Stage III Primary Retroperitoneal Yolk Sac Tumor with Sarcomatous Components in a 40-year-old Male: A Case Report

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Abstract

Primary yolk sac tumors (YST) are rare pre-pubescent tumors. A 40-year-old with chronic testicular swelling was diagnosed with YST with pulmonary metastasis, pulmonary embolism, and left leg DVT, undergoing 4 cycles of BEP chemotherapy and left orchiectomy. This report highlights the aggressive course but adequate treatment response to adult YSTs.

Introduction

The incidence of testicular germ cell tumors has increased from 5.7 to 6.8 cases per 100,000 individuals between 1992 and 2009 [1]. Testicular yolk sac tumors are rare in nature. They are a type of non-seminomatous germ cell tumors. In adults, yolk sac tissue in germ cell tumors is usually seen in combination with other types of germ cell tumors. They usually present during the second and third decades of life. Distant disease is also more frequent in post pubertal patients than the pediatric patients [2]. These yolk sac tumors are dangerous and without treatment, lead to rapid deterioration of clinical status.

Wang et al. [3] reported in 2015 that during their literature search, they only found < 20 cases of pure yolk sac tumors in adults. On further literature search, we found 2 more case reports published after 2015 reporting a case of pure yolk sac tumor [4] [5]. We report here a case of gonadal primary adult yolk sac tumor in a 40-year-old male.

Case Summary

A 40-year-old male presented to Rochester General Hospital in October 2021 with complaints of left leg swelling and pain. He had also noticed gradual non painful swelling of this scrotum over the last year with associated weight loss. Examination showed left sided scrotal swelling and left leg swelling. Doppler of the lower extremity showed left proximal DVT for which he was started on anticoagulation. A CT Abdomen and Pelvis was also ordered because of the left scrotal swelling. It showed a 30.1 cm (cranial caudal) lobulated mixed attenuating left lower pelvic retroperitoneal mass extending to the left hemiscrotum. Several pulmonary nodules measuring up to 1.9 cm were also seen. A subsequent CT Chest showed multiple pulmonary nodules as well as an extensive pulmonary embolus. Beta HCG was < 0.6 IU/ L (< 0.5 IU/ L) although AFP was raised to 74242.6 ng/mL (< 9 nm/mL) and LDH was raised to 1692 IU/L (100-190 IU/L). He subsequently underwent a needle biopsy of the retroperitoneal mass which showed histopathology most consistent with yolk sac tumor with tumor necrosis and sarcomatous pattern. It was also positive for chromosome 12p detection. He was diagnosed with stage III poor risk tumor (due to AFP >10,000) and completed BEP chemotherapy for 4 cycles. He underwent repeat CT scans post treatment in February 2022 showing complete resolution of the pulmonary nodules with evidence of chronic pulmonary embolus as well as dramatically favorable treatment response with the left retroperitoneal mass now measuring 4 cm in the greatest dimension. His AFP post treatment had also trended down to 72 ng/mL. After interdisciplinary

discussion, the decision was made to proceed ahead with radical orchiectomy in April 2022 given his AFP had risen to 109 ng/mL on surveillance follow up. He is still being actively followed by oncology and urology.

Discussion

Testicular tumors can be classified into germ cell tumors, sex cord stromal tumors and mixed germ cell and sex cord stromal tumors. The germ cell can further be classified into tumors derived from germ cell neoplasia in situ (GCNIS) or unrelated to germ cell neoplasia in situ. The former category can be further subdivided into seminomas and non-seminomatous germ cell tumors. Primary yolk sac tumor of the post pubertal type falls into the latter category [6]. Primary yolk sac tumors in the post pubertal age group are very rare [3]. Here we presented a case of a 40-year-old male diagnosed with a primary yolk sac tumor which had spread to involve retroperitoneum and metastasized to lungs on diagnosis. The patient also had a PE and DVT at the time of initial diagnosis.

Microscopic examination of yolk sac tumors consists of primitive cells with many histological types including microcystic, papillary, solid, festoon, poly-vesicular-vitelline, glandular, intestinal, endometroid, parietal, tubular or hepatoid. Schiller Duval bodies are a pathognomonic for yolk sac tumors and it appears like a glomerulus like structure with a fibrovascular core. There is no histopathological difference in the prepubertal and post pubertal yolk sac tumors, but they behave different clinically. The post pubertal yolk sac tumors are much more aggressive than their pre pubertal counterparts. The GCNIS tumors have a common finding of amplification of the 12p chromosome which our patient was also positive for [2].

There has been some report of similar cases in the literature. Medica [7] in 2001 reported a case of adult yolk sac tumor in a 44-year-old male whose AFP was 1733 ng/ml. His disease was localized, and he was treated with surgical resection of the testicular mass with lymph node dissection which was negative for metastatic disease. Behera et al. l [4] reported a similar case to ours in a 37-year-old male who was successfully treated with inguinal orchidectomy and BEP combination chemotherapy. Their patient also had a lung nodule on presentation and after 2 months of completion of chemotherapy, the lung nodule had regressed. Their patient was still doing well with a normal AFP at 4 years follow up at the time of publication. Murcia et al. [2] reported a case of primary yolk sac tumor as well, but their patient was younger (20-year-old) and had metastasis to the liver, retroperitoneum, and lung. Unfortunately, their patient had a precipitous hospital stay and passed away within 3 months of diagnosis.

The present-day treatment for yolk sac tumors is surgery and chemotherapy followed by active surveillance with serial examinations and laboratory follow up. Serum AFP seems to be an important biomarker of disease activity in these cases. The serial measurement of AFP is beneficial in monitoring the clinical course and response to treatment [8].

Conclusion

Primary yolk sac tumors are rare in the adult population. We described a case of a 40-year-old male here who presented with left testicular swelling and was found to have a primary yolk sac tumor with extension into the left retroperitoneal space as well as bilateral pulmonary metastasis. He was treated with BEP chemotherapy with good response and has gotten orchiectomy post treatment for residual disease. This report highlights the necessity of accelerated work up required for adult post pubertal patients presenting with possible testicular tumors given the possible aggressive nature of the underlying disease.

Authorship List

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Substantial contribution to conception and design, involved in drafting the manuscript and revising it critically, given final approval of the version to be published agreed to be accountable for all aspects of the work in ensuring questions related to accuracy or integrity are appropriately investigated

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