

Yolk Sac Tumor Masquerading As Pericarditis: A Case Report

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Abstract

Introduction: Yolk sac tumor is a rare form of a germ cell tumor (GCT). This case report describes an unusual presentation of a yolk sac tumor with pericarditis, pericardial effusion and bilateral microlithiasis (MT) in a young Vietnamese male with extragonadal GCT in the anterior mediastinum.

Presentation of Case: A 23-year-old Vietnamese male with one-pack year smoking history, but no other significant past medical history presented with nonproductive cough and pleuritic chest pain for 2 weeks. Chest pain was described as sharp and localized to the right-side of his substernal area. B-symptoms were present for 1 month and included: weight loss of 10lbs, night sweats and fever. Physical exam showed diminished breath sounds on the right side of the chest w/o accessory muscle use. CT of the chest revealed a large 14 cm round mass in the right middle lobe with possible extension to the mediastinum, moderate right pleural effusion and pericardial effusion. EKG showed a pattern consistent with pericarditis. Biopsy of the mediastinal mass was consistent with yolk sac tumor. LDH was 543, AFP 7200, and β -HCG < 2. Testicular ultrasound did not find any testicular masses, but there was bilateral testicular microlithiasis. The patient was started on BEP (Bleomycin + Etoposide + Cisplatin) chemotherapy with subsequent thoracotomy and mass resection.

Conclusion: Younger patients presenting with pleural effusions and pericarditis need a detailed history and physical examination in order to diagnose a yolk sac tumor.

Keywords: Yolk Sac Tumor; Microlithiasis; Pericarditis; Extragonadal Germ Cell Tumors

Received: February 24, 2019; **Accepted:** March 24, 2019; **Published:** March 29, 2019

Competing Interests: The authors have declared that no competing interests exist.

Consent: Consent was taken from the patient's next of kin for publication of this case report.

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Introduction

Yolk sac tumor or endodermal sinus tumor is a rare form of a germ cell tumor (GCT) that occurs primarily in the testes or ovaries(1). In the US, the incidence of GCTs is 56 per 1,000,000 in Caucasians and 10 per 1,000,000 in African-Americans with the highest prevalence occurring before age 4 (2). Other locations of tumor detection besides the gonads are termed extragonadal GCTs (EGGCT) with the most common sites in decreasing order: anterior mediastinum, retroperitoneum and pineal glands (3-4). The overall prevalence is less than 5% of all germ cell malignancies (3-4). In this case report, we describe an unusual presentation of yolk sac tumor with pericarditis and bilateral microlithiasis in a young Vietnamese male with extragonadal GCT in the anterior mediastinum.

Case Presentation

A 23-year-old Vietnamese male with one-pack year smoking history, but no other significant past medical history presented to our emergency department (ED) with nonproductive cough and pleuritic chest pain for 2 weeks. Chest pain was described as sharp and localized to right-side of his substernal area. Non-radiating and no aggravating factors were noted. Alleviated with Ibuprofen use. B-symptoms for 1 month included: weight loss of 10lbs, night sweats and fever. The patient had a recent visit to an out-of-state ED about 6 weeks before current presentation for cough with unremarkable diagnostics and normal chest radiography. He was discharged with symptomatic treatment. About one and half weeks prior to admission, patient went to an urgent care center for continued coughing and persistent symptoms described above and was given Keflex, Medrol Dosepak and Prilosec. He denied headache, visual changes, hemoptysis, dysuria or abdominal pain. No past surgical history. Patient does not have a family history of any malignancies, TB infection or COPD. Admitted to smoking 2 cigarettes a day for 12-months, but reported that he quit about 6-months ago. Uses marijuana once a day for 6-months and occasionally drinks alcohol. No IV drug use and has resided in the United States for five years. No known drug allergies and only uses Ibuprofen as needed for chest pain besides the medications provided to him during his urgent care visit. Initial vitals on presentation showed blood pressure of 103/72 mm Hg, pulse 141 beats per minute, temperature 101.6°F (38.7°C), respiratory rate of 21 per minute with an oxygen saturation of 96% on room air. Patient was 5 feet 2 inches tall and weighed 97 pounds (44kg) with BMI 18. Physical exam revealed young, chronically ill appearing Vietnamese male lying in his bed in mild distress. He had diminished breath sounds on the right side of the chest w/o accessory muscle use. Normal S1, S2 with tachycardia. No murmurs, rubs or gallops. Extremities were negative for edema, cyanosis or clubbing. On neuro exam patient was alert & oriented x3. Cranial nerves 2-12 grossly intact. CBC showed white count of 14.6, hemoglobin 10.8, hematocrit 32.4 and platelets of 499 (Figure 1). BMP unremarkable with exception of mild hyponatremia and CO2 of 33 (Figure 1). Lactic acid 1.8. Chest x-ray revealed opacified right lung up to the upper lung field (Figure 2) and a CT of the chest revealed a large 14 cm round mass in the right middle lobe with possible extension from the mediastinum, moderate right pleural effusion and pericardial effusion (Figure 3). EKG showed a pattern consistent with rightward axis, inferior and anterolateral leads with a concave upwards character with some PR depression suggestive of pericarditis (Figure 4). Echocardiogram revealed normal left ventricular size and systolic function with an ejection fraction of approximately 60-65% (Figure 5). Mild-to-moderate tricuspid regurgitation with estimated pulmonary artery systolic pressure is 44 mmHg (Figure 5). There was a small to

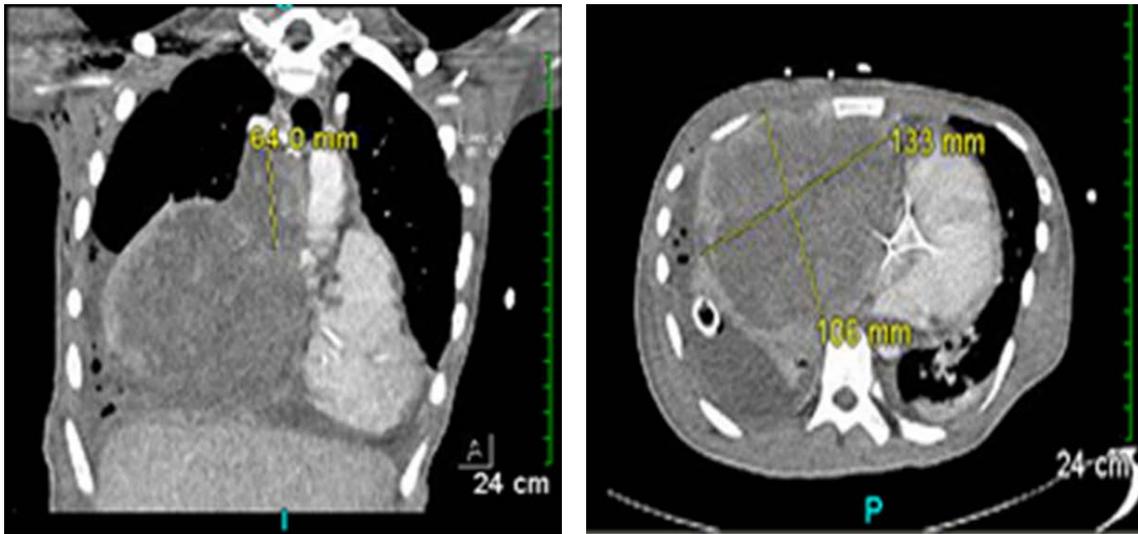


Figure 3 CT chest w/contrast. Mediastinum is deviated to the left due to a large anterior mediastinal mass measuring approximately 13.3 x 10.6 x 12.6 cm (right image). Also, there is an additional anterior mediastinal mass conglomerate measuring 6.4 x 3.9 x 6.4 cm (left image). Mass effect on the great vessels, heart, right lung and airway.

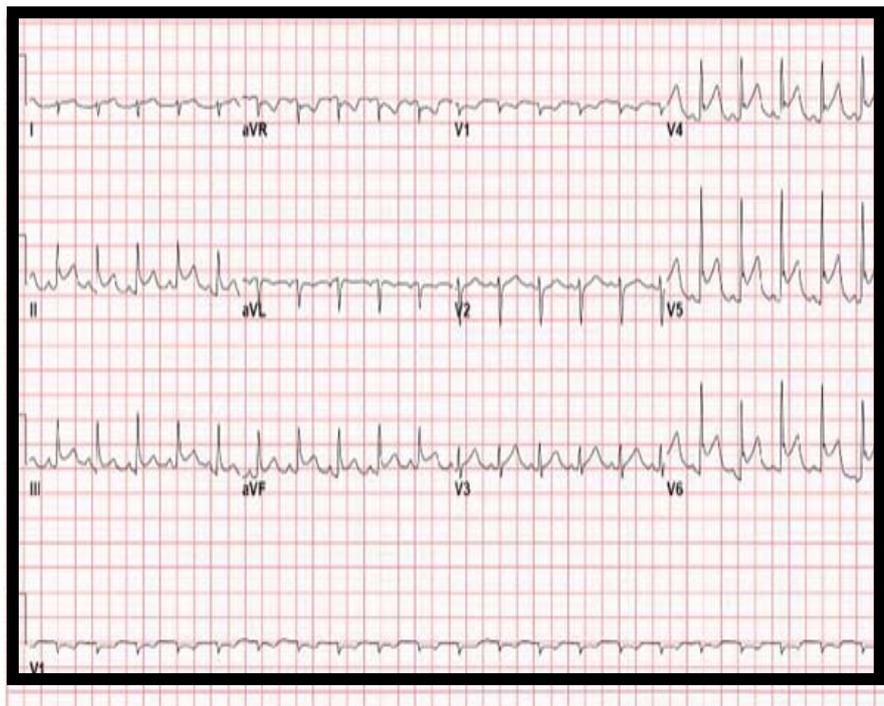


Figure 4 12-lead EKG. Ventricular rate 126. PR 110 ms & QRS 80 ms. Right axis w/diffuse ST elevation likely pericarditis.

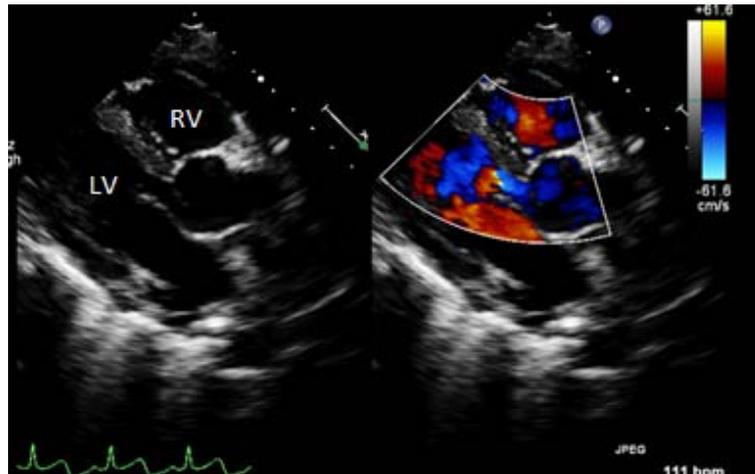


Figure 5 Transthoracic Echo. Normal left ventricular size and systolic function with an ejection fraction of approximately 60-65% on echo. Mild-to-moderate tricuspid regurgitation with estimated pulmonary artery systolic pressure is 44 mmHg. There was a small to moderate anterior and posterior pericardial effusion (RV= right ventricle, LV= left ventricle).



Figure 6 Hematoxylin & Eosin Stain. 400X Magnification. A. Tumor elements with associated eosinophilic hyaline globules (asterisk). B. & C. Endodermal sinus pattern with glomeruloid Schiller-Duval bodies (arrow).

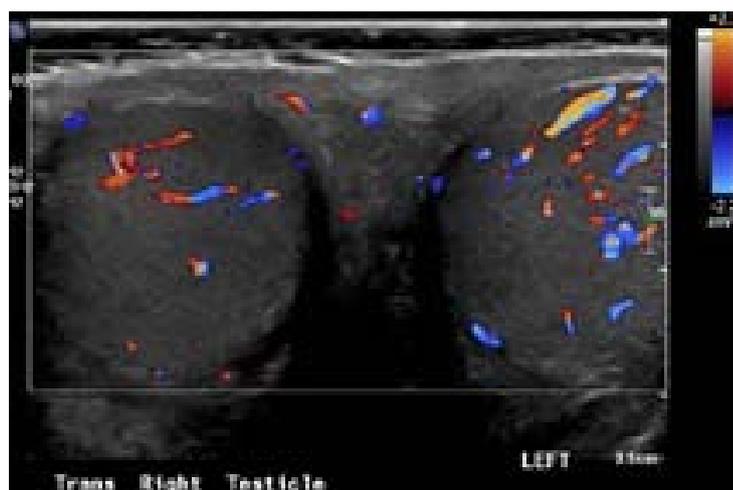


Figure 7 Testicular Ultrasound.No testicular masses. Equivocal bilateral testicular microlithiasis.

Discussion

Yolk sac tumor is a rare malignancy that likely arises from failed migration of germ cells during embryogenesis (5). The exact mechanism still remains unknown. The morphological pattern is of endodermal sinuses with Schiller-Duval bodies with diagnostic markers of alpha-fetoprotein, SALL4 and glypican-3 (6). The most common presentation is cough, weight loss, chest pain, and dyspnea (7). Ultrasound (US) is the diagnostic modality of choice as it can aid in distinguishing different testicular masses (9). Chest x-ray and CT should be performed if there is a high suspicion for metastasis (9).

The overall diagnosis for yolk sac tumors is very poor as the mass can metastasize rapidly to various parts of the body and is usually bulky/lobulated which can make it difficult to dissect (7-8). Treatment regimen include: bleomycin-etoposide-cisplatin (BEP) therapy or etoposide (Vepesid), ifosfamide, and cisplatin (VIP for at least 4 cycles (8).

MT is defined as • 5 testicular calcifications • 3 mm and associated with genetic syndromes such as Klinefelter's syndrome (10-11). It is caused by calcium deposits in seminiferous tubules from failed phagocytosis by Sertoli's cells within the tubules (10-12). Incidence in adults is around 5% and literature reviews performed have not found an association with MT and testicular tumors (12).

Mediastinal masses can present as common cardiac complaints including congestive heart failure with pleural effusions (13). Echocardiography is important to establish if either intra- or extrapericardial lesion is present (13). Leukemia, paraganglioma, schwannoma, carcinoid tumor, primitive neuroectodermal tumor, pheochromocytoma, and melanoma have been found to cause pericardial effusions (14). Most pericarditis cases are classified as idiopathic or viral induced (15). About 5% of pericarditis can be traced to malignancy such as mesotheliomas, fibrosarcomas, angiosarcomas, lymphoma, leukemia, lung, breast, ovary, prostate, colon, gastric, kidney, and bladder cancer (14-15). A detailed literature review performed, found limited reports of pericarditis cases induced by yolk-sac tumor.

Conclusion

Yolk sac tumor is a rare malignancy with an overall poor prognosis. Younger patients presenting with pleural effusions and pericarditis need a detailed history and physical examination in order to diagnose this rare condition.

Consent

A signed patient consent form was obtained.

Acknowledgement

We want to thank Dr. Prasad Garimella for his support and insights in the review of this case.

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