

An unusual case of pleural-based tumor with life-threatening post-operative complication

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Abstract

A 57-year-old male presented with hemoptysis of 4 years duration and a gradually increasing pleural mass on chest X-ray. The mass was causing pressure effects on the liver and the lungs. To rule out malignancy, thoracotomy was performed, which revealed large, thick-walled hematoma. Complete excision of mass was performed. Post-operative course was complicated by massive pleural bleeding requiring massive blood transfusions and re-exploratory thoracotomy. Subsequent tests revealed factor IX deficiency and, hence, he was managed with recombinant factor IX concentrate. This case stresses upon the fact that hereditary bleeding disorders may be diagnosed even in late adulthood with atypical presentations such as pseudotumor in pleural space. Moreover, hemophilia B may present with normal APTT levels making the diagnosis even more difficult.

Keywords: Factor IX deficiency, hemophilia B, pleural hematoma, pseudotumor

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Introduction

Differential diagnosis for a solitary pleural mass may include malignant or benign pleural tumors or mass related to ribs or chest wall. The metastatic involvement of the pleura is much more common than the primary pleural tumors.^[1] Loculated fluid in the pleural cavity may also radiologically present as a solitary pleural mass, and is referred to as a "pseudotumor." Here, we present a case of a 57-year-old male who presented with hemoptysis of 4 years duration and a gradually increasing pleural mass on chest X-ray (CXR) who developed life-threatening post-operative complication after thoracotomy, which was performed to rule out malignancy.

Case Report

A 57-year-old male, non-smoker, non-alcoholic and

with no co-morbidities or any significant past medical illness presented with complaints of mild, off and on hemoptysis and heaviness in the right chest for 4 years. He was diagnosed to have a right chest mass that had been increasing in size radiologically for the last 4 years. On examination, he was conscious, oriented and afebrile with a respiratory rate of 16/min, pulse of 74/min, blood pressure 120/80 mmHg and oxygen saturation of 97% on room air. Systemic examination was unremarkable except for reduced breath sound on right side of the chest. Routine laboratory reports were normal. INR was 1.13 and APTT was 30.2 s with a control of 29.0 s. CXR showed dense rounded pleural-based opacity in the right lower zone. Ultrasound revealed mild hepatomegaly and right lower pleural mass with no associated effusion. Computed tomography (CT) scan chest showed non-enhancing right pleural mass with specs of calcification compressing the superior surface of the liver [Figures 1].

One year back, CT-guided fine needle aspiration cytology (FNAC) was suggestive of an organized hematoma. About 1 month back, a Trucut biopsy had revealed dense fibrous tissue with marked hyalinization and inflammation with no evidence of granuloma or malignancy. Serial CXRs showed progressive

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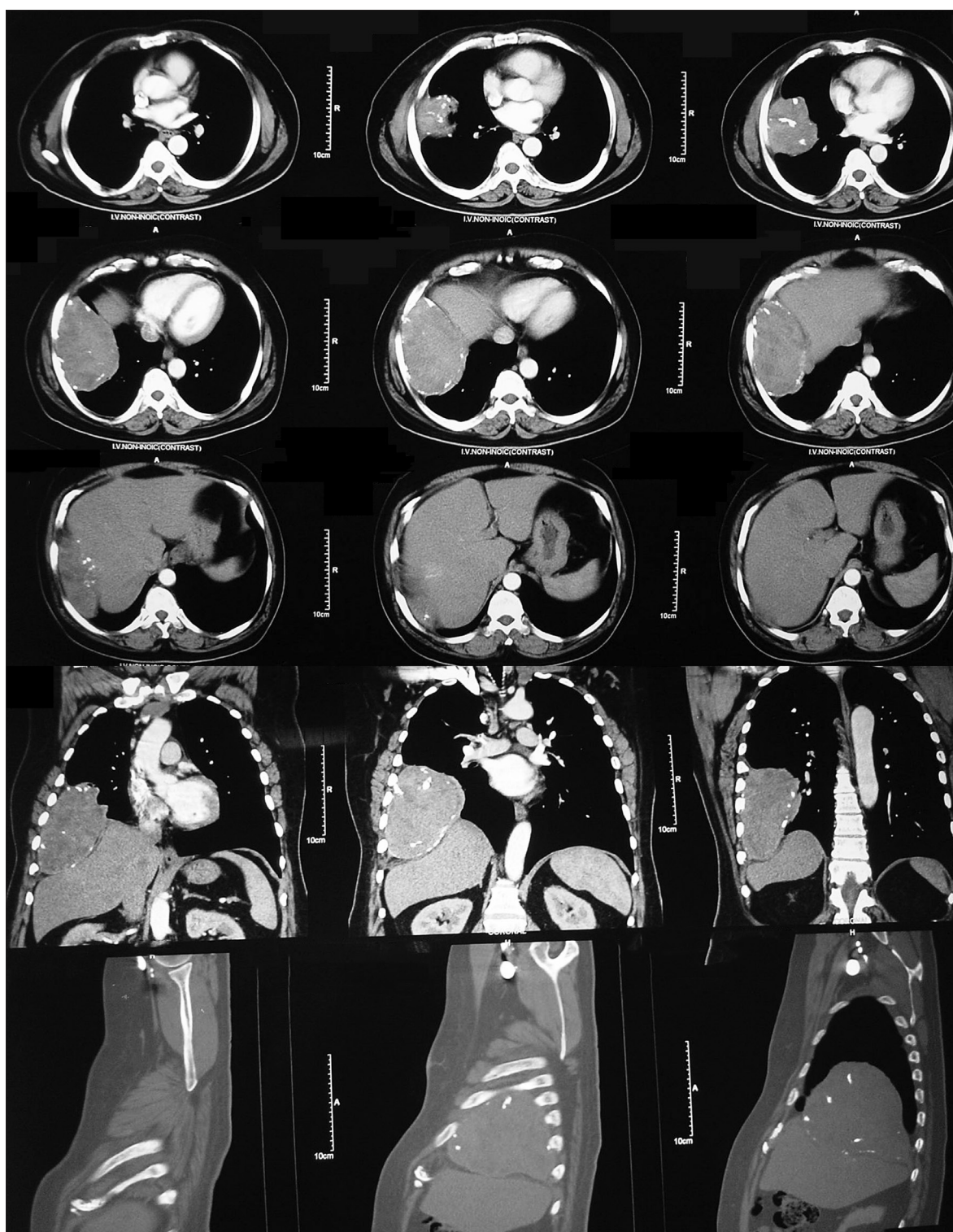


Figure 1: Computed tomography scan of the chest showing non-enhancing right pleural lesion abutting the anterior and lateral chest wall causing passive atelectasis of the underlying lung. Mild pleural thickening is also noticed

enlargement of the mass and, hence, a provisional diagnosis of a tumor of fibrous origin possibly undergoing malignant change was made. Therefore, a diagnostic and therapeutic exploratory thoracotomy was performed, which revealed a large, thick-walled hematoma in the

pleural space adherent to the lung. Complete excision of mass was carried out and an intercostal tube (ICT) was placed. Intraoperative hemostasis was achieved with great difficulty. In the first post-operative hour, ICT drained about 450 ml of frank blood. The patient

continued to bleed through ICT at a rate of 200–300 ml/h. He was managed with intravenous fluids, tranexamic acid, aprotinin, vitamin K and activated Factor VII in a dose of 7.2 mg, which was given empirically when bleeding could not be controlled with other medical measures. Despite multiple blood and blood product transfusions, bleeding did not stop. Re-exploration thoracotomy was done immediately. Per-operatively, more than 1 L of blood was seen in the pleural cavity with diffuse small oozes and with no evidence of any surgical bleed. Oozes were cauterized and complete hemostasis was ensured. After re-exploration, bleeding through ICT significantly reduced to 50 ml/h. Bleeding stopped gradually and the patient was extubated 48 h after re-exploration.

Histopathology of the pleural mass revealed fibrocollagenous pleural tissue with ulceration of mesothelial lining and foci of fibrin deposition [Figure 2], suggestive of an organizing hematoma. There was no evidence of granuloma or malignancy.

During the fourth to seventh post-operative days, the patient's general condition gradually worsened. He became progressively breathless and his hemoglobin dropped to 6.7 g/dl. There was progressive worsening of CXR suggestive of collection of clots on the right side, even though ICT was not draining anything. The hemoglobin continued to drop and he was unable to maintain adequate oxygen saturation with high-flow oxygen and, hence, was started on BiPAP support. Blood sample for factor VIII and factor IX activity was sent. He was again transfused packed cells and blood products. On the seventh post-operative day, Factor VIII levels were reported to be 103% of normal (60–150),

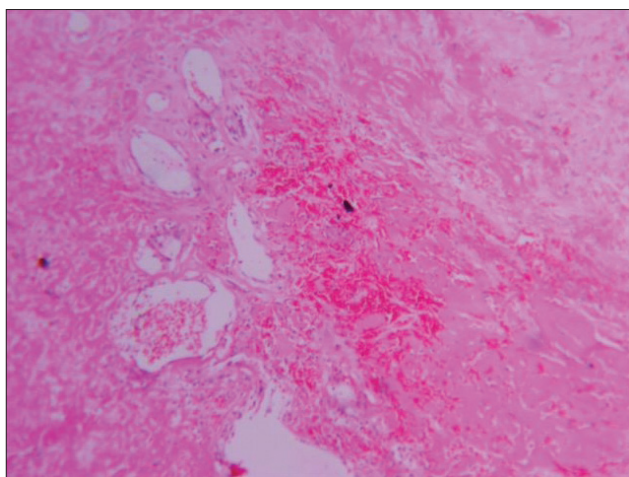


Figure 2: Histopathology specimen of the pleural mass showing blood clots and few newly formed blood vessels (x10 hematoxylin and eosin stain)

whereas Factor IX (functional) was only 24.5%. A repeat sample to another laboratory also showed only 18% Factor IX activity. Fibrinogen, FDP and Factor XIII levels were normal. Factor IX concentrate 1800 iu was given intravascularly, which immediately stopped the bleeding.

Factor IX concentrate 1800 iu once daily was continued for the next 4 days, after which repeat assay was 80% of normal. Dose of factor IX was gradually reduced to 600 iu and then stopped after 7 days. The patient continued to improve and was discharged on the 16th post-operative day and did not develop any further complications even after 6 months of follow-up. Complete familial work-up was done and his daughter and granddaughter were both found to be carriers for hemophilia B.

Discussion

Hemophilia is a genetic bleeding disorder that is inherited in a sex-linked recessive pattern. Two most common forms are hemophilia A and B, which are caused by a defect or deficiency of clotting factors VIII and IX, respectively. As these disorders are x-linked recessive, only males exhibit the disease and females are carriers. Worldwide, the incidence of hemophilia A and B is about 15–20 per 100,000 males.^[2] Hemophilia A is almost five-times more common,^[3] with the reported incidence of hemophilia B or Christmas disease being around 1 in 60,000.^[4]

The underlying pathophysiology of hemophilia B involves inadequate generation of thrombin, which causes reduced clot formation and reduced clot stabilization. Clinically, it commonly manifests as mucosal bleeding or as spontaneous or post-traumatic bleeding typically into the joints or deep muscles. Pulmonary involvement is rarely reported.^[5–7] The age at diagnosis is related to the level of factor IX activity. Patients with a milder form might not be diagnosed until later in life.

Only a few cases of hemophilia B with primary pulmonary involvement have been reported in the literature.^[5–7] O'Dwyer and DeLoughery reported a case of hemophilia B with pulmonary hemorrhage with bleeding inside a bulla mimicking a lung neoplasm.^[6] Grabczak *et al.* also reported a case of hemophilia B with hemothorax, which also developed a massive post-operative bleed after video-assisted thoracic surgery with lung decortication was performed to manage hemothorax.^[5]

Significantly prolonged APTT may typically suggest a diagnosis of hemophilia, but APTT may be normal or only minimally prolonged in patients with less-severe disease, which may make the diagnosis more challenging, as in our case.^[8] Hence, massive post-operative bleeding was totally unsuspected in our case. Confirmatory diagnosis can be established by demonstrating low-factor IX clotting activity. Normally, factor IX clotting activity is 50–150%.^[9] Hemophilia B has been classified as mild, when activity is between 5 and 30%, moderately severe when activity is between 1 and 5% and severe when it is less than 1%.^[10] According to this, our patient may qualify as having a mild disease with factor IX activity of 24.5%.

This case stresses upon the fact that hereditary bleeding disorders may be diagnosed even in late adulthood with atypical presentations. Moreover, hemophilia B may present with normal APTT levels, making the diagnosis even more difficult.

References

1. Leung AN, Müller NL, Miller RR. CT in differential diagnosis of diffuse pleural disease. *AJR Am J Roentgenol* 1990;154:487-92.
2. Bell B, Canty D, Audet M. Hemophilia: An updated review. *Pediatr Rev* 1995;16:290-8.
3. Kar A, Potnis-Lele M. Descriptive epidemiology of haemophilia in Maharashtra, India. *Haemophilia* 2001;7:561-7.
4. Mannucci PM. Hemophilia and related bleeding disorders: A story of dismay and success. *Hematology Am Soc Hematol Educ Program* 2002;1-9.
5. Grabezak EM, Krenke R, Jeleńska M, Komarow I, Ambroziak U, Fangrat A, *et al.* Hemorrhagic parapneumonic effusion in a 64 year-old patient as the first symptom of hemophilia B. *Pneumonol Alergol Pol* 2009;77:320-6.
6. O'Dwyer ME, DeLoughery TG. Pulmonary hemorrhage in a hemophiliac simulating a lung neoplasm. *Am J Hematol* 200;64:299-302.
7. Turiaf J, Battesti JP, Menault M, Le Brigand H. Spontaneous hemothorax revealing a case of B hemophilia. *Poumon Coeur* 1970;26:253-60.
8. Park CH, Seo JY, Kim HJ, Jang JH, Kim SH. A diagnostic challenge: Mild hemophilia B with normal activated partial thromboplastin time. *Blood Coagul Fibrinolysis* 2010;21:368-71.
9. Khachidze M, Buil A, Viel KR, Porter S, Warren D, Machiah DK, *et al.* Genetic determinants of normal variation in coagulation factor (F) IX levels: Genome-wide scan and examination of the FIX structural gene. *J Thromb Haemost* 2006;4:1537-45.
10. Brower CL, Thompson AR. Hemophilia B. In: Pagon RA, Bird TD, Dolan CR, Stephens K, editors. *GeneReviews* <http://www.ncbi.nlm.nih.gov/books/NBK1495/>. Seattle (WA): University of Washington, Seattle; 1993. [Last accessed on 2011 Mar 15].

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