

Hemoptysis in Children: Single Center Experience

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ABSTRACT

Objective: To review the causes of hemoptysis in children presenting to the respiratory department at King Hussein Medical Center.

Methods: A retrospective chart review of all children who presented with hemoptysis was conducted from 1st April 2002 to 1st April 2012. Diagnosis of pulmonary hemorrhage was based on radiological and bronchoscopic findings. Demographic data, age at presentation, number of attacks, and presence of another diagnosis were recorded. Radiological and laboratory data were included as well.

Results: A total of 60 children with 68 episodes of hemoptysis were reviewed. Diffuse pulmonary hemorrhage was seen in five patients, three of them had idiopathic pulmonary hemorrhage, one had celiac disease and another had Wagener disease. Of the 55 patients with localized pulmonary hemorrhage the most common cause was cystic fibrosis (30%) and congenital heart disease (27%), followed by pulmonary arteriovenous malformation (8%), ruptured hydatid cysts (7.3%) and retained foreign body (5.4%). Fifty one out of the 55 (92.7%) with localized hemorrhage had localized patch on the chest x-ray; 40% had previous x-ray within the previous two years showing the same radiological patch at the time of hemoptysis episodes. Massive hemoptysis was found in three patients. Diagnostic flexible bronchoscopy was done in all patients within one week of the episode of hemoptysis. High resolution chest CT-scan was done for all patients with the diffuse type. Dynamic chest CT-scan was done in 50% of patients with localized hemorrhage. Embolization was done in five patients with AV malformation and two patients with cystic fibrosis. Surgical lobectomy was done in those with sequestration, hydatid cyst and one patient with foreign body and another one with localised bronchiectasis.

Conclusion: Hemoptysis is not uncommon in children. As an entity it should always be thoroughly investigated. Etiology differs according to age. Idiopathic pulmonary hemorrhage still constitutes a major group.

Key words: Hemoptysis, Hemosiderin-laden macrophages, Idiopathic pulmonary hemorrhage.

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Introduction

Pulmonary hemorrhage in children is rather uncommon.⁽¹⁾ It can be either diffuse alveolar hemorrhage (DAH) or localized.⁽¹⁾ The former is

characterized by hemoptysis, dyspnea and pulmonary infiltrates on chest radiography with variable degree of anemia.⁽¹⁾

Hematemesis in children can be mistaken for

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hemoptysis.^(1,2) The accurate incidence in children is difficult to determine.⁽²⁾ Thus we reviewed children who presented with pulmonary hemorrhage to the pediatric pulmonology department in King Hussein Medical Center (KHMC).

Method

All children presenting with hemoptysis to KHMC who were at regular follow up at Queen Rania Al-Abdullah Hospital for Children from 1st April 2002 to 1st April 2012 were retrospectively reviewed. Demographic data, age, presentation, medical history, number of episodes and physical findings were recorded. Family history of chronic lung disease, tuberculosis, bleeding and renal disorders was noted. All patients had a chest X-ray done. Further radiological testing was done according to individual case. Laboratory tests were also included. Diagnostic flexible bronchoscopy was done for all patients within one week of presentation.

Patients between one month and 18 years of age were included. Patients were classified into three groups according to age (<5 years, 5-10 years and >10 years). Those with identified nasal bleeding, gastrointestinal bleeding, trauma, bleeding tendency and patients in the intensive care unit were excluded.

Pulmonary hemorrhage was defined as diffuse or localized based on radiological and bronchoscopy findings. Bronchoscopic diagnosis was made if active lower respiratory tract bleeding was found and/or there were hemosiderin-laden macrophages in the lavage (>30%). The amount of hemoptysis was estimated in 24 hours: massive bleeding was defined as acute bleeding more than 240ml/day or recurrent bleeds of substantial amount (>100ml/day).

Ethical committee approval at Royal Medical Services was obtained

Results

A total of 60 children, 58% (n=35) males and 42% (n=25) females, with 68 recorded episodes of hemoptysis were reviewed. Diffuse pulmonary hemorrhage was seen in only five patients, three of whom had idiopathic pulmonary hemorrhage (IPH), one adolescent had celiac disease and another had Wagener disease. Of the 55 patients

with localized pulmonary hemorrhage, the most common was cystic fibrosis (CF) (n=18, 30%) and congenital heart disease (CHD) (n=16, 27%), followed by pulmonary arteriovenous malformation (PAVM) (n=5, 8%) as shown in Table I.

Table I: Etiology of pulmonary hemorrhage in children with hemoptysis attending the pulmonology department

CF	18 (30%)
CHD	16 (27%)
PAVM	5 (8%)
Localized bronchiectasis	5 (8%)
Ruptured hydatid cyst	4 (7%)
IPH	4 (7%)
Foreign body	3 (5%)
Sequestration	2 (3%)
Pneumonia	2 (3%)
Wagener disease	1 (2%)

Table II: Etiology of pulmonary hemorrhage according to age

	1-5 years	5-10 years	10-18 years
IPH	3	0	1
CF	0	4	14
CHD	8	3	5
PAVM	1	3	1
Sequestration	0	1	1
FB	2	1	0
Hydatid cyst	0	2	2
Pneumonia	0	1	1
Localised bronchiectasis	0	2	3
Wagener disease	0	0	1
Total	14	17	29

CHD was more common in those younger than five years of age (Table II). Localized bronchiectasis was seen also in 8% (n=5) while ruptured hydatid cysts in 7% (n=4) and retained foreign body in 5% (n=3). Fifty one out of the 55 (92.7%) with localized hemorrhage had localized patch on the chest x-ray, 81.8% had cough, 54.5% had fever, 50% of them received antibiotic in the last 6 months. Forty percent had previous x-ray done within the previous two years showing the same radiological patch at the time of hemoptysis episodes.

Massive hemoptysis was found in three patients, one with CF, another with PAVM and one patient with sequestration. Diagnostic flexible bronchoscopy was done in all patients within one week of the episode of hemoptysis. Lobar localization of hemorrhage was identified in 90%

of those with localized hemorrhage and almost correlated with radiological findings. High resolution chest ct-scan was done for all patients with the diffuse type, dynamic chest ct-scan was done in 50% of patients with localized hemorrhage, embolization was done seven times in all patients with PAVM and four times in two patients with CF. Surgical lobectomy was done in those with sequestration, hydatid cyst and one patient with foreign body and another one with localized bronchiectasis. The indication for lobectomy in the latter was recurrent infection and hospitalization.

None of the four patients with DAH showed positive results for connective tissue diseases, but one patient had positive celiac screen (tissue transaminase). Control of the respiratory manifestations was adequately achieved on gluten-free diet. Bronchoalveolar lavage was diagnostic in three of them showing more than 30% hemosiderin-laden macrophages, but one was diagnosed by transbronchial biopsy. The other one with diffuse hemorrhage was already known with Wegener disease.

Blood transfusions were required in six patients, one with diffuse hemorrhage, one with sequestration and one PAVM, one with cystic fibrosis and two with CHD. Only the latter patient with the CHD died due to the pulmonary hemorrhage.

Discussion

Hemoptysis in children is a rare presentation of pulmonary disease.^(1,2) It can be massive and life threatening and thus worrisome to the patients, family and physicians as well.^(1,2) It should be differentiated from nasal or post nasal bleeding.^(1,2) Clinical onset can vary from acute fulminant bleeding to chronic cough, fatigue or only anemia.⁽³⁾ Adults have more pronounced respiratory symptoms while in children failure to thrive and anemia can be the only manifestations; thus there can be a delay in diagnosis.^(2,3)

There are no studies indicating the exact incidence of hemoptysis in children. We reviewed all children who attended the pulmonology clinic with hemoptysis.

The etiology of hemoptysis in children may vary according to race, age, region, and research methodology.^(2,3) To the best of our knowledge this is the first report on the etiology of

hemoptysis in Jordanian children. Etiology of hemoptysis in children is different than adults.⁽³⁾ IPH is recognized when the workup fails to reveal an underlying cause.⁽⁴⁾ In as many as 20% of patients, no diagnosis is found.⁽⁴⁾ Incidence has been reported in some populations to be 0.24-1.25 cases per million.^(4,5) IPH was found in four (7%) patients in our study, three of them were below the age of five years and had no serological or other laboratory tests suggestive of milk allergy or pulmonary-renal syndrome. IPH might cause severe and recurrent hemoptysis requiring transfusion, usually affecting children less than six years of age.⁽⁶⁾ Three children in our study had recurrent hemoptysis when immunosuppressive therapy was discontinued or tapered. Such children should be kept under continuous follow up as an underlying disease may unveil itself with time^(5,6) Few cases were reported to develop systemic vasculitis eight years after initial presentation.⁽⁷⁾ In the cohort reported by Sim *et al.* Heiner syndrome was found in four and IPH in one affecting the same age groups as ours.⁽⁷⁾ The one adolescent in our IPH group had positive serology for celiac disease. Immunological mechanism is suggested to play a role in both celiac disease and IPH.⁽⁸⁻¹⁰⁾ Bronchoalveolar lavage showing more than 30% hemosiderin-laden macrophages is diagnostic for IPH.⁽¹¹⁾ Identification of hemosiderin-laden macrophages in bronchoalveolar lavage however occurs after 72 hours and remains detectable till 4-8 weeks.^(11,12) This indicates that the bleeding is from the lungs and the process is active.⁽¹²⁾

In this study cystic fibrosis was the most common cause of hemoptysis (30%). Coss-Bu *et al.* found that CF also is the most common yet their percentage was much higher (65%).⁽¹³⁾ This can be partially explained by the older age of their patients or the fact that we have less number of CF patients. Major hemoptysis occurs in 1% of CF patients.⁽¹⁴⁾ CF has become the major cause of hemoptysis in older children and young adults in the developed world.^(13,14) Minor hemoptysis is common during their disease course but major bleeds are believed to be secondary to erosions in the abnormally newly formed vessels in the area of bronchiectasis.⁽¹⁴⁾ Efrati *et al.* in Israel reported 9% of CF patients developing hemoptysis at some stage of their disease.⁽¹⁵⁾ The majority had minor bleeding.⁽¹⁵⁾ Some patients respond to

conservative supportive treatment; others require bronchial artery embolization or surgical ligation.^(15,16)

The 2nd most common cause in our study was CHD (27%). This is nearly the same as in the Coss-Bu *et al.* study if we exclude those patients above 20 year age from their study.^(13,17) On the other hand in the Sim *et al.* study congenital heart disease was found in only 17.5%.^(7,17) In other studies hemoptysis in children with CHD was seen in <5% and was fatal.⁽¹⁸⁾ None of the studies related hemoptysis to postoperative complications.

Other causes of hemoptysis quite common in our study and found more frequently than previous studies is AV malformation (9%). One of our patients was operated for tracheoesophageal fistula and had massive hemoptysis at the age of eight years. She responded well to pulmonary artery embolization. PAVM are rare clinical entities and mostly present in the 3rd and 4th decades of life.^(17,18) PAVM usually present with dyspnea and unexplained hypoxemia.⁽¹⁹⁾ Pulmonary hemorrhage can be fatal in such cases.^(19,20) Medical treatment is usually ineffective.⁽²⁰⁾ In Coss-Bu *et al.* and Sim *et al.* AV malformation was found only in one patient each.^(13,17,20)

Ruptured hydatid cyst was seen in 7% of our cohort. This might reflect the high incidence of hydatid disease in our Jordanian population. In fact only few reported cases were mentioned in previous studies,⁽²¹⁾ 10-20% of hydatid disease present in those <16 years of age.⁽²¹⁾ In our cases flexible bronchoscopy showed the white laminated membrane in the bronchus draining the cyst which is diagnostic for the disease.⁽²¹⁾ Flexible bronchoscopy was also diagnostic for retained foreign bodies, which is scarcely reported previously as a cause of hemoptysis in children although it was responsible for 5% of hemoptysis cases in our study. This might be due to lack of care, ignorance, poor appreciation of the child symptomatology and disproportion between clinical and radiological findings after the choking episodes. In Godfrey *et al.* study, foreign body of vegetable origin was found in only one patient among 17 children with hemoptysis.⁽²²⁾

Pneumonia per se was found in only two patients (3.3%) in our study, in 5.75% in Coss-bu

et al. study and in 5% in Sin *et al.* study.^(7,13,23) This contrasts other studies originating from otorhinolaryngology department that found acute lower respiratory tract infections as the most common cause for hemoptysis in children.⁽²³⁾ This may be that most of their referred cases are acutely ill previously healthy children. Tuberculosis has been frequently implicated in endemic areas⁽²⁴⁾ however we did not have any cases.

Interestingly 40% of those with localized hemorrhage had previous x-rays done in the previous years showing the same pneumonic finding on the x-ray taken at time of hemoptysis. Pneumonia mostly resolves radiologically in three months time.⁽²⁵⁾ In case of persistence, flexible bronchoscopy and/ or dynamic chest ct-scan should be done to examine for lung anomalies, foreign bodies or hydatid cyst.^(25,26) Bronchoscopy can be crucial in localization of the source of hemorrhage in addition to documentation of the hemosiderin-laden macrophages and culture results.^(26,27)

Conclusion

Hemoptysis is rather uncommon in children. It may indicate the presence of a serious underlying disease and may be fatal if not appropriately managed. As an entity it should always be thoroughly investigated. Etiology differs according to age. Idiopathic pulmonary hemorrhage still constitutes a major group.

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