSA)were normal preoperatively. Only Carcino Embriologic Antigen(CEA) was increased. Thoraco-abdomino-pelvic Computer Tomography (CT) scan did not show any distant metastasis. A radical inguinal orchiectomy was performed. Histopathological and immunohistochemical analysis supported primary adenocarcinoma of the rete testis with complete resection. Indeed, histologically, the tumor was centered on the testicular hilum and there was transition from normal rete testis epithelium to dysplastic epithelium (Figure1). The tumor showed a large sheets and solid nests of pleomorphic cells. Central necrosis was observed (Figure 2). Furthermore, cross-sectional imaging did not identify any primary site which implies the existence

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of other metastatic adenocarcinoma. Immunohistochemical analysis showed positivity of tumor cells for keratin, Epithelial Membrane Antigen (EMA) and CEA and negativity for cytokeratin 39 and inhibin. Dosage of CEA was normal after the surgery. No adjuvant therapy was administered to the patient. Every six-months, dosage of β-HCG, α-fetoprotein and CEA was performed and a thoraco-abdomino-pelvic CT scan was done. Four years after the initial diagnosis, CT scan revealed several infracentimetric lumbo-aortic lymph nodes. The tumors markers were all negative. A complementary assessment by a FDG PET/CT revealed the presence of an abnormal hypermetabolic activity in front of these lymph nodes, highly suggestive of a tumor relapse. Lumbo-aortic lymph node dissection was performed. Three out of 15 lymph nodes were invaded by adenocarcinoma of the rete testis. The immunohistochemical analysis showed positivity of tumor cells for cytokeratinAE1/AE3 and focally forNeuron Specific Enolase (NSE) and negativity for Placental Alkaline Phosphatase (PLAP), β-HCG, α-fetoprotein, inhibin-α and calretinin. The FDG PET/CT was then proposed every 6 months with the aim to detect early recurrence. Repeatedly, this exam detected infracentimetric but hypermetabolic lymphadenopathies (Figure 3) always confirmed to be metastases of the adenocarcinoma of the rete testis after resection and histopathological analysis. Adjuvant treatment with external radiotherapy was administered twice after lymph node dissection. The patient never received adjuvant chemotherapy. Currently, after numerous surgeries, he never developed any visceral or bone metastasis (Table 1).

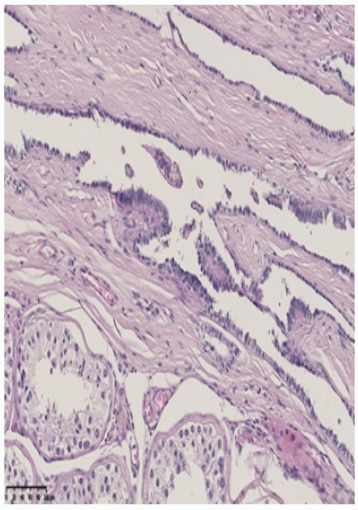

**Figure 1**: Histological analysis showed the transition from normal rete testis epithelium to dysplastic epithelium.

## Discussion

Primary adenocarcinoma of the rete testis is a rare testicular tumor with a poor prognosis [1]. Although cases have been described in men aged between 17 and 91, it occurs more frequently over 50 years with a peak of incidence around 70 years [2]. Clinically, the patient most often presents a painless testicular mass which can be accompanied by local signs (hydrocele, epididymitis, hematocele, etc.). The initial clinical presentation may be nonspecific since is a consequence of the presence of metastasis (lumbar pain, abdominal mass, groin mass, lymphangitis…). Diagnosis could therefore be delayed due to an atypical clinical presentation or symptoms suggesting an inflammatory process [2,3]. In some cases, there is a history of cryptorchidism or testicular trauma. The testicular tumor markers (β-HCG and α-fetoprotein) are normal. Several cases of adenocarcinoma of the rete testis with elevation of CEA and Carbohydrate Antigen 19-9 (CA19-9) have been reported in the literature, but actually there are no valid laboratory markers [6,7]. Given the rarity of this tumor, the

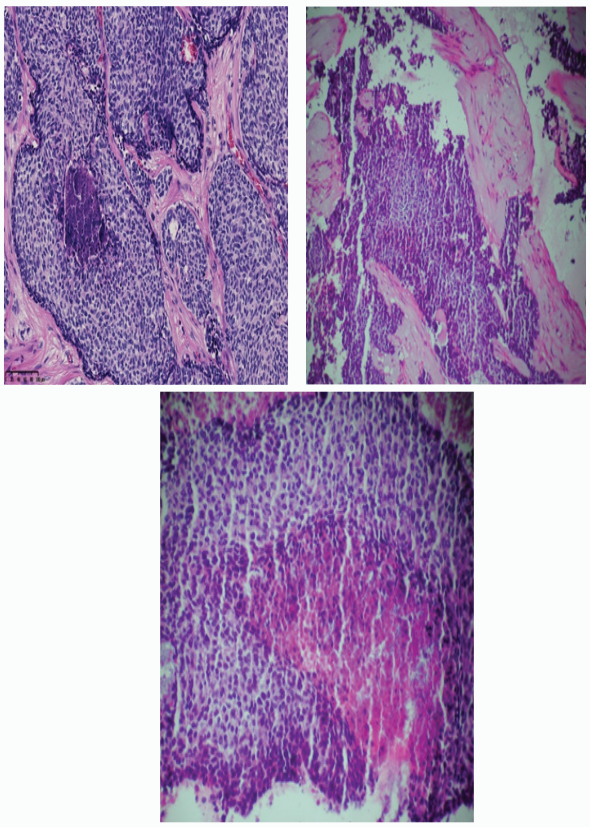

**Figure 2**: Histological analysis showed solid nests of pleomorphic tumoral cells with central necrosis.

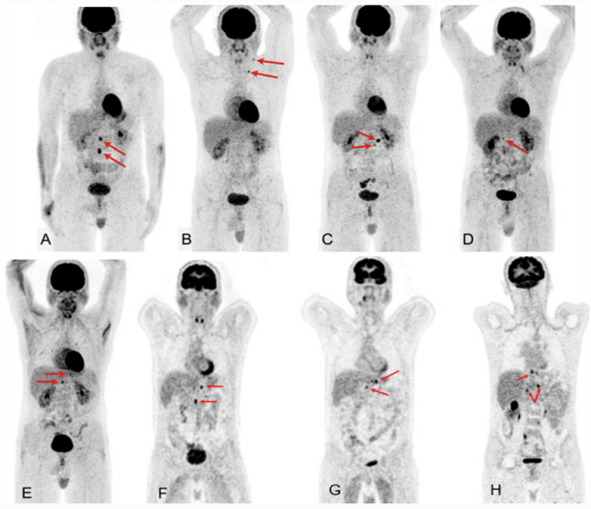


**Figure 3**: FDG PET/CT showing: (A) 2007: Lombo Aortic lymph nodes; (B) 2008: Cervical lymph nodes; (C) 2009: Retroperitoneal lymph nodes, near left renal vein; (D) 2010: Retroperitoneal lymph nodes; (E) 2012: Inter Aortico Caval And lesser gastric curvature lymph nodes; (F) 2016: Interaortocaval lymph nodes; (G) 2018: Cœliac trunk and lesser gastric curvature lymph nodes;(H) 2020: Coeliac trunk, hepatic hilus and mediastinal lymph nodes.
Table 1: Summary of localization, histological finding and adjuvant therapy of lymph node metastasis.

**Table 1:** Summary of localization, histological finding and adjuvant therapy of lymph node metastasis.

| Years | Lymph nodes localization on FDG PET/CT | Number of lymph nodes objectified on FDG PET/CT | Number of lymph nodes removed | Histological finding of lymph nodes | Adjuvant treatment after lymph node dissection |
|---|---|---|---|---|---|
| 2007 | Retroperitoneal: Lumbo-aortic | 2 | 15 | 3 lymph nodes invaded including 2 with capsular breakage | / |
| 2008 | Cervical | 2 | 14 | 5 lymph nodes invaded including 2 with capsular invasion | External radiotherapy |
| 2009 | Retroperitoneal: near renal vein | 2 | 25 | 5 lymph nodes invaded including 1 with capsular invasion | / |
| 2010 | Retroperitoneal: interaorticocaval | 1 | 5 | 5 lymph nodes invaded including 3 with capsular breakage | / |
| 2011 | Retroperitoneal and Mediastinal | 3 | 20 | 7 lymph nodes invaded | / |
| 2012 | Interaorticocaval + lesser gastric curvature | 3 | 5 | 4 lymph nodes invaded including 1 with capsular breakage | / |
| 2016 | Retroperitoneal: Interaorticocaval | 3 | 10 | 2lymph nodes invaded with capsular breakage and incomplete resection | External radiotherapy |
| 2018 | Cœliac trunk and small gastric curvature | 10 | 12 | 12 lymph nodes invaded | / |
| 2020 | Mediastinal | 3 | 10 | 2 lymph nodes invaded including 2 with capsular breakage | / |
| 2020 | Coeliac trunk and hepatic hilus | 2 | 4 | 2 lymph nodes invaded including 2 with capsular breakage | / |

anatomopathological and immunohistochemical characteristics are not yet well defined, which makes the diagnosis difficult and makes it most often an exclusion diagnosis. The challenge is to differentiate it from mesothelioma, sex cord-stromal tumor, germ cell tumor of the testis or any other metastatic extra-testicular adenocarcinoma. Five pathological criteria were proposed by Nochomovitz and Orenstein to identify adenocarcinoma of the rete testis: (1) a tumor centered on the testicular hilus, (2) proof of a transition between the normal rete testis and the rete testis invaded by the tumor, (3) the absence of invasion of the testicular vaginal, (4) the absence of a histologically similar extra-scrotal tumor and (5) a morphological appearance incompatible with any other testicular or para-testicular tumor [3]. Our case showed all these anatomopathological criteria. Immunohistochemical analysis is useful to exclude other metastatic adenocarcinoma, mesothelioma, sex cord-stromal tumor, germ cell tumor of the testis. Those expose variable degrees of positivity for cytokeratin (AE1/AE3, 7, etc.), CEA, EMA, vimentin, etc. In contrast, these analysis showed negativity for Octamer-binding Transcription factor ¾ (OCT3/4), Sal-Like Protein 4 (SALL4), glypican3, β-HCG, α-fetoprotein, PLAP, inhibin, calretinin, CK5/6, etc [8,9]. Adenocarcinoma of the rete testis has a poor prognosis. Sánchez-Chapado et al. [2] reported a disease-free survival of 49% at 3 years and 13% at 5 years. They also showed there is two predictors of survival: the size of the primary tumor (tumors smaller than 5cm have a better overall survival) and tumor confinement to the testis. Unfortunately, this aggressive tumor had a high potential for dissemination. Patients are often metastatic at the diagnosis or the metastases occur early during the course of the disease in lymph nodes, lungs, bones and liver. Thoraco-abdomino-pelvic CT scan may be useful for the detection of metastases. However, Musser et al. [10] illustrated the potential benefit of the FDG PET/CT in the early detection of metastases. Indeed, they reported a delayed detection of 3 months of suspicious lymphadenopathy with conventional imaging compared to FDG PET/CT. In our case, we also note that the FDG PET/CT allowed the early detection of metastatic lymphadenopathy however not suspicious on conventional imaging. Inguinal radical orchiectomy is the standard initial treatment of testicular tumor. Management of the adenocarcinoma of the rete testis after orchiectomy remains controversial because chemotherapy and external radiotherapy have only shown a low response rate in both localized and metastatic disease [2,7,9]. The benefit of prophylactic RPLND is debated because of the low number of reported cases. However, recently, some authors suggested RPLND after orchiectomy, even in absence of radiological metastasis, due to the risk of occult metastases [5,11]. Sánchez-Chapado and al. performed a meta-analysis of 40 patients and found that patients who underwent RPLND had an higher 3-year overall survival than those who did not (83% versus 42%, log rank P=0,034) [2]. But there is a selection bias, surgical treatment was performed mostly in patients with lower disease burden. In our patient, the aggressive surgical management of disease recurrence may improve the evolution of the disease. In appropriate candidates, adjuvant RPLND could be considered as a useful therapeutic modality such as surgical treatment of resectable metastases.

## Conclusion

Adenocarcinoma of rete testis is a rare tumor. The diagnosis is difficult and the therapeutic management is not yet well established. In absence of valid biomarkers, imaging is essential to detect metastases and disease recurrence. Close follow-up by FDG PET/CT seems to be the best modality to improve the early detection of metastases. Surgical management of resectable metastases is a valid therapeutic strategy because of the low response rate to systemic treatment and radiotherapy.

## References

1. Tian Y, Yao W, Yang L, Wang J, Wazir R, Wang K. Primary adenocarcinoma of the rete testis: A case report and review of the literature. Oncol Lett. 2014;7(2):455-7.

2. Sánchez-Chapado M, Angulo JC, Haas BP. Adenocarcinoma of the rete testis. Urology. 1995;46(4):468-75.

3. Nochomovitz LE, Orenstein JM. Adenocarcinoma of the rete testis: case report, ultrastructural observations, and clinicopathologic correlates. Am J Surg Pathol. 1984;8:625-34.

4. Cullen MH, Stenning SP, Parkinson MC, Fossa SD, Kaye SB, Horwich AH, et al. Short-course adjuvant chemotherapy in high-risk stage I nonseminomatous germ cell tumors of testis: a Medical Research Council report. J Clin Oncol. 1996;14(4):1106-13.

5. Maganty A, Fombona A, Bandari J, Lyon TD, Kulich S, Gingrich JR, et al. Aggressive surgical management of adenocarcinoma of the rete testis. Urol Case Rep. 2018;16:72-4.

6. Takeda S, Yamashita S, Kawamorita N, Mitsuzuka K, Fujishima F, Watanabe M et al. Primary adenocarcinoma of the rete testis with elevation of serum carcinoembryonic antigen: a case report. Int Cancer Conf J. 2017;6(2):84-7.

7. Kitano H, Sentani K, Goto K, Sekino Y, Yamanaka R, Nagasaka K et al. Primary adenocarcinoma of the rete testis with elevated serum CA19-9 antigen levels. Int Cancer Conf J. 2020;9(4):240-3.

8. Ballotta MR, Borghi L, Barucchello G. Adenocarcinoma of the rete testis. Report of two cases. Adv Clin Path. 2000;4(4):169-73.

9. Al-Obaidy KI, Idrees MT, Grignon DJ, Ulbright TM. Adenocarcinoma of the Rete Testis: Clinicopathologic and Immunohistochemical Characterization of 6 Cases and Review of the Literature. Am J Surg Pathol. 2019;43(5):670-81.

10. Musser JE, Ernest AJ, Thibault GP, McMann LP. Primary adenocarcinoma of the rete testis: improved staging accuracy with CT-PET. Urology. 2011;77:334.

11. Chovanec M, Mego M, Sycova-Mila Z, Obertova J, Rajec J, Palacka P, et al. Adenocarcinoma of the rete testis - A rare case of testicular malignancy. Klin Onkol. 2014;27:136-7.