Urology Case Reports 21 (2018) 116–118

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Oncology

Primary angiosarcoma of the testis with retroperitoneal metastasis

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Introduction

While uncommon, angiosarcomas predominately occur in superficial soft tissue and rarely present in the viscera. While also rare in the general population, testicular germ cell tumors (GCT) are the most common malignancy diagnosed in males 15–44 years old.[1] Primary angiosarcoma of the testis is extremely rare with the first published report by Hughes et al., in 1991[2] subsequently followed by sporadic case reports of primary or dedifferentiated testicular angiosarcoma.[3] Interestingly, Idress and colleagues published evidence suggesting that metastatic angiosarcoma may arise secondary to clonal progression following systemic treatment of mixed type GCT.[4] As it remains exceedingly rare, there is a paucity of data on the proper management of testicular angiosarcoma.

Case presentation

Our patient was a 58-year-old male without a significant past medical history. He presented to his primary care provider with a 3-month history of bilateral hip and low back pain. Eventually an MRI of his spine was performed, demonstrating a questionable lesion at the L5 level. A follow up CT revealed a large necrotic retroperitoneal mass causing right hydronephrosis and compression of the IVC [Fig. 1]. Urology was consulted and physical exam was significant for a large, bilobar, right testicular mass. Scrotal ultrasound confirmed a testicular mass with an adjacent scrotal mass and normal left testis.

Retrograde right ureteral stenting was attempted to relieve the right hydronephrosis. However, a stent was unable to be passed beyond the level of obstruction. Subsequently, a right nephrostomy tube was placed with improvement in the patient's renal function.

The patient underwent a right inguinal radical orchiectomy. Gross pathology demonstrated a 360 g testis with a 12 × 6.8 × 6.8 cm testicular tumor and a separate 3.5 × 2.4 × 2 cm tumor of the spermatic cord. Histologic evaluation of the specimen revealed angiosarcoma invading the testis, epididymis and spermatic cord with negative margins [Fig. 2]. Pathology demonstrated atypical tumor cells with elongated nuclei in a background of lymphocytes and histiocytes. Numerous stains were utilized to elucidate the pathologic diagnosis. Significantly, the specimen was negative for OCT3/4 and AFP staining suggesting no GCT component. The tumor was diffusely positive for CD10, CD31, and vimentin, and was negative for cytokeratin AE1/3, MDM-2, desmin and myoD1.

Given the presumed diagnosis of metastatic angiosarcoma, the patient was referred to Oncology for systemic treatment. Staging FDG PET/CT demonstrated avid uptake in the right pelvic lymph nodes, lung lesions, retroperitoneal mass, and numerous bone lesions [Fig. 3]. Biopsy of a bone lesion confirmed metastatic angiosarcoma with a final stage of pT3N1M1. The patient elected to pursue palliative chemotherapy and succumbed to his disease 6 months after initial presentation.

Discussion

Since the initial description in 1991[2], there have been scarce reports of primary testicular angiosarcoma. A review of the 5 reported cases was published in 2007 by Armah and colleagues.[3] They observed that patients under 30 with testicular angiosarcoma presented concurrently with pathological findings of germ cell teratoma. Patients older than 60 did not possess GCT components and pathologically appeared to exclusively contain elements of angiosarcoma.[3] It has been postulated that testicular angiosarcomas arise following a transformation from an original GCT. This transformation may occur *de novo* or following treatment with radio or chemotherapy.[3,5] Interestingly, Indress et al. demonstrated clonal progression from a mixed germ cell tumor in a 38 year old after 4 cycles of chemotherapy with the original germ cell tumor and the subsequent angiosarcoma demonstrated identical loss of heterozygosity within microsatellite markers.[4]

Our patient presented without a prior history of germ cell tumor nor prior chemotherapy or radiation. While the surgical margins were negative, the tumor was invasive into the epididymis and spermatic cord and found to be widely metastatic on subsequent imaging. There were no signs of a germ cell tumor in the orchiectomy specimen which appeared to exclusively contain invasive angiosarcoma. Significantly, the orchiectomy specimen was negative for OCT3/4 and AFP staining

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https://doi.org/10.1016/j.eucr.2018.09.014
Received 2 August 2018; Accepted 17 September 2018
Available online 19 September 2018

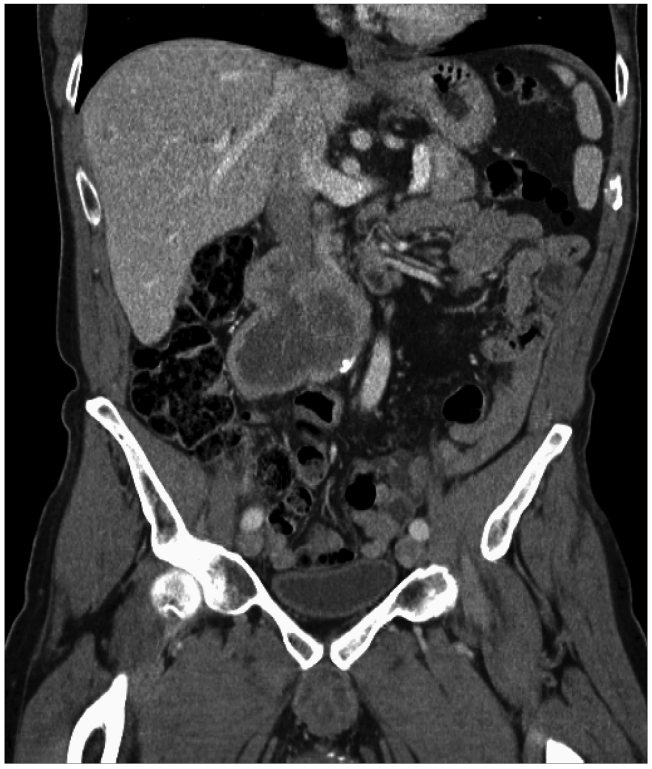

Fig. 1. Preoperative coronal CT scan demonstrating right retroperitoneal mass with encasement of right common iliac vessels and inferior vena cava.

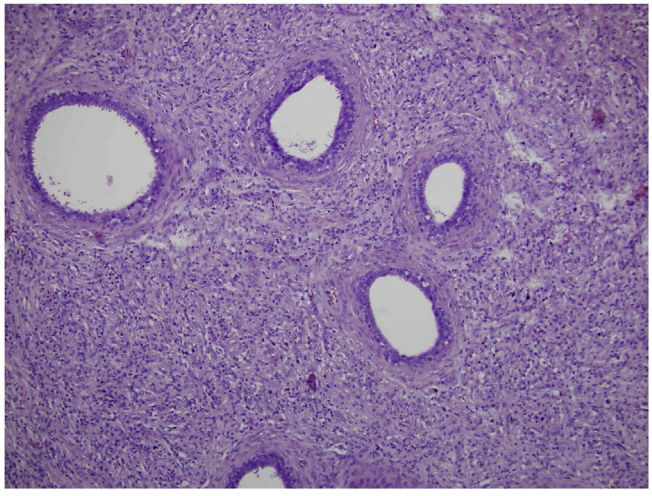

Fig. 2. Hematoxylin and eosin stain of right testicular tumor demonstrating tumor engulfment of epididymis tubules.

suggesting no germ cell component. The tumor was diffusely positive for CD10, CD31, and vimentin and was negative for cytokeratin AE1/3, MDM-2, desmin and myoD1. Similar to previous reports of testicular angiosarcoma this specimen stained positive for CD31 and negative for cytokeratin and CD45.[3,5] A dedifferentiated liposarcoma was excluded by a negative MDM-2 stain, and muscle tumors were excluded by negative desmin, muscle-specific actin, and myoD1 stains. Significantly in our case, despite an extensive search, no sign of a germ cell tumor could be located in the specimen.

At the time of the initial presentation the patient reported that he had not noted his testicular mass previously and that the mass was "new". He was unable to provide a more accurate timeline and review of outside records yielded no further information. There was no history

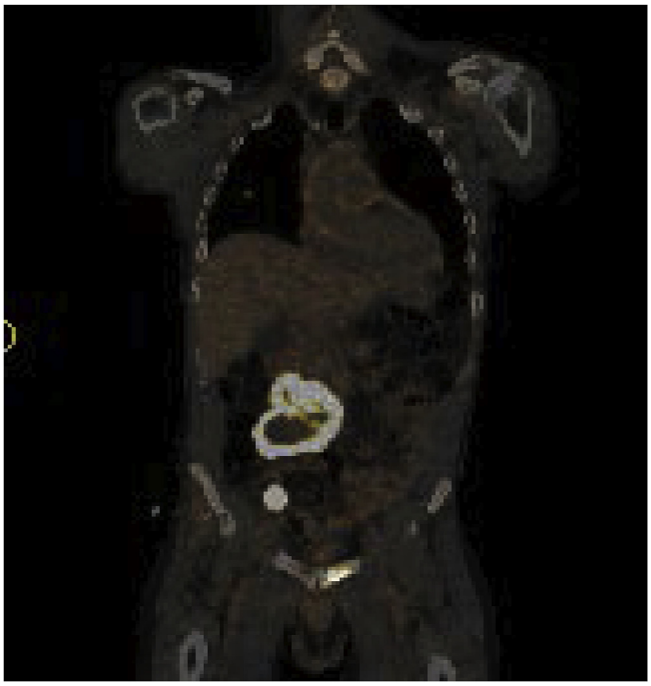

Fig. 3. Post-operative coronal PET/CT scan demonstrating avid uptake in the right pelvic lymph nodes and right retroperitoneal mass.

suggestive of chronic testicular or epididymal infection, persistent hydrocele, or other longstanding source of genitourinary inflammation. The patient denied any prior surgery, malignancy, or treatment with radiation. Thus, it is unclear if this testicular angiosarcoma was the result of progression from a long-standing germ cell tumor of the testis or a *de novo* presentation. The concordant presence of a right sided retroperitoneal mass surrounding the ureter and compressing the IVC circumstantially suggest a possible germ cell origin. However, definitive data is lacking.

Metastatic angiosarcoma carries a grim prognosis as do most metastatic soft tissue sarcomas. Localized angiosarcomas predominantly present in the lower extremities and are treated with aggressive surgical resection with or without adjuvant radiation depending on specific tumor location and final pathology. Unfortunately, in our case the patient was confirmed to be widely metastatic at the time of his presentation and was not a candidate for further surgical intervention. Consistent with his goals of care, the patient elected to undergo palliative chemotherapy with liposomal doxorubicin given the relatively low toxicity profile.

Conclusion

Angiosarcoma of the testis remains a rare diagnosis with few reports in the literature. It appears likely that certain cases of angiosarcoma of the testis may arise following a transformation of a preexisting GCT. However, the presence of a germ cell malignancy was not appreciated in our reported case of right-sided testicular angiosarcoma with associated right retroperitoneal and lung metastasis. Optimal management of testicular angiosarcoma has not been clearly defined with exact diagnosis unlikely prior to pathological evaluation of a radical inguinal orchiectomy specimen.

Consent

Obtained.

Conflict of interest statement

None.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.eucr.2018.09.014.

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