

Case Report

Report of Massive Bleed after Chest-drain Insertion in a Case of Job's (hyper-IgE) Syndrome

Prabudh Goel, Vishesh Jain, Sanjeev Kumar¹, Sanjiv Sharma¹, Minu Bajpai

Department of Pediatric Surgery and ¹Cardiac Radiology, All India Institute of Medical Sciences, New Delhi, India

ABSTRACT

Chest-drain insertion is considered to be a benign procedure. A 6-year-old girl who is a known case of hyper-IgE/Job's syndrome presented to us following three episodes of fresh bleed following chest-drain insertion for the left lung abscess. The site of bleed was localized to be the ruptured pseudoaneurysm of the anterior segmental branch of the left pulmonary artery. For successful management, various pitfalls and gray areas have been highlighted: (a) the procedure of chest-drain insertion has a potential for many complications some of which may be life-threatening, (b) the importance of proper history taking and a complete evaluation of all patients before surgical intervention, (c) role of ultrasound-guided chest-drain insertion, and (d) the importance of lateral and pragmatic approach with multidisciplinary involvement in such unusual and challenging cases cannot be overemphasized.

KEYWORDS: *Angioembolization, chest-drain insertion, hyper-IgE syndrome, intercostal tube-drain insertion, Job's syndrome, lung abscess, pseudoaneurysm*

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INTRODUCTION

Outcomes of incorrect chest-drain insertion such as rupture of the right^[1] and left ventricles,^[2] pericardial tamponade,^[3] mediastinal perforation, and contralateral hemothorax^[4] have been described. Collop *et al.*^[5] estimated a 10% complication rate even in expert hands. Doctors in nearly all specialties are exposed to chest-drain insertions at least occasionally; hence, there is a need for awareness on how to perform this procedure safely.

CASE REPORT

A 6-year-old girl with a week's history of high-grade fever, cough, and breathlessness consulted locally. Chest radiograph revealed a large cavity with air–fluid interface in upper half of the left hemithorax. Contrast-enhanced computed tomography (CT) of chest in lung [Figure 1b] and mediastinum [Figure 1a] windows demonstrated the presence of large collection (suspected abscess) with air–fluid (fluid–air: 220/40 ml) interface in the region of the left upper lobe with smooth compression of the left main bronchus (LMB) due to mass effect and a left-to-right mediastinal shift.

Chest drain inserted under local anesthesia drained 50 ml blood-tinged pus; there was a persistent collection on postdrain imaging [Figure 1c]. Replacement of chest drain resulted in a gush of fresh blood followed by clots and hypovolemic shock. Two more episodes of bleed occurred in the next 4 days; both episodes were preceded by bouts of unrestrained cough. The patient was resuscitated, chest drain removed, entry wound strapped, and referred to our center.

She presented to us in hemodynamic collapse.

She was a known case of genu valgum, H-type rectovestibular fistula, and hyper-IgE syndrome (HIES). HIES was diagnosed following the development of recurrent multiple abscesses at 2 months of age (IgE level – 13608.6 IU/mL against normal range of <393 IU/mL). Surgeries for anorectal malformation were also followed by prolonged wound infections. Additional hospital admissions for scalp and chest abscesses were

Address for correspondence: Prof. Minu Bajpai,
All India Institute of Medical Sciences,
New Delhi - 110 029, India.
E-mail: bajpai2b@gmail.com

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on record. The patient's mother was a known case of HIES and had succumbed to infections post-prolonged hospitalization. However, no genetic workup for the same was available.

Upon presentation, the primary assessment was followed by prompt initiation of stabilization measures. The review of CT films revealed the presence of hyperdensities within the collection, suggesting the possibility of a hematoma due to ruptured mycotic pseudoaneurysm [Figure 1b].

Localization of bleed

Multiphase CT angiogram was performed under sedation to evaluate the hematoma. Thin maximum intensity projection image documented a 2.2 cm × 1.8 cm contrast-filled structure near the medial wall of the abscess arising 1.2 cm from the anterior segmental branch of the left pulmonary artery, suggestive of pseudoaneurysm [Figure 1d]. The abscess cavity revealed the presence of heterogeneous contents.

Definitive management

Given patient's poor general condition, embolization of the pseudoaneurysm was considered appropriate. Selective digital subtraction angiography was performed with a 5 Fr vertebral artery catheter under general anesthesia via the right femoral route. The findings of CT angiography were confirmed and the arterial bleeding source was localized [Figure 2a and b].

The pseudoaneurysm neck was occluded with Reye® Embolization Coil (Cook India Medical Devices Pvt., Ltd.) and loss of flow confirmed on postembolization angiogram [Figure 2c].

Postprocedure and follow-up

Subsequently, the clinical course was uneventful and recovery smooth.

DISCUSSION

HIES or Job's syndrome was first described in 1966 by Davis, Wedgwood, and Schaller.^[6]

HIES is a complex primary immunodeficiency disorder characterized by the classic triad of recurrent staphylococcal abscesses, recurrent airway infections, and increased concentration of IgE in serum. Pneumonias are typically complicated by lung abscesses, bronchiectasis, bronchopleural fistulas, and formation of pneumatocele. This leads to colonization by opportunistic microorganisms, invasive aspergillosis, and aspergillum formation. Pulmonary sequel leading to chronic respiratory insufficiency and hemoptysis complicating lung abscess are the most common causes of death in these patients.

Several vascular anomalies in association with HIES have reported in literature, both congenital or acquired and venous or arterial.^[7] Such anomalies may also

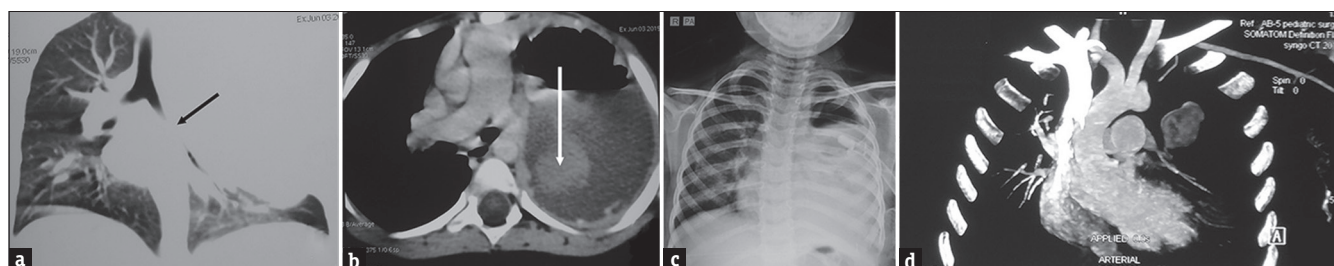


Figure 1: (a) Coronal section lung window high-resolution computed tomography chest: Space Occupying Lesion (SOL) in the left upper hemithorax compressing the left main bronchus (black arrow). (b) Axial section mediastinal window contrast-enhanced computed tomography chest: Large cystic lesion in the left upper hemithorax with air-fluid level suggestive of abscess. The presence of contrast in the abscess cavity is suggestive of active bleed/pseudoaneurysm (white arrow). (c) Anteroposterior view of X-ray chest: Persistence of the cavity with air-fluid level; chest tube *in situ*. (d) Coronal section chest computed tomography angiography: Pseudoaneurysm arising from anterior segmental branch of the left pulmonary artery

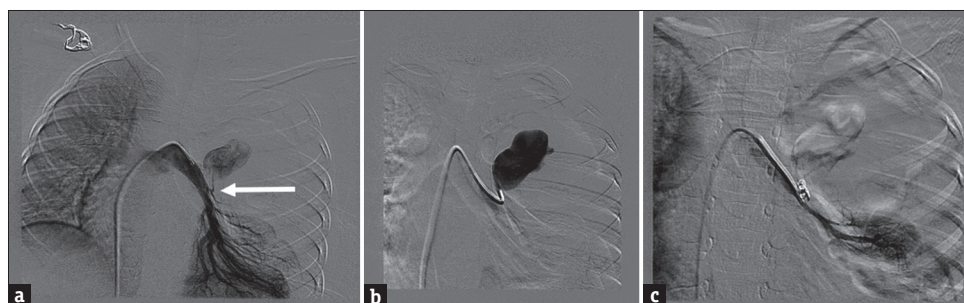


Figure 2: (a) Angiogram with the catheter in the left pulmonary artery showing the presence of a pseudoaneurysm arising from anterior segmental branch of the left pulmonary artery (white arrowhead). Normal flow to the left lower lobe is seen. (b) Demarcation of the pseudoaneurysm. (c) Angiogram after embolization of the vessel showing the absence of flow of the contrast in the pseudoaneurysm

be fatal. The exact pathogenetic mechanisms are not clear; factors – hypereosinophilia, vasculitis, and defective angiogenesis – have been implicated. Vascular anomalies such as aneurysms of various systemic arteries, pseudoaneurysms, vasculitis, vascular occlusion, and vascular ectasia have been described in HIES.^[7] Females were more predisposed with a ratio of 5:1 in the autosomal recessive group and 5:3 in the autosomal dominant group.

The index case is a known case of HIES with a positive family history in the mother. The prevalence of aneurysms and pseudoaneurysms of systemic arteries in these patients calls for additional caution; ultrasound-guided chest-drain insertion in these patients may be safer and may have averted this misfortune.

The event started with a simple chest infection, but progressed to the development of lung abscess. Probably, the inflammatory and necrotic processes during the evolution of abscess eroded through the wall of the preexisting pseudoaneurysm from the anterior segmental branch of the left pulmonary artery. The CT images before insertion of chest drain (elsewhere) are suggestive of the presence of hematoma in the abscess cavity and this finding went unnoticed inadvertently. Probably, the hematoma-induced heterogeneity of the abscess cavity was attributed to the presence of blood and necrotic material in the abscess cavity.

The findings of compressed LMB also point toward the presence of a ruptured pseudoaneurysm communicating with the abscess cavity. A normal abscess is less likely to generate enough pressure to compress the cartilaginous wall of the LMB as compared to this situation wherein the pressure inside the abscess is augmented by the arterial inflow. The same findings were not replicated in the follow-up CT angiogram done at our center since the pressure was being released intermittently in the form of Inter-Costal Drain(ICD) bleed.

It seems that the initial chest tube drained a loculus of collection not communicating with the main abscess cavity. Subsequently, when the chest drain was replaced, the disaster was precipitated. Ultrasound-guided chest-drain placement may be beneficial in similar situations.

CONCLUSIONS

- a. The procedure of chest-drain insertion is seemingly benign and is often delegated to the junior member of the team. However, we must understand and, more

importantly, emphasize on the trainees that it has the potential for serious complications which may be life-threatening

- b. The importance of proper history from every patient is highlighted. The prevalence of vascular anomalies such as pseudoaneurysms in HIES is known and merits caution
- c. Ultrasound-guided chest-drain insertion may be considered in such situations.

The authors also wish to highlight the fact that such unusual and challenging cases may not always be handled by the book. Often, a lateral and pragmatic approach with multidisciplinary involvement in an institutional setting may be lifesaving.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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