The effect of ankaferd blood stopper used for massive haemoptysis in a patient with mounier-kuhn syndrome: A case report

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Abstract
Mounier-Kuhn Syndrome (MKS) is a rare disorder derived from the muscular and elastic tissue defects of the trachea and the main bronchial walls, characterized by tracheobronchomegaly. Patients may present with complaints of cough, phlegm, dyspnoea and haemoptysis. Haemoptysis may be minor and mixed with phlegm or it may be massive. Establishment of airway patency is a priority in the management of massive haemoptysis. Cold saline solution, diluted adrenaline or tranexamic acid may be administered via the endobronchial route to stop haemorrhage while establishing the airway patency. Ankaferd Blood Stopper (ABS) has a haemostatic property and can be locally administered to the airway. In this report, we aim to highlight the effects of ABS administered via an endobronchial route for emergency palliation of a patient with MKS presenting with massive haemoptysis.

Keywords: Ankaferd blood stopper, Haemoptysis, Bronchoscopy, Mounier-Kuhn syndrome.

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Introduction
Mounier-Kuhn Syndrome (MKS) is a disorder derived from atrophy or absence of the elastic fibres and smooth muscle cells in the main bronchi or the trachea. It is characterized by dilatation of the trachea and main bronchi as well as recurrent infections of the lower respiratory tract. Patients may be asymptomatic or can present with cough, phlegm, dyspnoea or haemoptysis. Haemoptysis is usually secondary to bronchiectasis and may be fatal when massive. Patency of the airway should be ensured initially in cases with massive haemoptysis. Rigid bronchoscopy or in intubated patients, flexible bronchoscopy may be used to establish airway patency. Cold saline solution, diluted adrenaline, or tranexamic acid maybe administered for haemostasis in both the conditions. Ankaferd Blood Stopper (ABS) is another topical agent that has haemostatic properties, it has been used successfully in controlling the haemorrhage following procedures such as dental interventions, otorhinolaryngologic operations such as tonsillectomy and urologic procedures. Utilization of this agent in interventions concerning bronchial system is quite new and the results have only been stated in case reports so far.

Case Report
A 25-year-old male patient was admitted to the Adnan Menderes University, Emergency Department with complaints of cough, phlegm and haemoptysis in December 2016. Patient described a medical history of frequent bronchitis since childhood, indicating previously made diagnosis of bronchiectasis at a clinic in 2012. Patient also stated that he had been tested for cystic fibrosis at that time; however, no diagnosis concerning a genetical disorder was established. Physical examination revealed a conscious, cooperating and an oriented individual with a slightly impaired general condition. His blood pressure was 125/70 mmHg, pulse rate was 102/min and respiratory rate was 18/min. Oxygen saturation (SpO2) measured from the fingertip was found to be 95% (96%-99%) in normal room air. Laboratory tests revealed C-reactive protein level to be 23 mg/L (0-5), white blood cell count 10200/mm3 (4000-1000),

Figure-1: High resolution computed tomography of chest shows abnormal dilatation of trachea in anteroposterior diameter.
haemoglobin 13.5 gr/dL (12-16 gr/dL), thrombocyte count 393000/mm3 (100000-450000/mm3) and erythrocyte sedimentation rate was 47 mm/h (0-20/h). Physical examination of the thorax revealed bilateral crackles in the mid and lower regions. High resolution computed tomography (CT) of the chest revealed tracheobronchomegaly (Figure-1) together with diffuse bronchiectasis in both the lungs (Figure-2). Blood and phlegm cultures were obtained during the admission and patient was started on oxygen, intravenous fluids, antibiotics (Ampicillin-Sulbactam 4g/day) and intravenous Tranexamic acid. The patient developed massive haemoptysis of approximately two drinking cups (400 ml) during the follow-up period. He was transferred to the intensive care unit (ICU) and was intubated with an initiation of mechanical ventilation. Fiberoptic flexible bronchoscopy was performed through the endotracheal tube and an active bleeding was observed in the basal segments of the left lower lobe. Twenty ml of iced saline lavage, 5 ml of diluted adrenaline (0.5 mg adrenaline in 5 ml saline) and 20ml of diluted tranexamic acid (250 mg tranexamic acid in 15 ml saline) were administered to this site through the bronchoscope. However, as the bleeding continued, blood was aspirated and after 60 seconds of the agents applied time, 2 ml of ABS was administered to the bleeding site. A brown-colour, web-like layer was formed immediately after the administration and the bleeding stopped. No other bleeding episode was observed during follow-up period and the patient was extubated after three days. Bronchial embolization procedure was performed by an Interventional Radiology Department following extubation. Phlegm sample obtained during the hospitalization process was stained using the Ehrlich Ziehl Neelsen method. Both this method and the nonspecific cultures revealed no pathogen supporting an infection. Patient had no further complaints and was discharged from the hospital. During the follow-up period, although massive haemoptysis didn’t occur again, patient was directed to a transplantation centre for lung transplantation evaluation due to a history of bronchiectasis.

Discussion

The diagnosis of MKS is usually made based on its radiological findings. More than 30 mm transverse diameter of the trachea and the diameters of right and left main bronchi being more than 24 mm and 23 mm, respectively are considered for the diagnosing MKS. In our case, the transverse and the antero-posterior diameters were measured as 26.3 mm and 33.3 mm, respectively (Figure-1) and the antero-posterior diameter of the right main bronchus was measured to be 24 mm. Tracheobronchomegaly is usually accompanied by bronchiectasis in MKS cases. In our case, varicose bronchiectasis was present bilaterally, particularly in the lower lobes (Figure-2). Haemoptysis may develop due to the bronchiectasis. Although, no definite algorithm is present concerning the management of haemoptysis, bronchoscopy has proved to be an effective strategy as it allows rapid assessment of the patient’s airway to visualize bleeding sites and performing interventions when required. Rigid or flexible bronchoscopy can be used in cases of massive haemoptysis. Rigid bronchoscopy is a procedure requiring anaesthesia; however, it is more beneficial in control the bleeding. Although, flexible bronchoscopy is easier to use in controlling the haemorrhage, its benefits are more limited when compared to rigid bronchoscopy. If flexible fiberoptic bronchoscopy is planned in patients with massive haemoptysis, it is necessary to perform endotracheal intubation. Thus, both the patency of the airway is provided and the physician can perform flexible bronchoscopy via the tube if necessary. After massive haemoptysis is diagnosed, we intubated the patients and then iced saline, diluted adrenaline and tranexamic acid was administered. There is no standard procedure for administration of adrenaline and tranexamic acid in case of airway bleeding. However, it can be administered 1 mg adrenaline diluted in up to 20mL of saline and 500 tranexamic acid mg diluted in up to 20mL of saline. In the event that bleeding continuous medication can be repeated 3 times. In our case due to an ongoing bleeding, 2 ml of ABS was given endobronchially through a flexible
bronchoscopy after 60 seconds of the agent's applied time. A brownish, web-like layer was formed at the bleeding site terminating the bleeding. ABS is a topical haemostatic agent. It is composed of a standardized mixture of Thymus vulgaris, Glycyrrhiza glabra, Vitis vinifera, Alpinia officinarum, and Urtica dioica plants. Each of these plants have been known to affect the endothelium, blood cells, angiogenesis, cellular proliferation, vascular dynamics, and/or cellular mediators.8 The mechanism of action of ABS concerning haemostasis is to form a specific protein-web for erythrocyte aggregation. ABS induces basic erythrocyte membrane proteins (ankyrin, spectrin, actin); thus leading to erythrocyte aggregation due to its effect on the fibrinogen gamma A.9 ABS manifests its preventive effects on the bleeding site by creating a specific protein-web within a short period of time such as 1 second. In a study conducted by Arslan et al.10 it was reported that ABS had successfully stopped the bleeding within seconds in a 62-year-old patient with an endobronchial tumour that had occurred due to a biopsy. An article published by Uzun et al.,5 reported a 40-year-old male patient with massive bleeding due to welding fumes; bleeding decreased after administration of a 2 ml of ABS through a fiberoptic bronchoscope and no recurrence was noted in the subsequent days. It was also reported that successful results were obtained with an endobronchial ABS administration in 23 out of 25 patients with haemoptysis hospitalized with various diagnoses.6

A few articles have reported successful haemostasis by an endobronchial delivery of the ABS for palliative treatment of the patients with bronchiectasis or other causes. In this report, we aim to share our experiences by emphasizing on the use of ABS which is a method amongst a limited number of treatment choices that can be used to achieve rapid palliation in acute episode of massive haemoptysis.

Conclusion

Efficacy of ABS in a patient with MKS having massive haemoptysis has been discussed in our case. Episode of massive haemoptysis can occur in a MKS patient and should be considered. After the establishment of a patent airway, haemostatic agents can be applied into the airway. ABS is also an agent that can be applied into the airway which has the benefit of preventing bleeding. This method can be used besides other haemostatic agents that are used in patients with massive haemoptysis.

To the best of our knowledge, this is the first case in which ABS has been applied into the airway of a MKS patient with massive haemoptysis in order to control the bleeding.

Limitation

ABS can decrease the severity of bleeding and facilitate the effect of ABS even if they didn't stop it completely.

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References


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