
In the Name of God

A case with Hemoptysis

By : Saeedeh Khajouie Nejad , MD

Case Presentation

- **Chief Complaint :** hemoptysis
- **Present illness :** A 37 Y/O male presented with intermittent episodes of mild hemoptysis for about 4 years. There was no history of dyspnea, chest pain, weight loss, arthritis & skin lesions .
- **PMH :** Seasonal allergies
- **DH :** Anti-histamines
- **SH :** C/S _ , O/A _ , A/A _
- **FH :** DM +, HTN + , HLP + , Blood Malignancies +
- **AH :** Seasonal allergies , Shrimp
- **Occupation :** Civil Engineer

Case Presentation

- **GA :** the patient was young, alert & oriented . He was not ill or in respiratory distress .
- **Vital Signs :** Stable
- **Skin :** not pale / not icteric / no cutaneous lesion or ecchymosis or petechiae
- **H&N :** no cervical or supraclavicular LAP
- **Chest :**

✓Heart : S1 & S2 heard without murmur

✓Lung : Bilateral clear

- **Abdoman :** NL
- **Extremities :** NL , no clubbing

What did we do ?

1. Medicine Therapy : Antihistamines, Antibiotics, …
2. Medical Tests
3. ENT Consult
4. Chest CT scan
5. Bronchoscopy
6. Open Lung Biopsy

Lab Data

BS	**94**	**WBC**	**6.2**
BUN	17	Hb	16.8
Cr	1.08	PLT	179
Acid Uric	6.7	PT	13
Ast	15	INR	1.1
Alt	9	PTT	33
LDH	345	U/A	NL
ESR	5	CPK	103
CRP	0.2	Total Pro	7

Lab Data

ANA	0.1	C3	NL
C-ANCA	8.7	C4	NL
P-ANCA	2.4	DsDNA	Neg
Lupus Anti Coagulant	Neg	IgA	143
Phospholipid Ab	Neg	IgM	78
Cardiolipin Ab	Neg	B2Glycopro	Neg
IgG4	NL	NBT	98 (80%PMN)
Anti RO	Neg	IgG	1300

Aldolose	**6.4 NL**
ACE	**40 NL**
TSH	**1.41**
IGRA	**Neg**
Sputum for B/K	**Neg**
Pcr for COVID19	**Neg**

ENT Consult

- Laryngoscopy was NL
- No upper respiratory hemorrhage

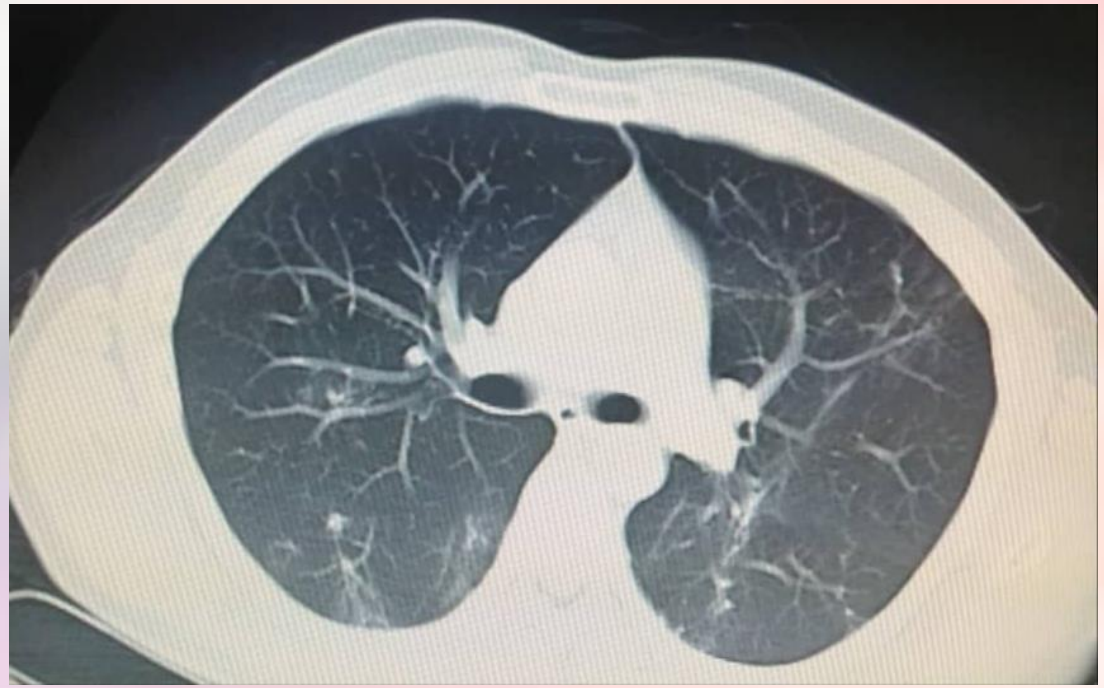
Chest CT Scan

- 99/2/27 :
  - Multiple GGO in both lungs
  - High suggestive for COVID19
- 1400/7/20 : Chest CT Angiography
  - No PTE
  - No Arteriovenous Malformation
  - Multiple reticulonudolar opacities
  - GGO opacities
  - Peripheral thickening

Chest CT Scan

- Before Thalidomide
- 1400/5/13
  - Multiple small opacity & GGO in both lungs
  - Both lungs are emphysematous
  - Pneumonia
  - Reticular pattern
  - No mediastinal LAP
  - Suspected for covid19

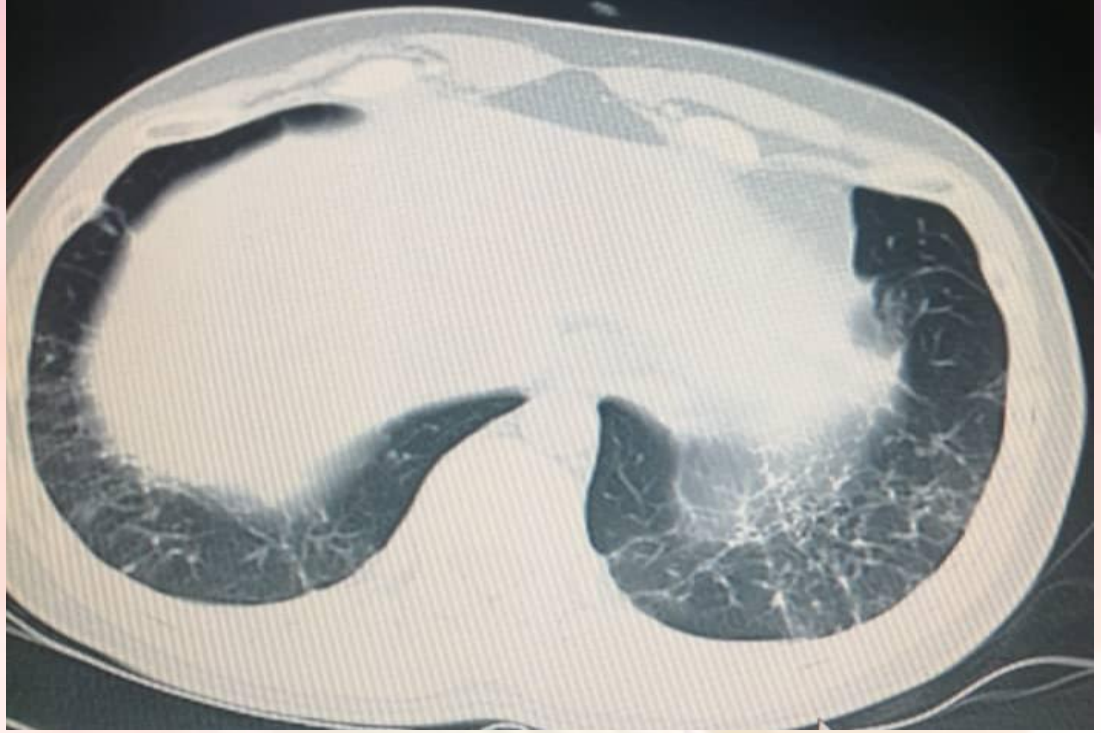








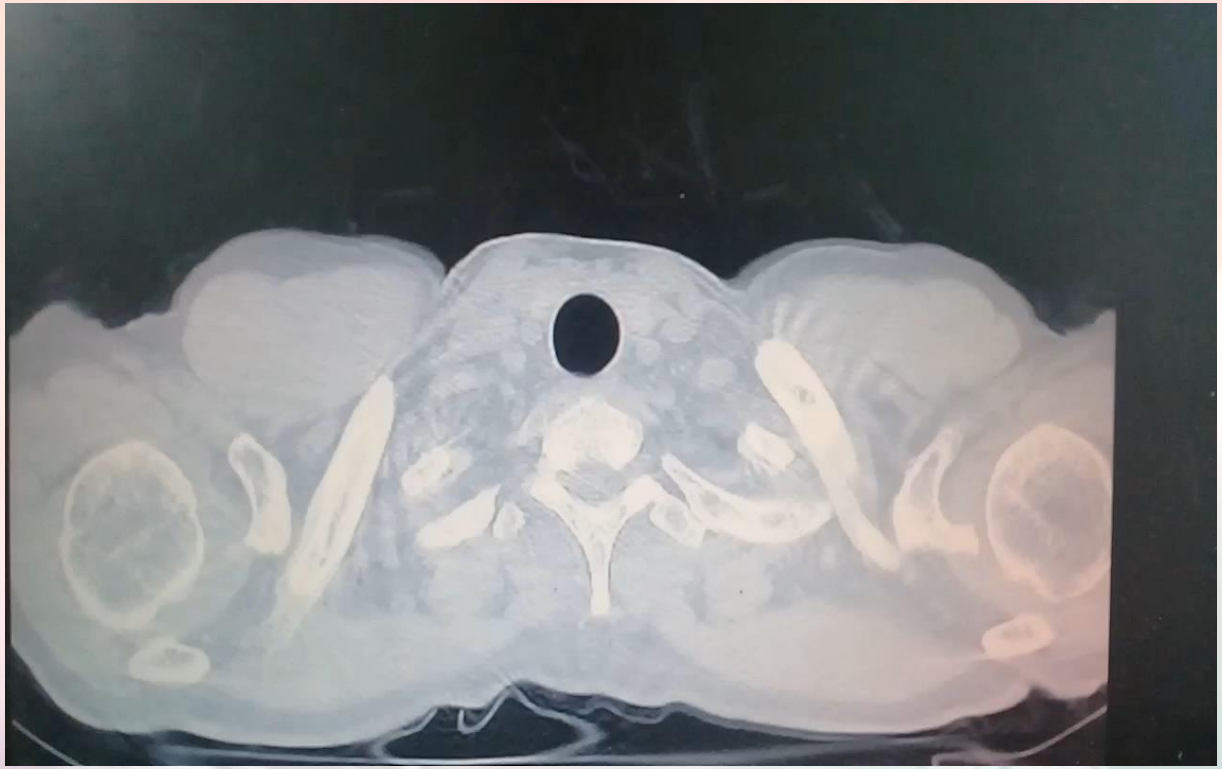


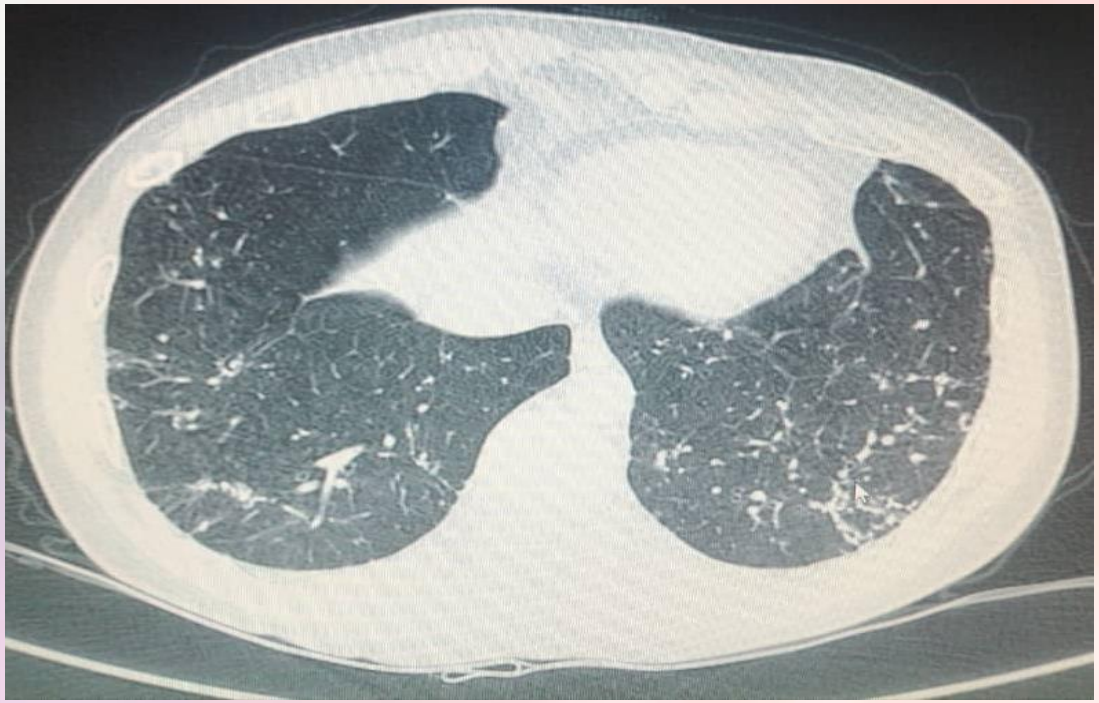


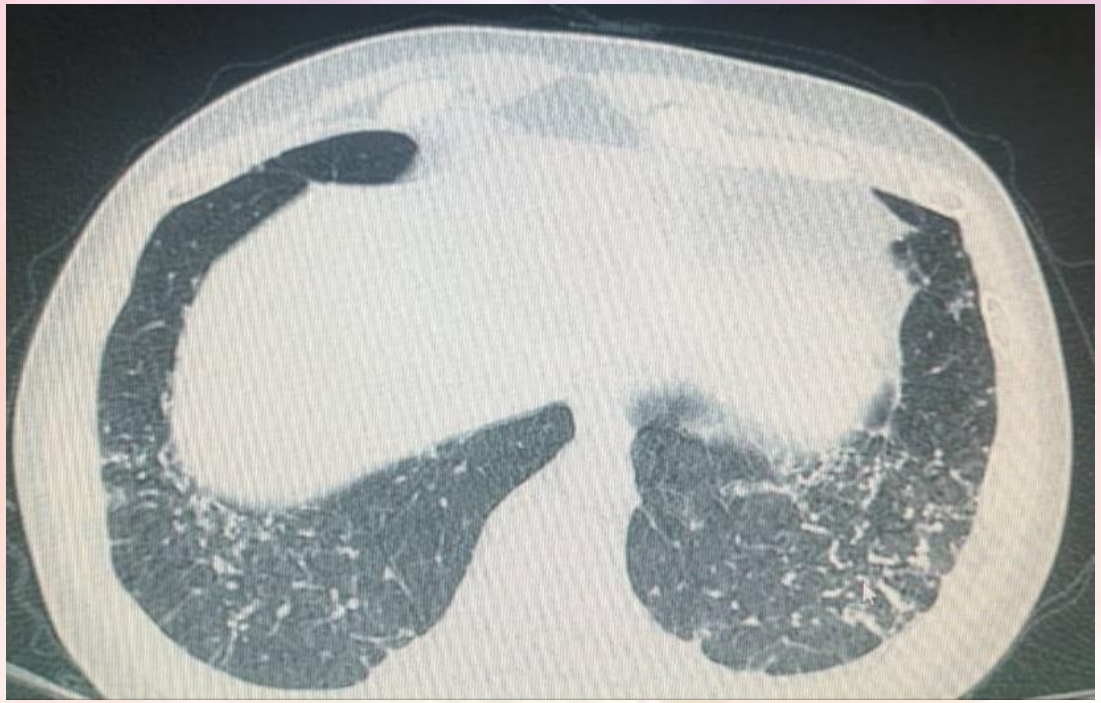


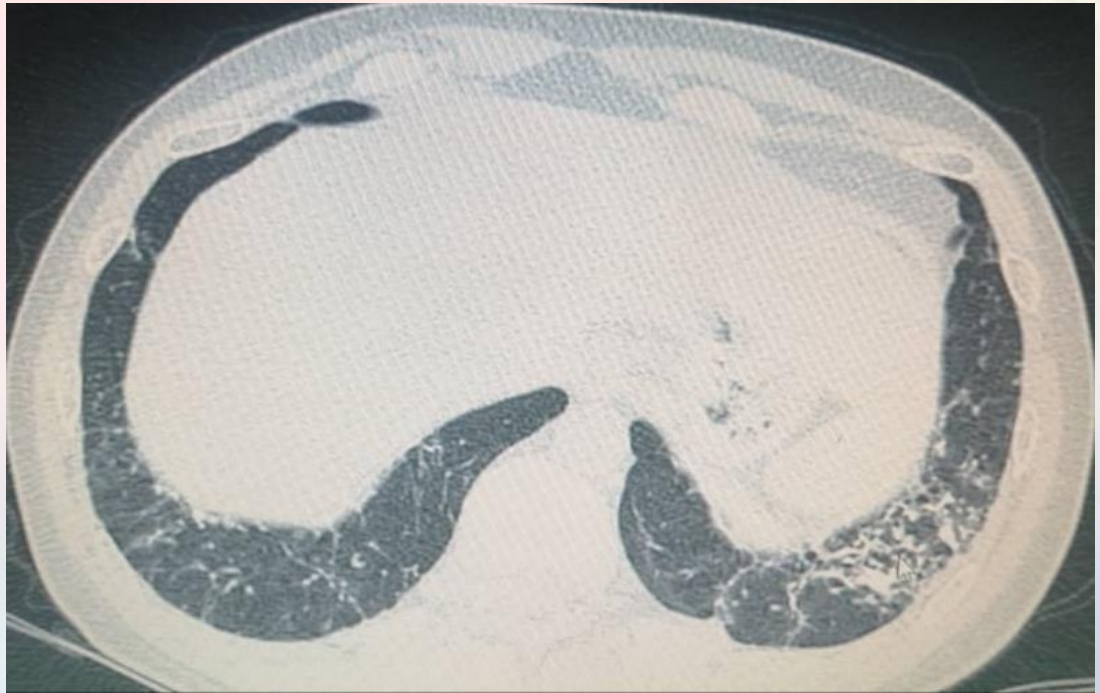
Chest CT Scan

- After Thalidomide
- 1401/5/18
  - Multiple small opacity in both lungs
  - Pripheral calcification in LT lung
  - Reticulonodular pattern





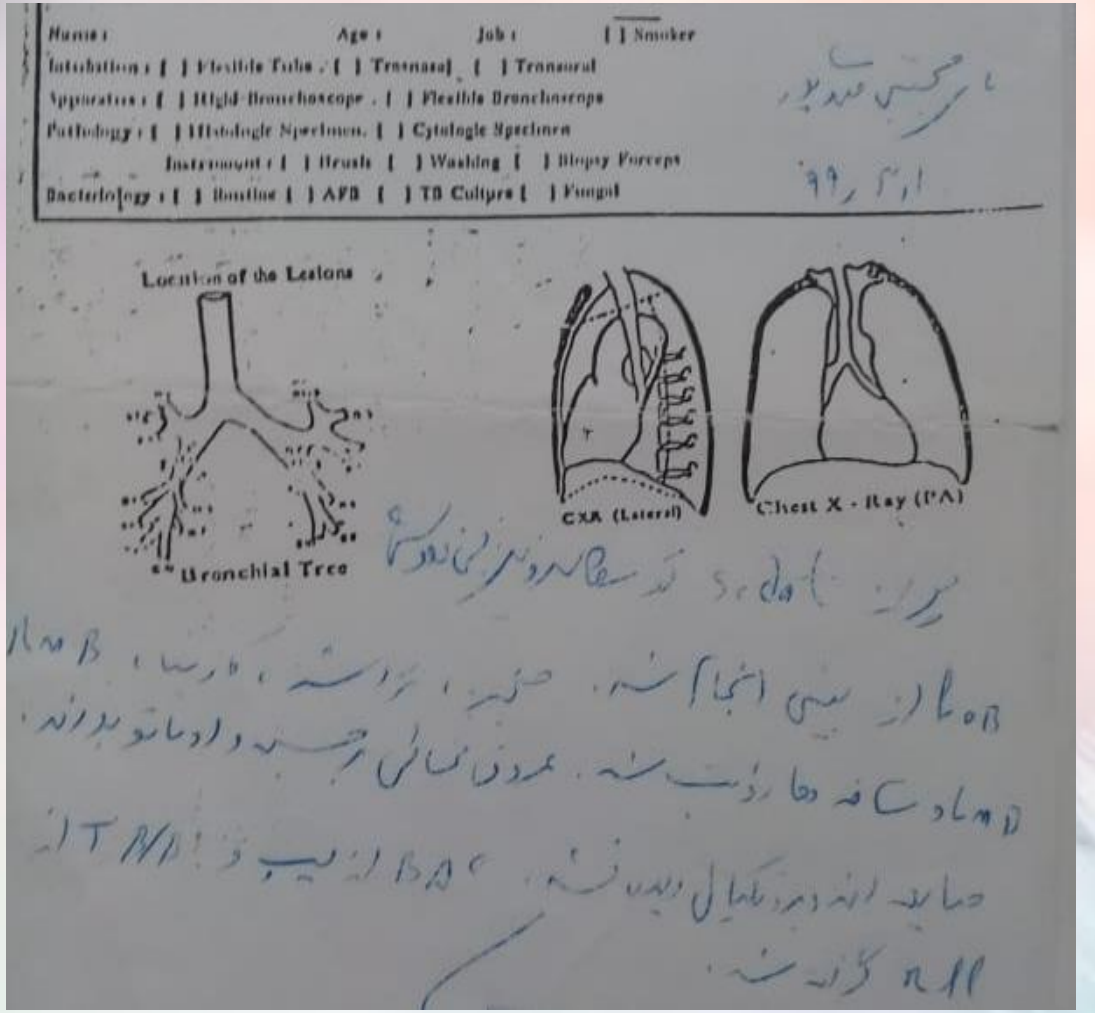


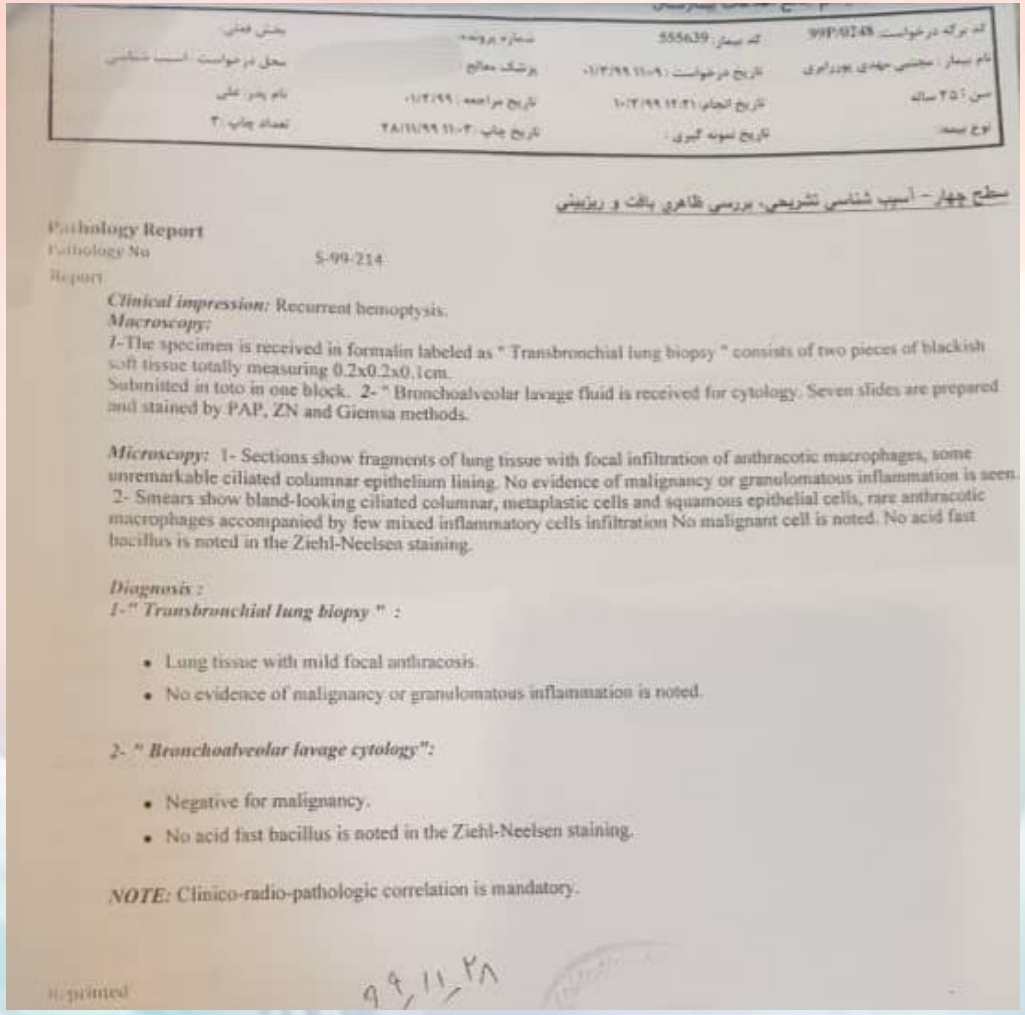


Bronchoscopy

- 99/3
- Prominent and edematous mucosal vessels
- No endobronchial lesions
- TBLB :
  - Lung tissue with mild focal anthracosis
  - No evidence of malignancy or granulomatous inflammation
- BAL cytology:
  - Neg for malignancy
  - No acid fast bacillus is noted in the Ziehl-Neelsen staining

Bronchoscopy





Pathology Report

Pathology No S-99-214

Report

Clinical impression: Recurrent hemoptysis.

Macroscopy:

*1-*The specimen is received in formalin labeled as " Transbronchial lung biopsy " consists of two pieces of blackish soft tissue totally measuring 0.2x0.2x0.1cm. Submitted in toto in one block. 2- " Bronchoalveolar lavage fluid is received for cytology. Seven slides are prepared and stained by PAP, ZN and Giemsa methods.

Microscopy: 1- Sections show fragments of lung tissue with focal infiltration of anthracotic macrophages, some unremarkable ciliated columnar epithelium lining. No evidence of malignancy or granulomatous inflammation is seen. 2- Smears show bland-looking ciliated columnar, metaplastic cells and squamous epithelial cells, rare anthracotic macrophages accompanied by few mixed inflammatory cells infiltration No malignant cell is noted. No acid fast bacillus is noted in the Ziehl-Neelsen staining.

Diagnosis :

1-" Transbronchial lung biopsy " :

- Lung tissue with mild focal anthracosis.
- No evidence of malignancy or granulomatous inflammation is noted.

2- " Bronchoalveolar lavage cytology":

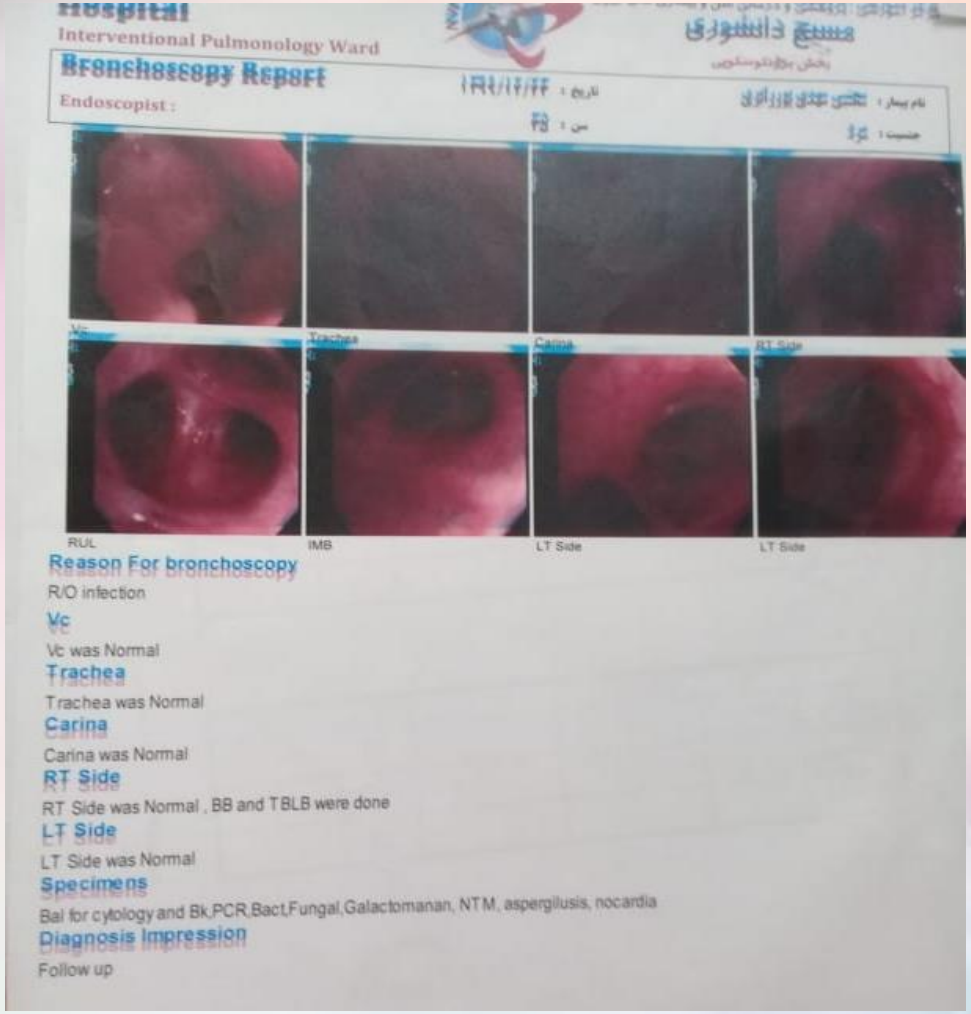
- Negative for malignancy.
- No acid fast bacillus is noted in the Ziehl-Neelsen staining.

NOTE: Clinico-radio-pathologic correlation is mandatory.

Bronchoscopy

- 99/12
- Normal
- BAL for cytology, BK PCR, bacteria, fungal, NTM, aspergilusis, nocardia was Negative .
- BAL for Galactomanan : 0.92 (>0.5 pos)
- Smear & cell block : no malignancy seen
- Bronchial biopsy : framnents of mildly inflamed respiratory mucosa.
- TBLB : lung parenchyma with anthracotic pigment deposition and presence of hemosiderin laden macrophages in few alveolar spaces.

Bronchoscopy



Hospital

Interventional Pulmonology Ward

Bronchoscopy Report

Endoscopist :

RUL IMB LT Side LT Side

Reason For bronchoscopy

R/O infection

Vc

Vc was Normal

Trachea

Trachea was Normal

Carina

Carina was Normal

RT Side

RT Side was Normal , BB and TBLB were done

LT Side

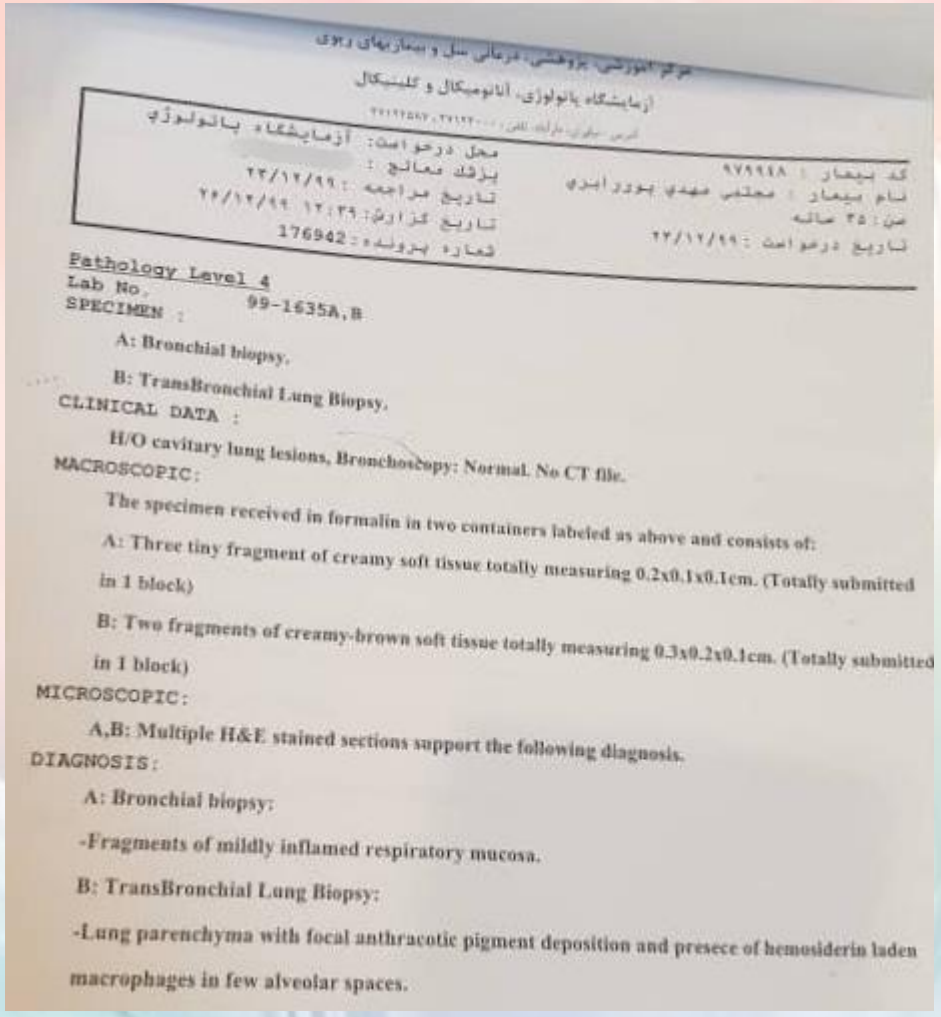
LT Side was Normal

Specimens

Bal for cytology and Bk,PCR,Bact,Fungal,Galactomanan, NTM, aspergilusis, nocardia

Diagnosis Impression

Follow up



زایشگاه پاتولوژی، آناتومیکال و کلینیکال

محل درخواست: آزمایشگاه پاتولوژی

بزشک معالج :

تاریخ مراجعه : ۲۲/۱۲/۹۹

تاریخ گزارش: ۲۲/۱۲/۹۹ ۱۲:۳۹

شماره پرونده: 176942

کد بیمار : ۹۷۹۹۱۸

نام بیمار : مجتبی مهدی پوردابری

سن: ۳۵ ساله

تاریخ درخواست : ۲۲/۱۲/۹۹

Pathology Level 4

Lab No. 99-1635A,B

SPECIMEN :

A: Bronchial biopsy.

B: TransBronchial Lung Biopsy.

CLINICAL DATA :

H/O cavitary lung lesions, Bronchoscopy: Normal. No CT file.

MACROSCOPIC:

The specimen received in formalin in two containers labeled as above and consists of:

A: Three tiny fragment of creamy soft tissue totally measuring 0.2x0.1x0.1cm. (Totally submitted in 1 block)

B: Two fragments of creamy-brown soft tissue totally measuring 0.3x0.2x0.1cm. (Totally submitted in 1 block)

MICROSCOPIC:

A,B: Multiple H&E stained sections support the following diagnosis.

DIAGNOSIS:

A: Bronchial biopsy:

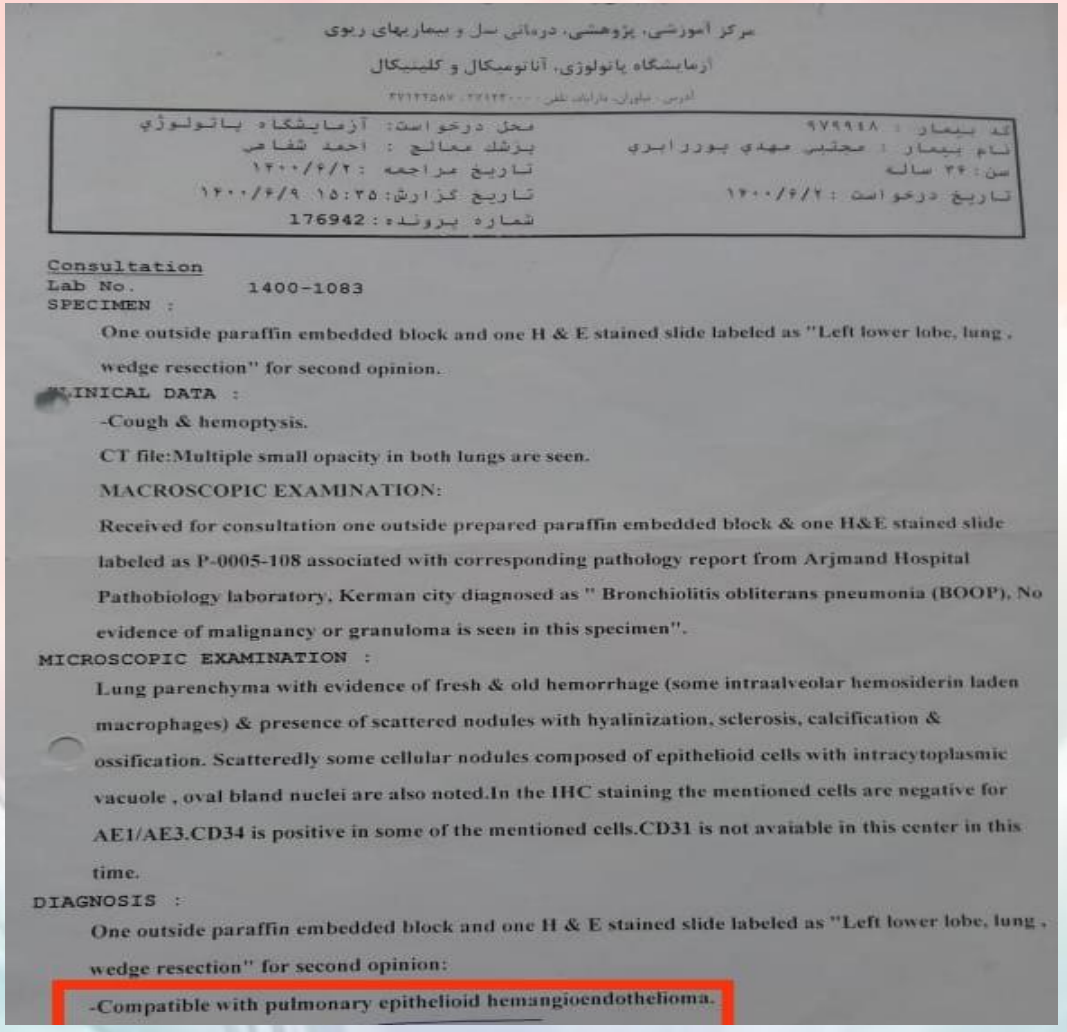
-Fragments of mildly inflamed respiratory mucosa.

B: TransBronchial Lung Biopsy:

-Lung parenchyma with focal anthracotic pigment deposition and presece of hemosiderin laden macrophages in few alveolar spaces.

Open Lung Biopsy

- Pulmonary Epithelioid Hemangioendothelioma



مرکز آموزشی، پژوهشی، درمانی سل و بیماریهای ریوی

آزمایشگاه پاتولوژی، آناتومیکال و کلینیکال

کد بیمار : ۹۷۹۹۴۸	محل درخواست: آزمایشگاه پاتولوژی
نام بیمار : مجتبی مهدی پوررابری	پزشک معالج : اجمه شفاهر
سن: ۴۴ ساله	تاریخ مراجعه : ۱۴۰۰/۶/۲
تاریخ درخواست : ۱۴۰۰/۶/۲	تاریخ گزارش: ۱۵:۳۵ ۱۴۰۰/۶/۹
	شماره پرونده: 176942

Consultation

Lab No. 1400-1083

SPECIMEN :

One outside paraffin embedded block and one H & E stained slide labeled as "Left lower lobe, lung , wedge resection" for second opinion.

CLINICAL DATA :

-Cough & hemoptysis.

CT file:Multiple small opacity in both lungs are seen.

MACROSCOPIC EXAMINATION:

Received for consultation one outside prepared paraffin embedded block & one H&E stained slide labeled as P-0005-108 associated with corresponding pathology report from Arjmand Hospital Pathobiology laboratory, Kerman city diagnosed as " Bronchiolitis obliterans pneumonia (BOOP), No evidence of malignancy or granuloma is seen in this specimen".

MICROSCOPIC EXAMINATION :

Lung parenchyma with evidence of fresh & old hemorrhage (some intraalveolar hemosiderin laden macrophages) & presence of scattered nodules with hyalinization, sclerosis, calcification & ossification. Scatteredly some cellular nodules composed of epithelioid cells with intracytoplasmic vacuole , oval bland nuclei are also noted.In the IHC staining the mentioned cells are negative for AE1/AE3.CD34 is positive in some of the mentioned cells.CD31 is not avaiable in this center in this time.

DIAGNOSIS :

One outside paraffin embedded block and one H & E stained slide labeled as "Left lower lobe, lung , wedge resection" for second opinion:

-Compatible with pulmonary epithelioid hemangioendothelioma.

INTRODUCTION

Hemoptysis is the expectoration of blood from the lower respiratory tract.

The first step in evaluation is to ascertain whether the bleeding is coming from the respiratory tree or instead originating from the nasal cavities (i.e.,epistaxis) or the gastrointestinal tract (i.e., hematemesis) as the therapies for these etiologies will be significantly different.

Once established as hemoptysis, the exact nature of the expectoration is important as the term can be applied to blood-tinged phlegm, the pink frothy sputum of pulmonary edema, or frank blood.

Next steps include identifying the source and etiology of bleeding.

ANATOMY AND PHYSIOLOGY OF HEMOPTYSIS

Hemoptysis can arise from anywhere in the respiratory tract; from the glottis to the alveolus.

Most commonly, bleeding arises from the bronchi or medium sized airways, but a thorough evaluation of the entire respiratory tree is often necessary.

A unique feature of the lung that predisposes to hemoptysis of varied severity is its dual blood supply—the pulmonary and bronchial circulations.

Continuation

The former is a low-pressure system that is essential to gas exchange at the alveolar level; in contrast, the bronchial arteries originate from the aorta and are under systemic pressure.

The bronchial arteries supply the airways and have the ability to neovascularize tumors, dilate airways of bronchiectasis, and cavitary lesions.

Most hemoptysis is due to vessels in the bronchial circulation and is, therefore, under systemic pressure, making it more challenging to arrest the bleeding.

ETIOLOGY

Hemoptysis commonly results from :

infection,

malignancy,

or vascular disease;

however, the differential for bleeding from the respiratory tree is varied and broad.

Infections

Most blood-tinged sputum and small-volume hemoptysis is due to viral bronchitis.

Patients with chronic bronchitis are at risk for bacterial superinfection with organisms such as Streptococcus pneumoniae, Haemophilus influenzae, or Moraxella catarrhalis, increasing airway inflammation and potential for bleeding.

Similarly, patients with bronchiectasis are prone to hemoptysis with exacerbations of disease.

Continuation

Due to recurrent bacterial infection, bronchiectatic airways are dilated, inflamed, and highly vascular, supplied by the bronchial circulation.

In several case series, bronchiectasis is the leading cause of massive hemoptysis and subsequent death.

Tuberculosis had long been the most common cause of hemoptysis worldwide, but it is now surpassed in industrialized countries by bronchitis and bronchiectasis.

Continuation

In patients with tuberculosis, development of cavitary disease is frequently the source of bleeding but rarer complications such as the erosion of a pulmonary artery aneurysm into a preexisting cavity (i.e., Rasmussen's aneurysm) can also be the source.

Other infectious agents such as endemic fungi, Nocardia, and non-tuberculous mycobacteria can present as cavitary lung disease complicated by hemoptysis.

In addition, Aspergillus species can develop into mycetomas within preexisting cavities, with neovascularization to these inflamed spaces leading to bleeding.

Continuation

Pulmonary abscesses and necrotizing pneumonia can cause bleeding by devitalizing lung parenchyma. Common responsible organisms include Staphylococcus aureus, Klebsiella pneumoniae, and oral anaerobes.

Paragonimiasis can mimic tuberculosis and is another significant cause of hemoptysis seen globally; it is common in Southeast Asia and China, although cases have been reported in North America from raw crayfish ingestion. It should be considered as a cause of hemoptysis in recent immigrants from endemic areas.

Vascular

Hemoptysis commonly results from pulmonary edema due to elevated left ventricular end-diastolic pressure.

While the classic description of the sputum expectorated in pulmonary edema is “pink and frothy,” a spectrum of hemoptysis including frank blood can be seen.

A pulmonary embolism with parenchymal infarction can present with hemoptysis, although most pulmonary emboli do not cause hemoptysis and will present with other signs and symptoms.

An ectatic vessel in an airway or a pulmonary arteriovenous malformation can be a source of bleeding.

Continuation

While rare, rupture of an aortobronchial fistula can result in massive bleeding and sudden death; these fistulae arise in the setting of aortic pathology such as aneurysm or pseudoaneurysm and can cause small bleeding episodes that herald massive hemoptysis.

Diffuse alveolar hemorrhage (DAH), despite causing significant bleeding into the lung parenchyma, uncommonly results in hemoptysis.

A range of insults cause DAH, including immune-mediated capillaritis from diseases such as systemic lupus erythematosus, toxicity from cocaine and other inhalants, and stem cell transplantation.

Continuation

DAH more commonly presents with diffuse ground glass opacities on imaging and anemia, so the absence of hemoptysis should not exclude the diagnosis.

The so-called “pulmonary-renal” syndromes, including granulomatosis with polyangiitis and anti-glomerular basement membrane disease, may lead to both hemoptysis and hematuria (though one manifestation may be present without the other).

Malignancy

Bronchogenic carcinoma of any histology is a common cause of hemoptysis (both massive and non-massive) in modern published series.

Hemoptysis often indicates airway involvement of the tumor and can be a presenting symptom of carcinoid tumors, vascular lesions that frequently arise in the proximal airways.

Small cell and squamous cell carcinomas are frequently central in nature and more likely to erode into major pulmonary vessels, resulting in massive hemoptysis.

Pulmonary metastases from distant tumors (e.g.,melanoma, sarcoma, adenocarcinomas of the breast and colon) can also cause bleeding.

Kaposi’s sarcoma, seen in advanced acquired immunodeficiency syndrome, is very vascular and can develop anywhere along the respiratory tract, from the bronchi to the oral cavity.

Mechanical and Other Causes

In addition to infection, vascular disease, and malignancy, other insults to the pulmonary system can cause hemoptysis.

Pulmonary endometriosis causes cyclical bleeding known as catamenial hemoptysis.

Foreign body aspiration can lead to airway irritation and bleeding.

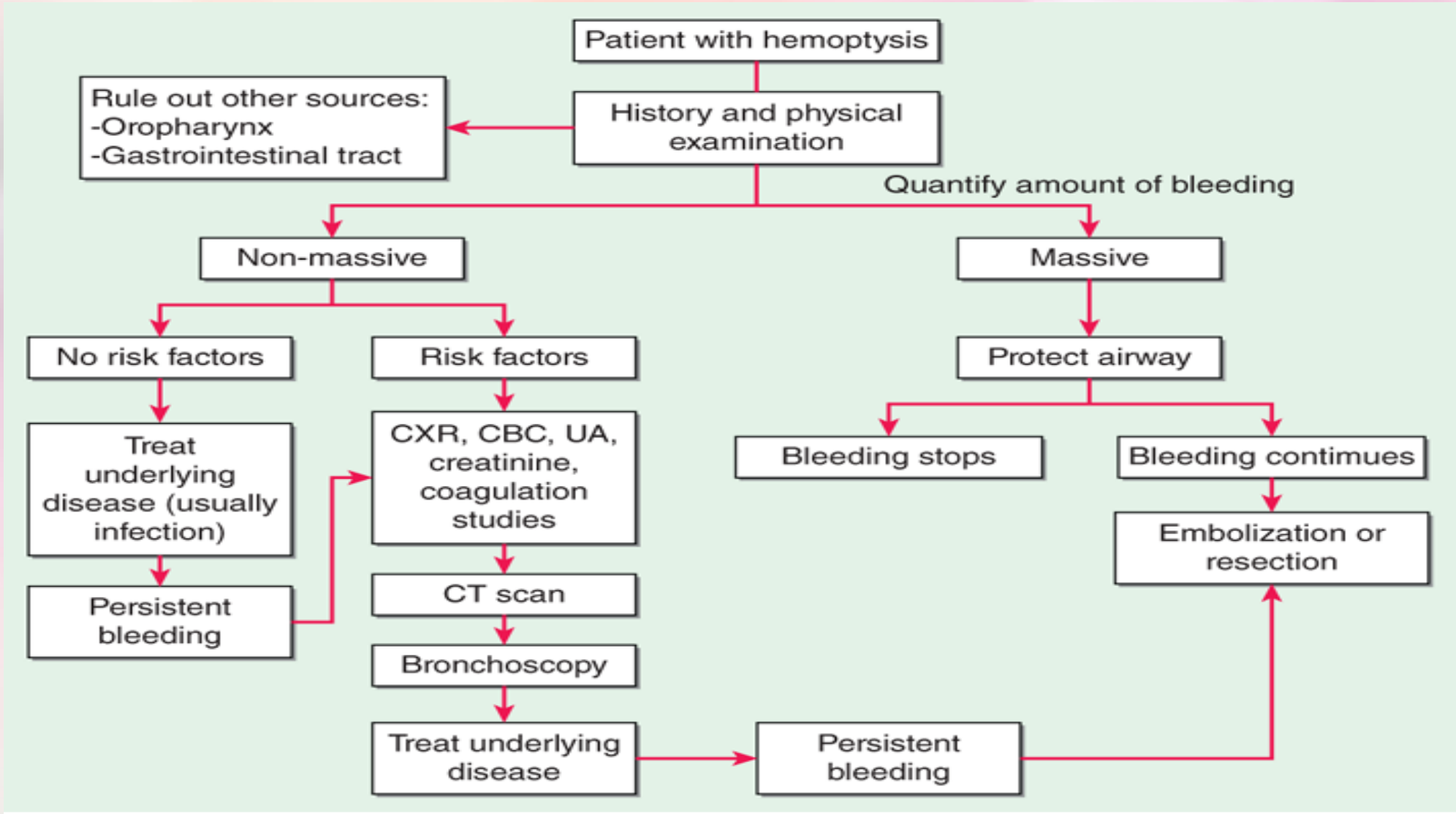
Diagnostic and therapeutic procedures are also potential offenders: pulmonary vein stenosis can result from left atrial procedures, such as pulmonary vein isolation, and pulmonary artery catheters can lead to rupture of the pulmonary artery if the distal balloon is kept inflated.

Finally, in the setting of thrombocytopenia, coagulopathy, anticoagulation, or antiplatelet therapy, even minor insults can cause hemoptysis.

EVALUATION AND MANAGEMENT

The first step in evaluating hemoptysis is to determine the amount or severity of bleeding.

A patient’s description of the sputum (e.g., flecks of blood, pink-tinged, or frank blood or clot) is helpful if you cannot examine it.



Source: J.L. Jameson, A.S. Fauci, D.L. Kasper, S.L. Hauser, D.L. Longo, J. Loscalzo: Harrison's Principles of Internal Medicine, 20th Edition
Copyright © McGraw-Hill Education. All rights reserved.

Continuation

It is crucial to determine whether the amount of blood expectorated is massive; while there is no agreed-upon

volume, blood loss of 400 mL in 24 hours or 100–150 mL expectorated at one time are considered massive hemoptysis.

These numbers derive from the volume of the tracheobronchial tree (generally 100–200 mL).

This determination is clinically important as patients rarely die of exsanguination and, instead, are at risk of death due to asphyxiation from blood filling the airways and airspaces.

Most patients cannot describe the volume of their hemoptysis in mL, so using referents like cups (one U.S. cup is 236 mL) can be helpful.

Continuation

Fortunately, massive hemoptysis only accounts for 5–15% of cases of hemoptysis.

Careful history may point to the cause of hemoptysis. Fever, chills, or antecedent cough may suggest infection.

A history of smoking or unintentional weight loss makes malignancy more likely.

Patients should be asked about inhalational exposures.

A thorough medical history with careful attention to chronic pulmonary disease should be obtained, and the clinician should determine risk factors for malignancy and bronchiectatic lung disease (e.g., cystic fibrosis, sarcoidosis).

Physical Examination

Reviewing the vital signs is an important first step.

The presence of hypoxemia, tachypnea, and tachycardia should raise concern.

Clinicians should examine the nasal and oral cavities; observe the patient's breathing pattern, with careful attention to any respiratory distress; and auscultate the lungs.

Clubbing can suggest underlying lung disease such as lung cancer or cystic fibrosis.

Signs of bleeding diathesis (e.g., skin or mucosal ecchymoses and petechiae) or teleangectasias may suggest other predispositions to hemoptysis.

Diagnostic Studies

Initial studies should include measurement of a complete blood count to assess for infection, anemia, or thrombocytopenia, coagulation parameters, measurement of electrolytes and renal function, as well as urinalysis to exclude pulmonary-renal disease.

In patients with small, non-massive hemoptysis, outpatient evaluation can be pursued.

All patients with hemoptysis need chest imaging. A chest radiograph is usually obtained first, though it frequently does not localize bleeding and can appear normal.

In patients without risk factors for malignancy and with a normal chest radiograph, treating for bronchitis and ensuring close follow-up is a reasonable strategy, with further diagnostic workup if bleeding persists.

Continuation

In contrast, patients with risk factors for malignancy (i.e., age >40 or a smoking history) should undergo additional testing.

First, chest computed tomography (CT) should be obtained to better identify masses, bronchiectasis, and parenchymal lesions.

Following CT, a flexible bronchoscopy should be performed to exclude bronchogenic carcinoma unless imaging reveals a lesion that can be sampled without bronchoscopy.

Small case series show that patients with hemoptysis and unrevealing bronchoscopies have good outcomes.

Interventions

When the amount of hemoptysis is massive, there are three simultaneous goals:

first, protect the non-bleeding lung;

second, locate the site of bleeding;

and third, control the bleeding.

Protecting the airway and non-bleeding lung is paramount in the management of massive hemoptysis, since asphyxiation can happen quickly.

If the side of bleeding is known, the patient should be positioned with the bleeding side down, to use gravitational advantage to keep blood out of the non-bleeding lung.

Endotracheal intubation should be avoided unless truly necessary, since suctioning through an endotracheal tube is a less effective means of removing blood and clot than the cough reflex.

Continuation

If intubation is required, take steps to protect the non-bleeding lung either by selective intubation of one lung (i.e., the non-bleeding lung) or insertion of a double-lumen endotracheal tube.

Locating the bleeding site is sometimes obvious, but frequently it can be difficult to determine the source of hemoptysis.

A chest radiograph, if it shows new opacities, can be helpful in localizing the side or site of bleeding, though this test is not adequate by itself.

CT angiography helps by localizing active extravasation.

Flexible bronchoscopy may be useful to identify the side of bleeding (although it has only a 50% chance of locating the site).

Continuation

Experts do not agree on the timing of bronchoscopy, though in some cases—cystic fibrosis, for instance—bronchoscopy is not recommended because it may delay definitive management.

Finally, proceeding directly to angiography is also a reasonable strategy given that it has both diagnostic and therapeutic capabilities.

Controlling the bleeding during an episode of massive hemoptysis can be accomplished in one of three ways:

from the airway lumen, from the involved blood vessel, or by surgical resection of both airway and vessel involved.

Bronchoscopic measures are generally only temporizing: a flexible bronchoscope can be used to suction clot and insert a balloon catheter that occludes the involved airway.

Continuation

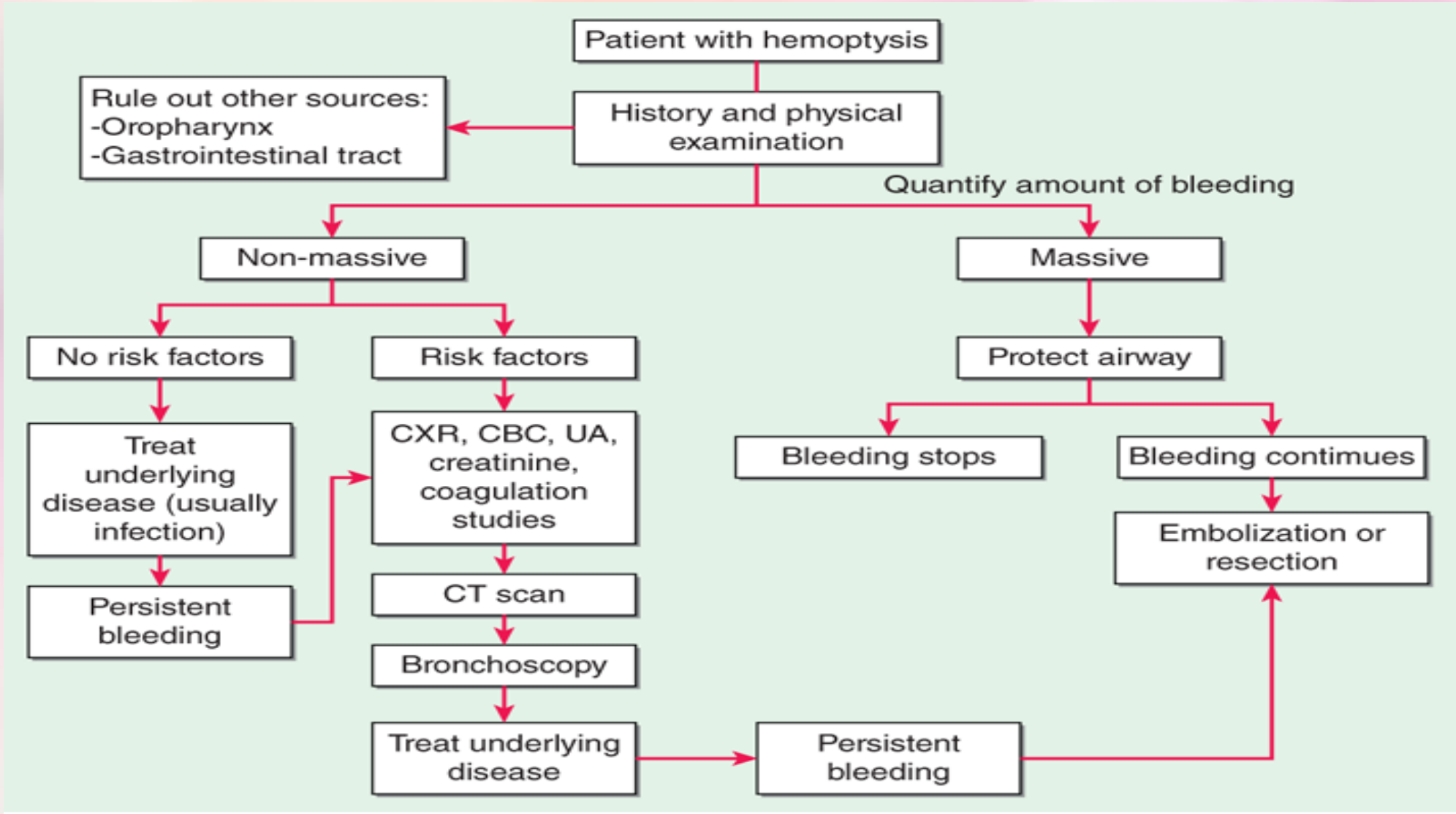
Rigid bronchoscopy, done by an interventional pulmonologist or thoracic surgeon, may allow therapeutic interventions of bleeding airway lesions such as photocoagulation and cautery.

Because most massive hemoptysis arises from the bronchial circulation, bronchial artery embolization is the procedure of choice for control of massive hemoptysis.

It is not without risk—embolization of the anterior spinal artery is a known complication—but is generally successful in the short term, with >80% success rate at controlling bleeding immediately, though bleeding can recur if the underlying disease (e.g., a mycetoma) is not treated.

Surgical resection has a high mortality rate (up to 15–40%) and should not be pursued unless initial measures have failed and bleeding is ongoing.

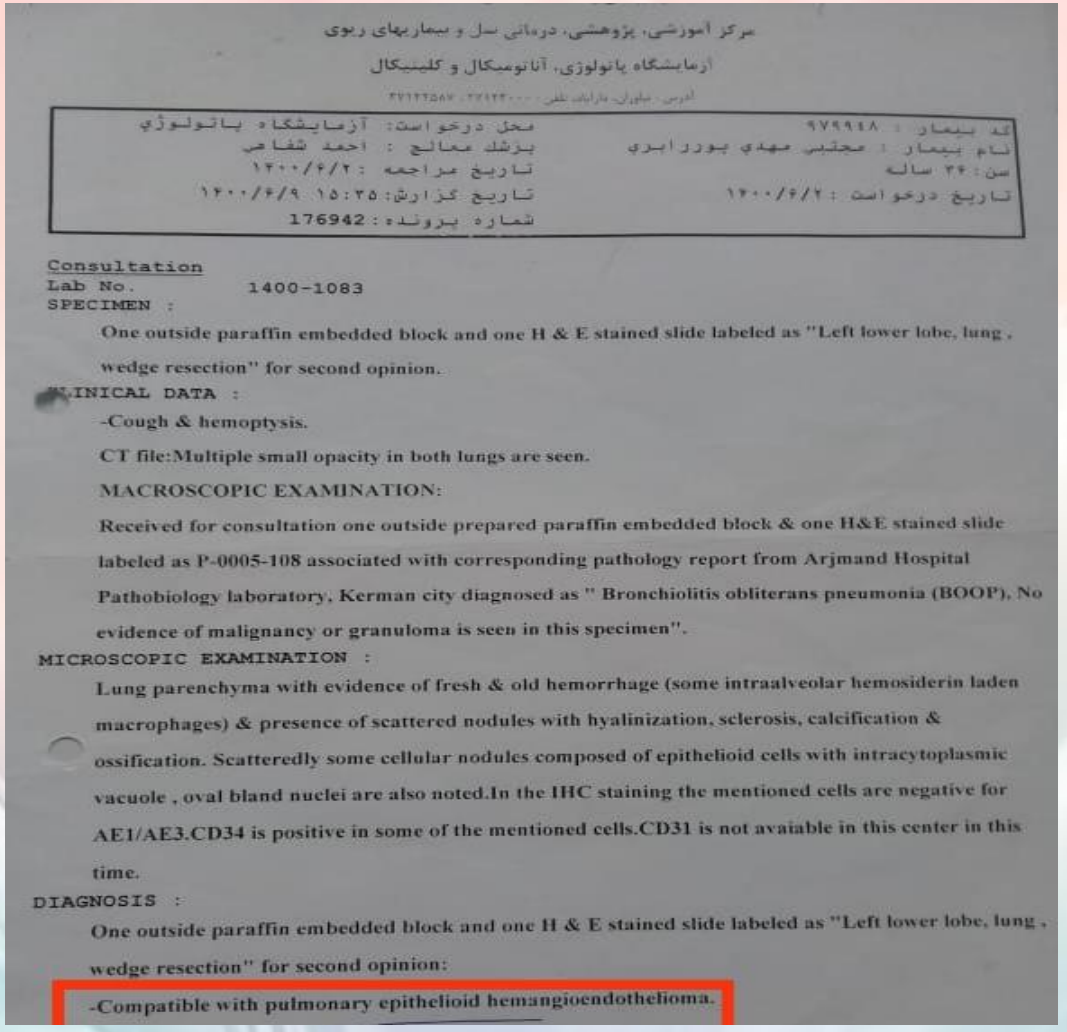
Ideal candidates for surgery have localized disease but otherwise normal lung parenchyma.



Source: J.L. Jameson, A.S. Fauci, D.L. Kasper, S.L. Hauser, D.L. Longo, J. Loscalzo: Harrison's Principles of Internal Medicine, 20th Edition
Copyright © McGraw-Hill Education. All rights reserved.

Open Lung Biopsy

- Pulmonary Epithelioid Hemangioendothelioma



مرکز آموزشی، پژوهشی، درمانی سل و بیماریهای ریوی

آزمایشگاه پاتولوژی، آناتومیکال و کلینیکال

کد بیمار : ۹۷۹۹۴۸	محل درخواست: آزمایشگاه پاتولوژی
نام بیمار : مجتبی مهدی پوررابری	پزشک معالج : اجمه شفاهر
سن: ۴۴ ساله	تاریخ مراجعه : ۱۴۰۰/۶/۲
تاریخ درخواست : ۱۴۰۰/۶/۲	تاریخ گزارش: ۱۵:۳۵ ۱۴۰۰/۶/۹
	شماره پرونده: 176942

Consultation

Lab No. 1400-1083

SPECIMEN :

One outside paraffin embedded block and one H & E stained slide labeled as "Left lower lobe, lung , wedge resection" for second opinion.

CLINICAL DATA :

-Cough & hemoptysis.

CT file:Multiple small opacity in both lungs are seen.

MACROSCOPIC EXAMINATION:

Received for consultation one outside prepared paraffin embedded block & one H&E stained slide labeled as P-0005-108 associated with corresponding pathology report from Arjmand Hospital Pathobiology laboratory, Kerman city diagnosed as " Bronchiolitis obliterans pneumonia (BOOP), No evidence of malignancy or granuloma is seen in this specimen".

MICROSCOPIC EXAMINATION :

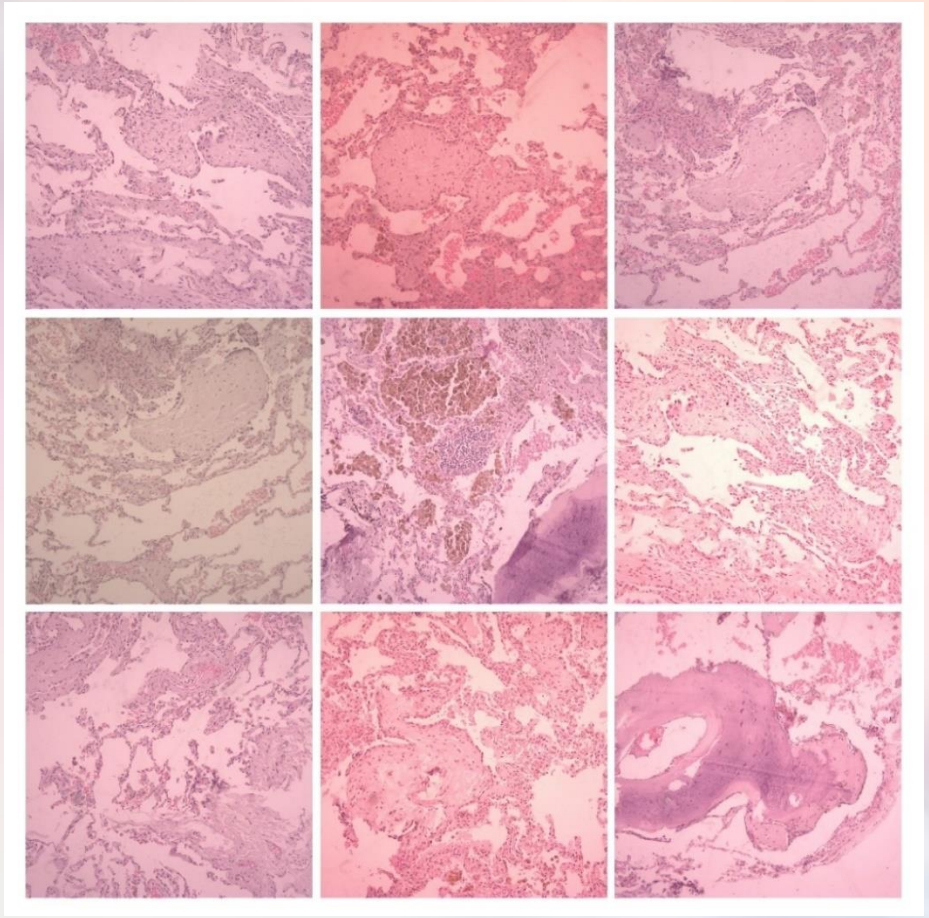
Lung parenchyma with evidence of fresh & old hemorrhage (some intraalveolar hemosiderin laden macrophages) & presence of scattered nodules with hyalinization, sclerosis, calcification & ossification. Scatteredly some cellular nodules composed of epithelioid cells with intracytoplasmic vacuole , oval bland nuclei are also noted.In the IHC staining the mentioned cells are negative for AE1/AE3.CD34 is positive in some of the mentioned cells.CD31 is not avaiable in this center in this time.

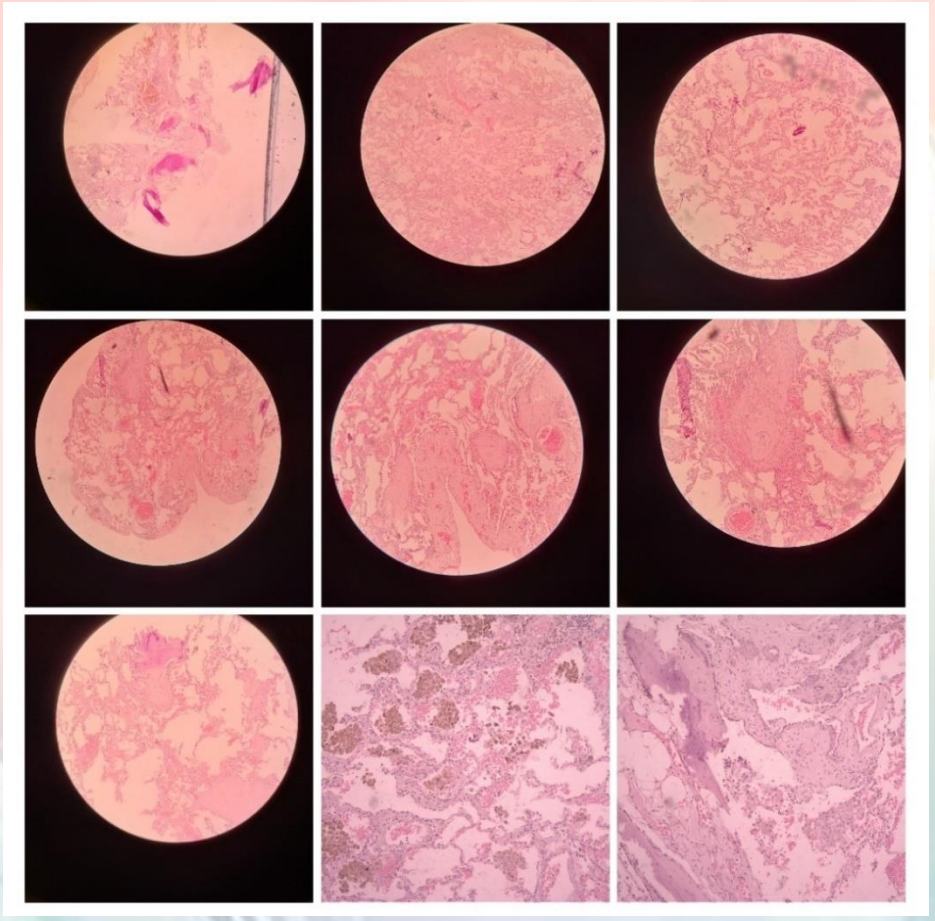
DIAGNOSIS :

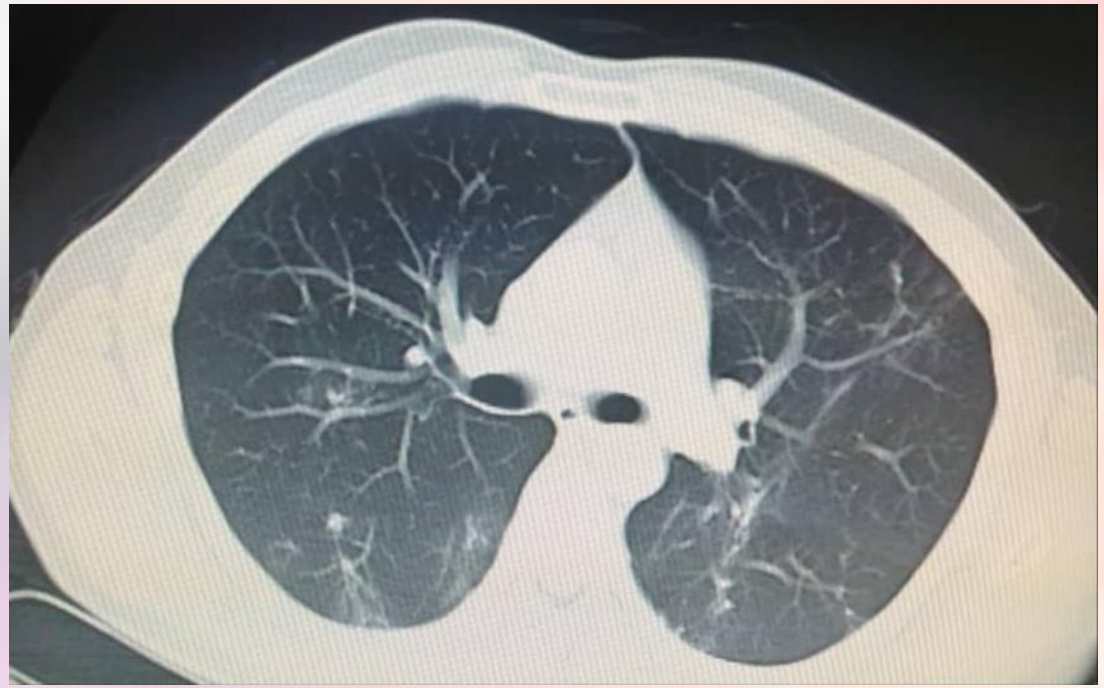
One outside paraffin embedded block and one H & E stained slide labeled as "Left lower lobe, lung , wedge resection" for second opinion:

-Compatible with pulmonary epithelioid hemangioendothelioma.

Pathology



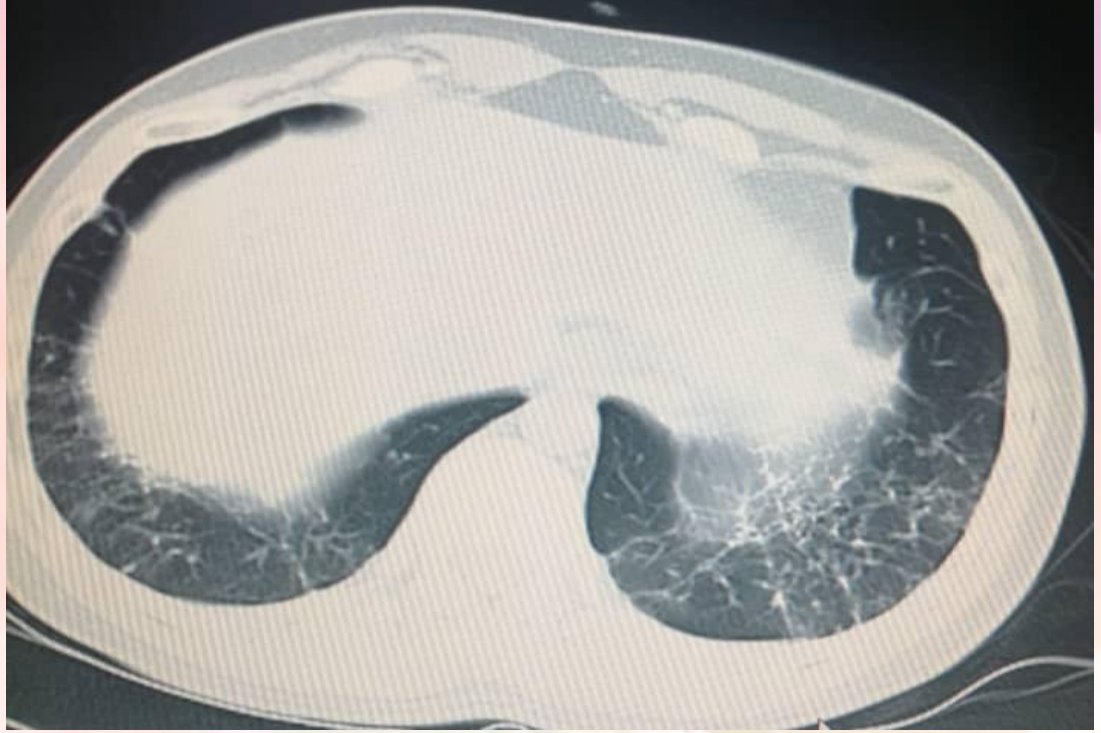




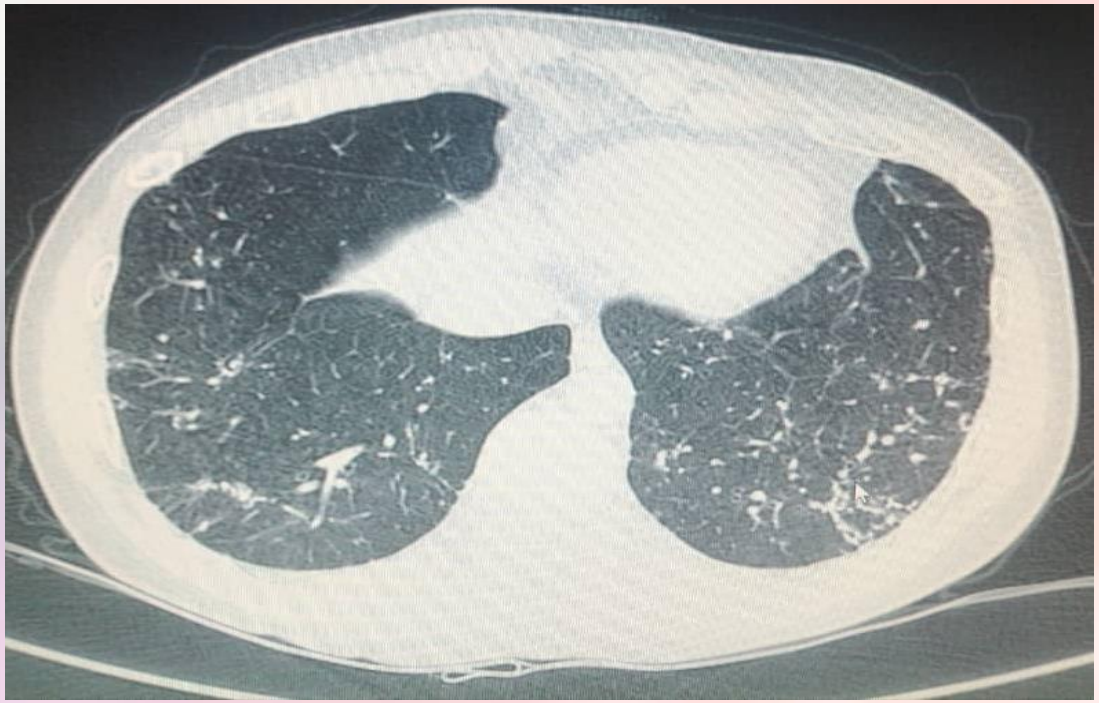


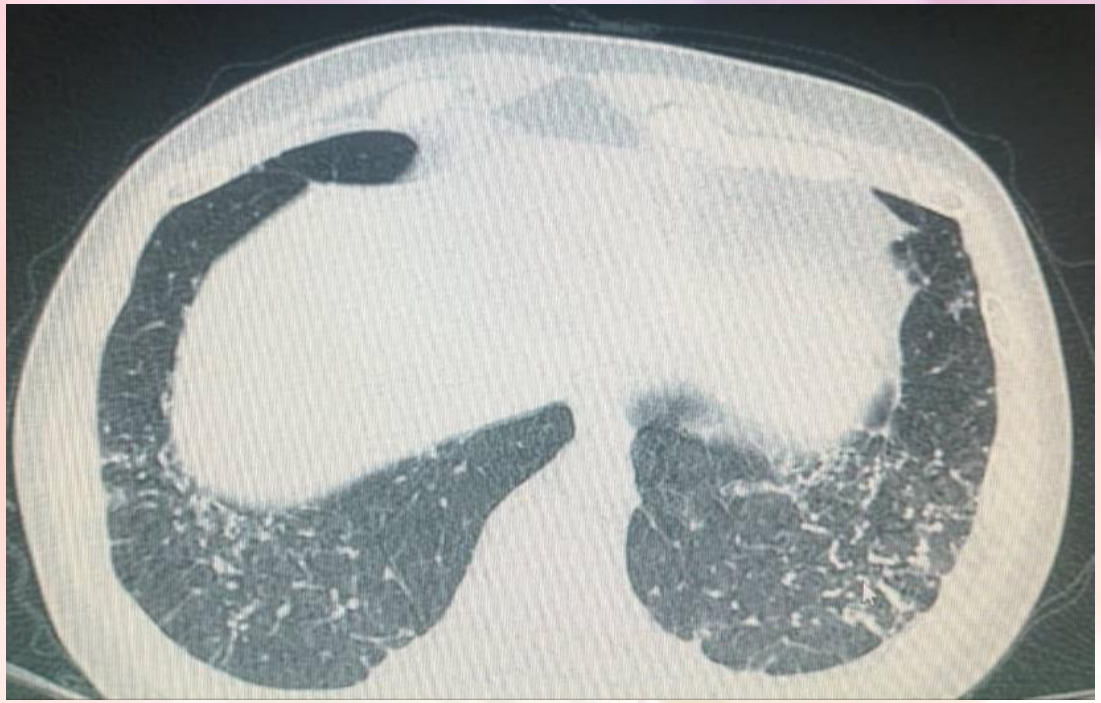


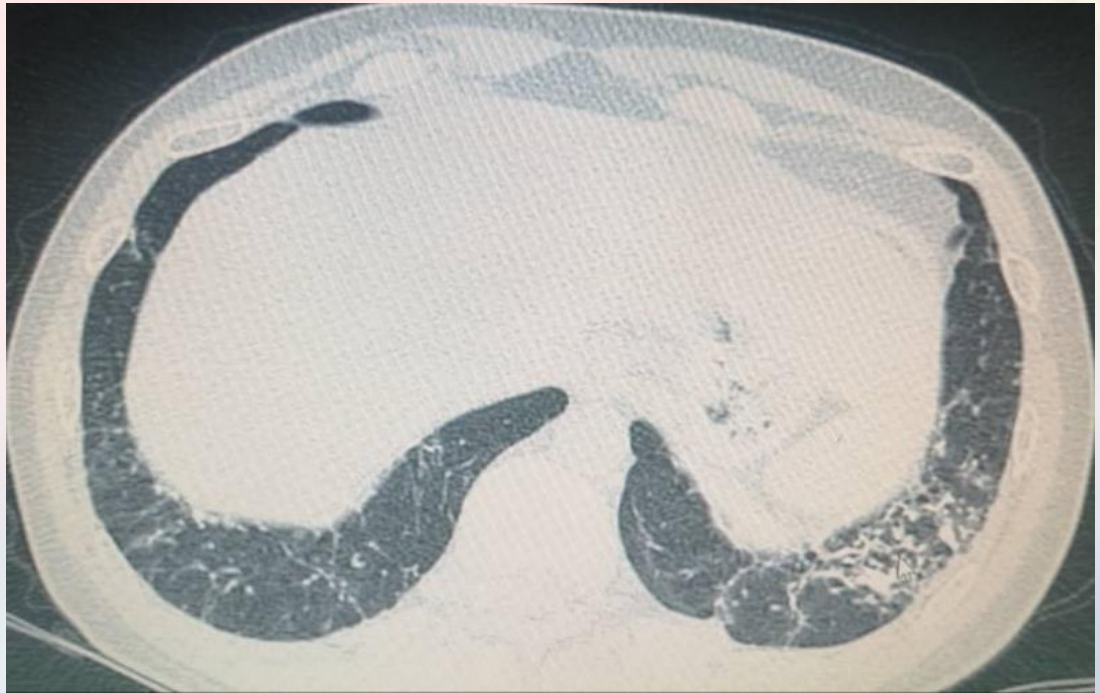












Pulmonary Epithelioid Hemangioendothelioma

Epithelioid hemangioendothelioma (EHE) is an uncommon neoplasm of vascular origin.

Typically occurring in young women, it is considered a tumor of borderline malignant potential.

We present a rare case of a young man with pulmonary EHE.

The tumor involved the mediastinum, endobronchial mucosa, pulmonary parenchyma and pleural surfaces.

Pulmonary EHE is an extremely rare illness that can mimic many rather common pulmonary disorders.

Awareness of this entity and a thorough work-up is the key to diagnosis.


THANK YOU
FOR YOUR
ATTENTION