CLINICAL VIGNETTE

Paratesticular Serous Papillary Carcinoma

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A 36-year-old male presented with a history of a left testicular mass. The mass had been noticed years ago and was determined to be a hydrocele based on exam and serial ultrasounds. There was never any change until about two years ago when he noted growth of the mass. Repeat imaging and follow up still suggested a hydrocele. However, over the present year the patient became more concerned and opted to proceed with a left orchiectomy. Pathology demonstrated a 6.6-centimeter, high grade serous papillary carcinoma located in the upper pole of the testis. The tumor invaded into the hilar soft tissue and scrotal wall, but the epididymis and testicular parenchyma were unremarkable for malignant involvement. Immunohistochemistry ruled out other etiologies such as lymphoma and melanoma. Interestingly, the tumor was negative for alpha-fetoprotein, calretinin, CK5/6, D240, glypican-3, inhibin-A, and SALL-4 and was positive for p16, estrogen receptor, CA125, WT-1, and PAX-8. In whole, the findings were consistent with a paratesticular papillary serous carcinoma. Computed Tomography (CT) of the chest demonstrated no metastatic disease. CT of the abdomen and pelvis showed post-operative changes and likely liver cysts and pelvic bone islands. His basic labs were unremarkable.

He was otherwise healthy with no other medical problems or prior surgeries. His father had a history of early stage prostate cancer at 60 years old. He was married with one 2-year-old daughter. He quit smoking a year prior to consultation, but he never engaged in heavy alcohol or illicit drug use.

He was asymptomatic other than low back pain that he attributed to the surgery. It waxed and waned but was improving with time and did not require any analgesics. His vital signs and exam were unremarkable.

Germ cell tumors are the most common malignancy when cancer is discovered in the testicle.[1] Ovarian type epithelial carcinomas of the testicle are not common with only case reports noted in the literature.[2,3] When present, serous papillary borderline tumors are the most common pathology, but invasive mucinous, endometrioid, and clear cell types have been documented.[2] Presentations are similar to that presented here with a new scrotal mass or fullness commonly noted at presentation.[1] Very frequently a hydrocele is associated with the disease.[1,2,3] The age of presentation noted here is typical for diagnosis as the mean age for invasive cancer is 31 years old, although older (56 years old) for borderline tumors.[1,2,3] These tumors can arise in the testis or more commonly in the paratesticular tissue.[2,3] More specifically, they involve the visceral tunica vaginalis or, as with our patient, involve the upper pole of the testis in the testiculoepididymal area.[2] The higher grade of this patient's disease was a bit unusual as grade 1-2 disease is usually seen, although an aggressive clinical course is not unusual.[2] In one series of six patients, two patients were later found to have peritoneal or metastatic disease.[2]

The exact cell of origin for development is not exactly known, but it is hypothesized that it may be related to Mullerian duct remnants (for example, the appendix testis and obliterated remnants of the connective tissue around the epididymis or spermatic cord).[1,2] Support for the appendix testis includes the fact that a large number of these tumors are seen in the upper pole of the testis.[2] Another theory is that the cells from the mesodermal epithelium maintain the capacity for differentiation when malignant changes occur.[1] Thus, some propose that these serous tumors derive from structures such as the tunica vaginalis or tunica albuginea.[1,2]

Given the rarity of this tumor type, it is imperative to rule out other rare cancers such as mesothelioma and metastatic carcinomas.[1,2] Generally, immunohistochemistry is used in this regard but, given overlap with other rare tumors and the rarity of this tumor type, results can be difficult to interpret, requiring experienced pathology assessment.[1] While labs and imaging can help support the diagnosis and guide surgery, there are no specific findings to suggest the diagnosis.[1] CA125 tumor marker can be elevated in some patients as noted in ovarian cancer patients.[2] Pathology is the most crucial feature to make the diagnosis.[1,2]

There are no data on the best treatment approach. It is fairly consistent throughout the various case reports that radical orchiectomy is performed.[1,2,3] Many of these patients do very well long-term with no relapse of disease.[1,2] However, borderline tumors and invasive tumors are often grouped together, making determination of outcomes harder to interpret. There are no guidelines for the role of adjuvant therapy in invasive disease.[1] Some reports state that these tumors are resistant to chemotherapy and radiation.[1] Most of the literature recommends treating them akin to ovarian cancer given the similar pathology.[1] In the case above, given his young age and high grade disease, adjuvant platinum-based therapy was recommended with six cycles of carboplatin and Taxol.[1] The patient opted to proceed and remained in the midst of treatment upon completion of this report.

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