Adenocarcinoma of the testicular adnexa presenting with metastatic disease

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JOOS ZP, PRITCHETT TR, PORTER CR, ISACSON C, PICOZZI V, CORMAN JM. Adenocarcinoma of the testicular adnexa presenting with metastatic disease. The Canadian Journal of Urology. 2006;13(1):2990-2992.

Primary epididymal adenocarcinoma is a rare malignancy with fewer than 30 documented cases. We report a case of a 57-year-old patient with a paratesticular mass in the presence of retroperitoneal metastatic disease. Histology confirmed the presence of primary paratesticular adenocarcinoma. In this report we review the index case, the pertinent literature and discuss adjuvant therapy.

Key Words: paratesticular neoplasm, epididymis, adenocarcinoma

Case report

The patient is a 57-year-old Caucasian male who noted a gradually increasing painful lump in or around his right testicle 12 months prior to presentation. A scrotal ultrasound demonstrated the right epididymis to be diffusely larger than the left with an isoechoic focal area situated in the epididymal tail (1.3 cm x 0.8 cm x 0.8 cm). The patient underwent inguinal exploration and radical orchiectomy.

On exploration a paratesticular mass was encountered. The tumor was a moderately differentiated adenocarcinoma, whose epicenter was the epididymis. The tumor was accompanied by abundant fibrosis and

Accepted for publication January 2006

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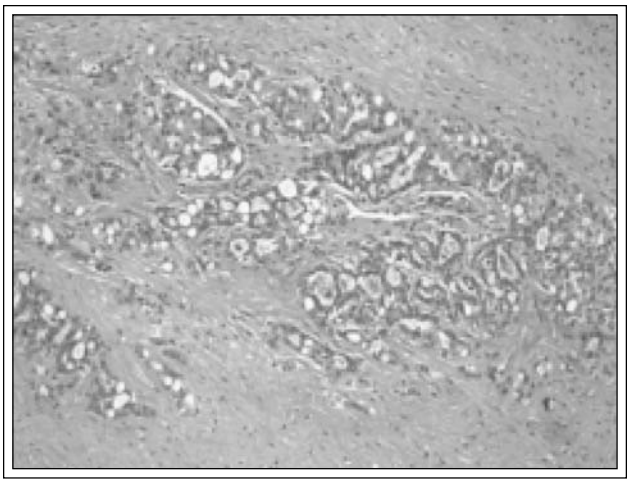

Figure 1. 100x H+E stain. Epididymal mass showing poorly differentiated adenocarcinoma.

focal ischemic necrosis, Figure 1. The tumor infiltrated between the seminiferous tubules in the hilum, extending from the epididymis at this junction. Immunohistochemical staining was performed. The tumor cells were cytokeratin (CK) 7 and CK20 strongly positive and negative for all other antigens. Basement membrane stain with collagen type IV in the epididymis/rete junction shows that the tumor nests did not have definite basal lamina. There was no evidence that the tumor contained markers specific for prostate, lung, thyroid or GI tract. Rigorous clincal evaluation revealed no evidence of an alternative site of primary malignancy. Final evaluation confirmed the tumor as a primary adenocarcinoma of the epididymis.

Initial staging studies included a PET/CT which demonstrated a single 15-mm periaortic lymph node with an SUV of 7. The patient elected to undergo a retroperitoneal lymph node dissection 10 weeks following the PET/CT scan, revealing metastatic adenocarcinoma involving intra-aortocaval, para-aortic and left renal hilar disease with extranodal extension. Repeat PET/ CT 4 weeks later demonstrated interval progression of disease with new pulmonary and nodal metastases throughout the chest, abdomen, and pelvis. In light of widely disseminated disease, the patient began a course of cisplatinum, 5- FU.

Comment

Since 1924, 24 cases of primary epididymal carcinoma have been reported.[1,2] While metastatic disease to the epididymis from prostate, lymphatic, pulmonary or biliary primaries is more commonly noted,[3-7] primary epididymal neoplasms are typically benign.[3] Large reviews suggest that fewer than 25% of epididymal neoplasms represent malignancies. The most common lesion is benign adenomatoid tumors.[4] Other benign tumors of the epididymis include leiomyoma, papillary cystadenoma and lipoma. Reported histologic variants of primary paratesticular carcinoma include: adenocarcinoma, mucinous carcinoma and squamous cell carcinoma.[3] Other primary malignancies include leiomyosarcoma, teratoma, embryonal carcinoma, rhabdomyosarcoma, fibrosarcoma and lymphoma.[2]

Primary adenocarcinoma of the epididymis may present as either a painful or painless paratesticular mass, often encompassed by a hydrocele. Scrotal involvement has not been reported. There is no reported propensity for left versus right sidedness. Paratesticular adenocarcinoma has been reported in patients 22 to 82 years old.[2] Metastatic epididymal carcinoma has only been reported in men with right-sided lesions. Environmental exposures linked to the disease include: asbestos and radiation exposures.[1,7] There is no known hereditary, genetic or family linkage to the disease, nor has the tumor been linked to neoplasmic syndromes.

Histologically, typical tumors have either tubular or tubulopapillary features lined by clear cells. Some paratesticular adenocarcinomas are, however, more cystic with papillary projections lined with columnar cells.[8] Differentiating adenomatoid tumor from adenocarcinomas may be challenging in that both display characteristic signet ring cells. Unlike epididymal carcinomas, adenomatoid tumors have little mitotic activity.[2] In order to correctly diagnose a tumor as a paratesticular primary, one must exclude the possibility of a metastasis. Immunohistochemical stains for TTF1, PSA and thyroglobulin must be conducted in order to assess the nature of the tumor. Most scrotal mesotheliomas are epithelioid; therefore, stains for calretinin, WT-1 and CK5/6 can be performed to rule out mesothelioma. With further staining for B72.3, CK7, CK20 and CEA one can conclude that the tumor is adenocarcinoma.

Of the 24 cases in the literature, 12 patients have ultimately developed metastatic disease. Of these, 6 had metastatic disease on presentation. Metastases have involved retroperitoneal lymphatics, pulmonary metastasis, and in one case, tumor thrombus.[1-3,5-7] Yamamoto et al reported that in post-mortem examinations, metastases were observed along the course of the inguinal and pelvic lymphatics with or without invasion into adjacent organs.[3]

Both trans-scrotal as well as inguinal approaches

have been advocated for paratesticular masses. Because patients greater than 50 years old are likely to have benign lesions, Beccia et al[4] suggests that scrotal exploration is sufficient. Because of the increased likelihood of malignancy in patients 20 to 50 years, inguinal exploration is conducted. Ganem suggests that all solid epididymal masses be explored inguinally especially those that have recently changed in character.[2]

The present case represents the 6th report of paratesticular adenocarcinoma presenting with metastatic disease. Five of six patients underwent orchiectomy shortly after presentation. Of the five previous patients, two are reported NED up to 30 years following therapy. Of these patients, both presented with retroperitoneal lymphadenopathy. One patient was successfully treated with a retroperitoneal lymph node dissection; the other received cis-platin based chemotherapy. Because of the scarce reports of this disease, ideal adjuvant therapy is unclear. For our index case, multidisciplinary mangement was employed, with the decision to treat the patient in a manner similar to a primary testicular neoplasm. Historically, the most commonly employed chemotherapy agent has been carboplatin with or without paclitaxel. In one case, gemcitabine salvage chemotherapy was initiated, but did not prove effective.

Given the rarity of primary paratesticular adenocarcinoma, ideal therapy is still unknown. Because these neoplasms have a high mortality rate, adjuvant radiation and chemotherapy are strongly advised. Since this tumor is easily mistaken for less aggressive disease, a high index of suspicion is required. □

References

1. Chauhan RD, Gingrich JR, Eltorky M, Steiner MS. The natural Progression of Adenocarcinoma of the Epididymis. *J Urol* 2001;166:608-610.
2. Ganem JP, Jhaveri FM, Marroum MC. Primary adenocarcinoma of the epididymis: case report and review of the literature. *Urol* 1998;52:904-908.
3. Yamamoto M, Myake K, Mitsuya H et al. A case of primary carcinoma of the epididymis. *Acta Urol Jpn* 1987;33:1139-1141.
4. Beccia DJ, Krane RJ, Olsson CA. Clinical management of non-testicular intrascrotal tumors. *J Urol* 1976;116:476-479.
5. Jones MA, Young RH, Scully RE. Adenocarcinoma of the Epididymis: a report of four cases and review of the literature. *Am J Surg Pathol* 1997;21:1474-1480.
6. Lioe TF, Biggart JD. Tumours of the Spermatic Cord and Paratesticular Tissue: a clinicopathological study. *Br J Urol* 1993;71:600-606.
7. Gisser SD, Nayak S, Kaneko M, Tchertkoff V. Adenocarcinoma of the Rete Testis: a review of the literature and presentation of a case with associated asbestosis. *Hum Pathol* 1977;8:219-224.
8. Ulbright TM, Amin MB, Young RH. Tumors of the testis, adnexa, spermatic cord and scrotum. In: atlas of tumor pathology. Fasc 25. 3rd series. Washington, DC: Armed Forces Institute of Pathology, 1999.
9. Chia-Cheng Y, Hong-Khing H, Hung C, Ming Tsun C, Chang LS. Papillary Cystadenocarcinoma of the Epididymis: a case report and review or the literature. *J Urol* 1992;147:162-165.
10. Kher MM, Kherdekar MS, Grover S, Sharma KD. Carcinoma of Epididymis: report of a case. *Ind J Cancer* 1973;10:475-478.
11. Arisan S, Akbulut, ON, Cakir OO, Ergenekon E. Primary Adenocarcinoma of the Epididymis: case report. *Int Urol Nephrol* 2004;36:77-80.