Hemoptysis is defined as the expectoration of blood that originates from the lower respiratory tract. Pseudohemoptysis is the expectoration of blood from a source other than the lower respiratory tract such as the nares, oropharynx, larynx, or the gastrointestinal tract. Massive hemoptysis is defined as expectoration of blood exceeding 200 to 1,000 mL over a 24-hour period, with expectoration of greater than 600 mL in 24 hours being the most commonly used definition (1).

In practice, the rapidity of bleeding and ability to maintain a patent airway are critical factors; life-threatening hemoptysis can alternatively be defined as the amount of bleeding that compromises ventilation (2). Only 3% to 5% of patients with hemoptysis have a massive bleed, with the mortality rate ranging from 20% to as high as 80% in some case series (3–6). Most patients who die from massive hemoptysis do so from asphyxiation (3,7,8).

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ETIOLOGY OF MASSIVE HEMOPTYSIS

The causes of massive hemoptysis are listed in Table 145.1. Virtually all causes of hemoptysis may result in massive hemoptysis. Infection associated with bronchiectasis, tuberculosis, lung abscess, and necrotizing pneumonia are commonly responsible for the massive bleeding. Other common causes include bronchogenic carcinoma, mycetoma, invasive fungal diseases, chest trauma, cystic fibrosis, pulmonary infarction, and coagulopathy. Although massive hemoptysis is usually due to bleeding from the bronchial circulation, alveolar hemorrhage due to conditions such as Wegener granulomatosis and Goodpasture syndrome may occasionally cause massive hemoptysis (Table 145.2).

ANATOMIC SOURCES OF HEMOPTYSIS

The sources of lower respiratory tract bleeding include the pulmonary and bronchial circulations. The pulmonary circulation is a low-pressure circuit when normal pulmonary artery pressures are present. The pulmonary arteries supply blood to the pulmonary parenchyma. The bronchial circulation consists of the bronchial arteries, which originate from the aorta and have systemic arterial pressures, and the bronchial veins, which drain into the systemic veins to the right side of the heart. The bronchial and pulmonary circulations are normally interconnected by a bronchopulmonary anastomosis near the junction of the terminal and respiratory bronchioles. The bronchial arteries are the main source of blood to the airways, large branches of the pulmonary vessels, and supporting structures of the lung. The bronchial arteries feeding the proximal airways, such as the trachea and mainstem bronchi, drain into bronchial veins, which empty into the right side of the heart. Bronchial arteries serving the intrapulmonary airways and lung parenchyma drain through the bronchopulmonary anastomosis into the pulmonary veins, which empty into the left side of the heart. Angiographic studies of patients with active hemoptysis have demonstrated that the bronchial artery circulation
TABLE 145.1

POTENTIAL CAUSES OF MASSIVE HEMOPTYSIS

<table>
<thead>
<tr>
<th>Category</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>NEOPLASM</td>
<td>Bronchogenic cancer, Metastasis (parenchymal or endobronchial), Carcinoid, Leukemia</td>
</tr>
<tr>
<td>INFECTIOUS</td>
<td>Lung abscess, Bronchietasis, Necrotizing pneumonia, Fungal pneumonia, Septic pulmonary emboli, Mycetoma (aspergilloma)</td>
</tr>
<tr>
<td>PULMONARY</td>
<td>Bronchiectasis, Cystic fibrosis, Tuberculosis, Fungal pneumonia, Aspergilloma</td>
</tr>
<tr>
<td>CARDIAC/VASCULAR</td>
<td>Mitral stenosis, Pulmonary embolism/infarction, Arteriovenous malformation, Bronchoarterial fistula, Ruptured aortic aneurysm</td>
</tr>
<tr>
<td>CONGESTIVE HEART FAILURE</td>
<td>Pulmonary arteriovenous fistula</td>
</tr>
<tr>
<td>IATROGENIC/TRAUMATIC</td>
<td>Blunt or penetrating chest trauma, Bronchoscopy, Pulmonary artery rupture from pulmonary artery catheter, Endotracheal tube suctioning trauma</td>
</tr>
<tr>
<td>HEMATOLOGIC</td>
<td>Coagulopathy, Disseminated intravascular coagulation, Thrombocytopenia</td>
</tr>
<tr>
<td>DRUGS/TOXINS</td>
<td>Anticoagulants, Antiplatelet agents, Thrombolytic agents, Crack cocaine</td>
</tr>
</tbody>
</table>

is responsible for bleeding in approximately 90% of cases (9). Bronchial arteries arise directly or indirectly from the thoracic aorta at the level of the third through the eighth thoracic vertebrae, originating most commonly at the level of the fifth and sixth vertebrae.

The bronchopulmonary anastomoses may increase in size due to chronic inflammatory conditions such as bronchiectasis, cystic fibrosis, and tuberculosis (10). New collateral vessels from bronchial arteries or other intrathoracic systemic arteries may also develop in chronic inflammatory conditions.

TABLE 145.2

CAUSES OF ALVEOLAR HEMORRHAGE

<table>
<thead>
<tr>
<th>Category</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Goodpasture syndrome</td>
<td>Wegener granulomatosis, Microscopic polyangiitis, System lupus erythematosus, Mixed connective tissue disorder, Systemic sclerosis (scleroderma)</td>
</tr>
<tr>
<td>Vasculitis/collagen vascular disease</td>
<td>Rheumatoid arthritis, Henoch-Schonlein purpura, Mixed cryoglobulinemia, Behcet syndrome</td>
</tr>
<tr>
<td>Diffuse alveolar damage</td>
<td>Antiphospholipid syndrome, Idiopathic pulmonary hemosiderosis</td>
</tr>
<tr>
<td>Antiphospholipid syndrome</td>
<td>Hematopoietic stem cell/bone marrow transplantation</td>
</tr>
<tr>
<td>Coagulopathy</td>
<td>Mitral stenosis, Lymphangioleiomyomatosis</td>
</tr>
<tr>
<td>Drugs/toxins</td>
<td>Isocyanates, Trimelitic anhydride, D-penicillamine, Nitrofurantoin, All-trans retinoic acid, Crack cocaine</td>
</tr>
</tbody>
</table>

INITIAL EVALUATION

A detailed history and physical examination should be performed. Patients with a history of tuberculosis may have bleeding from a ruptured pulmonary artery aneurysm in the cavity lumen, known as a Rasmussen aneurysm, or by breakdown of bronchopulmonary anastomoses within the wall of old cavities (11). Bronchogenic carcinoma should be suspected in smokers older than 40 years of age. Repeated episodes of hemoptysis over months to years suggest bronchietasis or a carcinoid tumor. Chronic sputum production predating the hemoptysis implies a diagnosis of chronic bronchitis, bronchiectasis, or cystic fibrosis. Pulmonary embolism should be suspected in patients with a history of deep venous thrombosis or risk factors for pulmonary thromboembolism. A febrile illness with sputum production, night sweats, and weight loss suggests a lung abscess or tuberculosis. Excessive anticoagulation, thrombolytic therapy, and coagulopathy may also cause hemoptysis (12,13).

In children with hemoptysis, the most likely diagnoses are carcinoid tumors, vascular anomalies, and aspiration of foreign bodies (14,15). Alveolar hemorrhage should be suspected in patients with dyspnea, hypoxemia, and diffuse pulmonary infiltrates. The triad of upper airway disease, lower airway disease, and renal disease suggests Wegener granulomatosis (16). Goodpasture syndrome should be suspected in young men with alveolar hemorrhage and microscopic or macroscopic hematuria (17). Patients with a history of systemic lupus erythematosus may develop alveolar hemorrhage at any time during the course of their disease, and alveolar hemorrhage may be
the initial manifestation (18). Alveolar hemorrhage should be considered in patients with diffuse pulmonary infiltrates who have recently undergone hematopoietic stem cell or bone marrow transplantation (19). Although an uncommon cause of hemoptysis, a tracheoinnominate artery fistula is an important consideration in patients with tracheostomy (20,21). The peak incidence is between the first and second week, although hemorrhage can occur as early as 48 hours and as late as 18 months after the procedure. A sentinel self-limited bleed is observed in 35% to 50% of patients. Trauma from suctioning, particularly in the setting of abnormal coagulation, may also cause hemoptysis in patients with a tracheostomy tube or in those who are intubated with an endotracheal tube. The possibility of traumatic rupture of a pulmonary artery should be considered in patients with a pulmonary artery catheter in place (22,23).

Physical Examination

The physical examination may provide clues to the diagnosis of massive hemoptysis. A saddle nose deformity and/or septic perforation suggest Wegener granulomatosis. Stridor or unilateral wheezing indicates a possible laryngeal tumor, tracheobronchial tumor, or airway foreign body. Pulmonary embolism should be considered in patients with tachypnea, a pleural friction rub, and lower extremity edema. Diffuse rales on examination raise the possibility of diffuse alveolar hemorrhage, diffuse parenchymal lung disease, or cardiac disease as the cause of the hemoptysis. The presence of telangiectasias of the skin or mucous membranes suggests hereditary hemorrhagic telangiectasia or a connective tissue disease as the cause. Ecchymoses or petechiae suggest a hematologic abnormality or coagulopathy. Clubbing of the fingers may be a sign of a lung carcinoma, bronchiectasis, or cystic fibrosis. The finding of pulsation of the tracheostomy tube is of concern for the development of a tracheoinnominate fistula.

Laboratory Studies

Laboratory studies, including a complete blood count (CBC), coagulation studies, urinalysis, and chest radiograph, should be obtained in all patients. The CBC may suggest an infectious process or hematologic disorder as the cause of hemoptysis and indicates the need for blood transfusion. Coagulation studies may provide evidence for a hematologic disorder as the cause for the hemoptysis, or may identify a coagulopathy that is causing or contributing to the bleeding from another disease. Hematuria may be noted on urinalysis, which suggests the diagnosis of Goodpasture syndrome, Wegener granulomatosis, or another systemic vasculitis.

Chest Radiograph

The chest radiograph is an important study to identify the cause and side of bleeding. The chest radiograph may demonstrate abnormalities such as lung masses, cavitary lesions, atelectasis, focal infiltrates, and diffuse infiltrate. Single or multiple pulmonary cavities suggest neoplasms, tuberculosis, fungal disease, lung abscess, septic pulmonary emboli, parasitic infection, or Wegener granulomatosis as the cause for hemoptysis. The presence of a mass within a cavitary lesion indicates a possible mycetoma (aspergilloma). The appearance of a new air-fluid level in a cavity or infiltrate around a cavity is suggestive of the site of bleeding. A solitary pulmonary nodule may be an arteriovenous malformation. Diffuse pulmonary infiltrates suggest diffuse alveolar hemorrhage (Table 145.2), bleeding from coagulopathy, lung contusions from blunt chest trauma, hemorrhage with multiple areas of aspiration, or pulmonary edema with a cardiac cause for hemoptysis. Chest radiographs may be normal or nonlocalizing in 20% to 45% of patients (24,25).

Computed Tomography

The role of computed tomography (CT) in the management of massive hemoptysis is somewhat controversial. CT may demonstrate abnormalities that are not visible on the chest radiograph. It is helpful in the diagnosis of bronchiectasis (26), although abnormalities from bronchiectasis can usually be appreciated on the chest radiograph. CT with contrast may detect pulmonary emboli, thoracic aneurysms, or arteriovenous malformations. CT scans may also demonstrate cavitary with a surrounding infiltrate, the halo sign, which suggests a necrotizing infection such as aspergillosis or mucormycosis (27,28). Some authors have noted that CT scanning before bronchoscopy may increase the yield of bronchoscopy (29). In one retrospective study of 80 patients with large or massive hemoptysis, chest CT was superior to chest radiograph or bronchoscopy in determining the cause of bleeding and was similar to bronchoscopy in successfully localizing the site of bleeding (30). Some authors have argued that transport of the potentially unstable patient with massive hemoptysis may not be judicious; however, the patient should be adequately stabilized prior to obtaining a chest CT.

Angiography

Angiography can determine the site of bleeding in 90% to 95% of cases. However, in one case series, the routine use of diagnostic angiography provided a diagnosis not identified on bronchoscopy in only 4% of patients (31). Angiography can be helpful in detecting a pseudoaneurysm that has formed after healing of a pulmonary artery tear from pulmonary artery catheterization (32). As previously noted, the bronchial arteries and other collateral systemic arteries account for the source of bleeding in most cases of massive hemoptysis. Pulmonary angiography is usually performed only when there is suspicion for pulmonary aneurysms, arteriovenous malformations, and pulmonary embolism. Technetium-labeled red blood cell or collod studies rarely provided any information that is not obtained by bronchoscopy and chest CT. The use and timing of bronchoscopy will be discussed in a subsequent section.

Other Studies

Depending on the suspected causes of massive hemoptysis, additional studies may be indicated. For potential infections, sputum and bronchoscopic specimens should be sent for bacterial
cultures, fungal stains and cultures, viral cultures, acid-fast bacilli stains, and mycobacterial cultures. Bronchoalveolar lavage (BAL) specimens may also be sent for cytology, with special stains to evaluate for fungi, Pneumocystis, viruses, protozoa, and parasites. Bronchoscopic specimens should be obtained if a neoplasm is suspected. Echocardiography may be performed if a cardiac cause is possible. If diffuse alveolar hemorrhage syndromes are suspected, laboratory testing, including antiglomerular basement membrane antibody, antineutrophil cytoplasmic antibody, antinuclear antibody, rheumatoid factor, complement levels, cryoglobulins, rheumatoid factor, and antiphospholipid antibodies, should be performed depending on the causes that are being considered. Transbronchial lung biopsy, open lung biopsy, or kidney biopsy may be indicated in some cases of alveolar hemorrhage to establish a diagnosis.

MANAGEMENT OF MASSIVE HEMOPTYSIS

Airway Protection and Stabilization

Once the diagnosis of massive hemoptysis is established, the initial priorities are to protect the airway and stabilize the patient. In general, the patient with massive hemoptysis should be monitored in the ICU setting, even if intubation and mechanical ventilation are not required. Large-bore IV access should be established and supplemental oxygen provided. Blood should be drawn for a CBC, arterial blood gas analysis, coagulation studies, electrolytes, renal function tests, and liver function tests. The patient should be type and cross-matched for blood, studies, electrolytes, renal function tests, and liver function tests.

Airway patency must be ensured in patients with massive hemoptysis, as deaths from this process are predominantly due to asphyxiation. Most patients with ongoing massive hemoptysis will require intubation and mechanical ventilation, although select patients who are not hypoxemic and are able to keep the airway clear on their own may not require intubation. Although intubation generally preserves oxygenation and facilitates blood removal from the lower respiratory tract, the endotracheal tube (ET) can become obstructed by blood clots, leading to the inability to oxygenate and ventilate the patient. The largest possible ET should be inserted to allow the use of bronchoscopes with a 2.8 to 3.0 mm working channel for more effective suctioning and to allow for better ventilation with the bronchoscope in the airway for prolonged periods of time. In severe cases, the mainstem bronchus of the nonbleeding lung can be selectively intubated under bronchoscopic guidance to preserve oxygenation and ventilation from the normal lung.

Some authors have recommended the use of a double-lumen ET to isolate the normal lung and permit selective intubation. Although double-lumen endotracheal tubes have been used successfully in the airway management of massive hemoptysis, there are several potential pitfalls. First, placement of a double-lumen ET is difficult for less-experienced operators, particularly with a large amount of blood in the larynx and oropharynx. Second, the individual lumens of the ET are significantly smaller than a standard ET and are at significant risk of being occluded by blood and blood clots. Last, positioning of the double-lumen ET and subsequent bronchoscopic suctioning of the distal airways requires a small pediatric bronchoscope with working channels of 1.2 to 1.4 mm. Adequate suctioning of large amounts of blood and blood clots through such bronchoscopes is extremely problematic. In one series of 62 patients with massive hemoptysis, death occurred in 4 of 7 patients managed with a double-lumen ET due to loss of tube positioning and aspiration (35). In general, we do not recommend the use of double-lumen ETs for airway management in massive hemoptysis. As an alternative to selective mainstem bronchial intubation or intubation with a double-lumen ET, an ET that incorporates a bronchial blocker, such as the Univent tube, may be used.

Localization of Source and Cause of Hemoptysis

Once the patient is stabilized and airway patency is achieved, the source of bleeding should be localized as precisely as possible, and the cause of bleeding should be determined. Identification of the cause and location of the bleeding potentially allows for more specific therapy. Methods of localization include patient history, physical examination, chest radiograph, chest CT, bronchoscopy, and angiography. In one study of 105 patients with hemoptysis, patients themselves were able to localize the side of bleeding in 10% of cases but with an accuracy of 70% when able to do so (34). Localization by a physical examination performed by a physician was possible in 43% of patients. Chest radiographs were able to localize bleeding in 60% of cases. Bronchoscopy was accurate in localizing the source of bleeding in 86% of patients. In another study, 9 of 24 patients were able to accurately localize the side of their bleeding (35). Chest radiographs should be routinely obtained to help localize the source of bleeding and determine the cause. As discussed earlier, chest CT may provide additional information beyond the chest radiograph, and may be more accurate in localizing the bleeding and determining the cause, although concerns about transporting a potentially unstable patient out of the ICU exist (36,37). Bronchoscopy and angiography remain the modalities for localizing the source of hemoptysis and offer potential therapeutic intervention.

Early—rather than delayed—bronchoscopy should be performed to increase the likelihood of localizing the source of bleeding. Bronchoscopy performed within 48 hours of bleeding onset successfully localized bleeding in 34% to 91% of patients, depending on the case series, as compared to successful localization in 11% to 52% of patients if delayed bronchoscopy was performed (38). Bronchoscopy performed within 12 to 24 hours may provide an even higher yield. Besides flexible bronchoscopy should not be performed to establish a diagnosis of a tracheoarterial fistula such as a tracheostemominate fistula (39,40). Bronchoscopy may be performed in the patient with a tracheostomy tube and hemoptysis to exclude bleeding from suction trauma, tracheitis, granulation tissue, and lower respiratory tract disorders. If no other causes for hemoptysis...
can be found, or if the observation that anterior and downward pressure on the cannula at the level of the stoma site or over-inflation of the tracheostomy tube slows down the bleeding, a surgical consultation should be obtained. The tracheostomy balloon should not be deflated, and the tracheostomy tube should not be removed without protecting the airway below the tracheostomy tube. The patient should be brought to the operating room for further examination with preparations for surgical repair in place.

Bronchoscopic Therapies to Control Hemoptysis

Endobronchial tamponade via flexible bronchoscopy can prevent aspiration of blood into the contralateral lung and preserve gas exchange in patients with massive hemoptysis. Endobronchial tamponade can be achieved with a 4 French Fogarty balloon-tipped catheter. The catheter may be passed directly through the working channel of the bronchoscope, or the catheter can be grasped with biopsy forceps placed through the working channel of the bronchoscope prior to introduction into the airway of the bronchoscope and catheter. The catheter is held in place adjacent to the bronchoscope by the biopsy forceps, and both are then inserted as a unit into the airway. Care must be taken not to perforate the catheter or balloon by the forceps. The catheter tip is inserted into the bleeder bronchus with the Fogarty catheter remaining in position, thus providing endobronchial hemostasis (41–43). The catheter can safely remain in position until hemostasis is ensured by surgical resection of the bleeding segment or bronchial artery embolization. Right heart balloon catheters have been used in a similar fashion (44). A modified technique for placement of a balloon catheter has been described using a guidewire for insertion. A 0.035-inch soft-tipped guidewire is inserted through the working channel of the bronchoscope into the bleeding segment. The bronchoscope is withdrawn, leaving the guidewire in place. A balloon catheter is then inserted over the guidewire and placed under direct visualization after reintroduction of the bronchoscope (45). The use of endobronchial blockers developed for unilateral lung ventilation during surgery may hold promise for management of massive hemoptysis in tamponading bleeding and preventing contralateral aspiration of blood (46). The Arndt endobronchial blocker is placed through a standard ET and directed into place with a self-contained steering mechanism under bronchoscopic visualization. At this time, there is limited published experience with these blockers in the setting of massive hemoptysis, although the author has successfully used them for this application. The prolonged use of endobronchial blockers may cause mucosal ischemic injury and postobstructive pneumonia.

Additional bronchoscopic techniques may be useful as temporary measures in patients with massive hemoptysis. Bronchoscopically administered topically applied, such as iced sterile saline lavage or topical 1:10,000 or 1:20,000 epinephrine solution, may be helpful (47). Direct application of a solution of thrombin or a fibrinogen-thrombin combination solution has been used (48). The use of bronchoscopic-guided topical hemostatic tamponade therapy using oxidized regenerated cellulose mesh has recently been described (49). Although anecdotal, the author has had success with topical application of a sodium bicarbonate solution.

For patients who have hemoptysis due to endobronchial lesions, particularly endobronchial tumors, hemostasis may be achieved with the use of neodymium-yttrium-aluminum-garnet (Nd:YAG) laser photothermolysis, electrocautery, or cryotherapy via the bronchoscope.

Angiography and Embolization

Angiography can identify the bleeding site in more than 90% of cases. As noted, the bronchial arteries are the most frequent source of bleeding in massive hemoptysis. In some cases, systemic vessels other than the bronchial arteries can be the source of bleeding (50). The pulmonary arteries may be the source for massive hemoptysis in 8% to 10% of cases (9). Visualization of extravasated dye from a vessel is relatively uncommon. Signs that suggest a particular vessel is the source of bleeding include vessel tortuosity, increased vessel diameter, and aneurysmal dilatation.

Bronchial artery embolization is considered the most effective nonsurgical modality for treatment of massive hemoptysis. The immediate success rates from bronchial artery embolization range from 51% to 90.0% (3,51–65). Embolization has been performed with Gelfoam, polyurethane particles, polyvinyl alcohol particles, and vascular coils. Sclerosing agents may cause subsequent lung necrosis and should be avoided. Recurrence of bleeding, although usually nonmassive, has been noted in 16% to 46% of patients (51,52). Repeat embolization may be required in some patients (57,60,62,66). Complications include chest pain, fever, vessel perforation and intimal tears, and embolization of material to mesenteric and extremity arteries. The most serious complication is embolization of the anterior spinal artery, which may arise from the bronchial artery, with subsequent spinal artery infarction and paraparesis. The risk of this occurrence is less than 1%.

Rupture of the Pulmonary Artery

The pulmonary artery may potentially be ruptured from right heart catheterization. This complication should be suspected in patients who develop hemoptysis with a pulmonary artery catheter in place. Balloon tamponade and contralateral selective intubation should be performed (67). The catheter should be withdrawn 5 cm with the balloon deflated, and the balloon is then inflated with 2 mL of airway and allowed to float back into the ruptured vessel to occlude it. Patients who stop bleeding should undergo angiographic evaluation to localize the tear and identify the formation of a pseudoaneurysm (32,68). If a pseudoaneurysm is identified, embolization of the affected vessel should be considered to prevent subsequent hemorrhage.
Surgery

Emergency surgery for control of massive hemoptysis is performed less often due to the advent of bronchial artery embolization. Mortality rates for surgical management of massive hemoptysis range from 1% to 50% (3,69–74). Surgical resection of the source of bleeding offers definitive treatment as long as the lesion can be completely resected and the patient is able to tolerate resectional surgery. It is often difficult to accurately determine if these patients will be able to tolerate surgery, as they are often too ill to undergo pulmonary function tests, or are intubated and thus unable to perform pulmonary function tests. Surgical resection may be considered in patients when bronchial artery embolization is unavailable, if bleeding continues despite embolization, or if the cause of the hemoptysis is unlikely to be controlled with embolization.

Diffuse Alveolar Hemorrhage

Patients with diffuse alveolar hemorrhage syndromes are not candidates for bronchial artery embolization or surgery. Treatment for these groups of patients is pharmacologic. Corticosteroids are typically used and are effective for a wide range of the alveolar hemorrhage syndromes (75). Doses of 1 to 2 mg/kg per day of methylprednisolone have been most commonly used. For life-threatening alveolar hemorrhage, initial doses of 500 to 1,000 mg per day of methylprednisolone have been recommended. For Goodpasture disease, Wegener granulomatosis, and other vasculitides, adjunctive cytotoxic therapy or plasmapheresis may be considered.

References

Section XIII: Respiratory Disorders


