measure her total lung capacity (TLC) using a planimetric method as described by Harris et al* which demonstrated a loss in TLC from 4.4 L to 2.6 L in the period from 1966 to 1972. During this period, it is of interest that the appearance of the lung parenchyma on the chest roentgenogram remained normal. Arterial blood gas determinations done at rest and mild to moderate exercise in 1966 and 1970 gave normal results. These studies strongly suggest that the patient's abnormal pulmonary-function test results were due to musculoskeletal chest deformities rather than the more typical parenchymal fibrosis.

In summary, this unusual manifestation of a decrease in TLC and VC associated with marked bone resorption of the ribs seen on chest roentgenogram must be added to the possible pulmonary complications of PSS.

REFERENCES

Airway Obstruction due to Spontaneous Retropharyngeal Hemorrhage

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A patient is described with polycythemia vera who was taking anticoagulants and developed a spontaneous retropharyngeal hemorrhage after a violent sneezing episode. This progressed to tracheal compression with stridor after he had taken some aspirin for relief of the neck pain. An emergency tracheostomy was life saving.

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Spontaneous retropharyngeal hematoma causing respiratory distress and stridor is a relatively rare event, reported only as a complication of hemophilia.\textsuperscript{1,2} This is believed to be the first reported case of retropharyngeal and mediastinal hemorrhage causing respiratory compromise in a patient with polycythemia vera who was taking anticoagulants.

**CASE REPORT**

A 60-year-old white man was admitted to the UCLA Hospital in July, 1974 after an emergency tracheostomy in the emergency room to relieve an upper airway obstruction caused by a retropharyngeal hemorrhage.

The patient was known to have had polycythemia vera for five years, currently being treated by phlebotomy only. He had severe arteriosclerotic vascular disease with numerous ulcerations of the fingers and paronychia of the lower extremities, for which he underwent a sympathectomy in 1969 with little improvement. Several years prior to admission, he was placed on bishydroxycoumarin (Dicumarol) because of his vascular disease and one week prior to admission, the dose was increased from 300 to 350 mg weekly.

Three days prior to admission, the patient had a violent episode of sneezing, after which he experienced a sharp pain in the left side of his neck. Mild pain persisted for three days. On the morning of admission he had a sore throat and some dysphagia and took two aspirins to relieve the pain. He went to the emergency room at UCLA, where he noticed that his neck had begun to swell, that he was unable to swallow his secretions, and, soon thereafter, he began to have difficulty breathing. His neck was found to be swelling and mildly tender; ecchymosis of the posterior pharynx was noted. When the patient rapidly developed stridor, an emergency tracheostomy was performed.

\begin{figure}
\centering
\includegraphics[width=\textwidth]{image1.png}
\caption{Chest x-ray film on admission showing widening of the upper mediastinum and mild deviation of the trachea to the right.}
\end{figure}

\begin{figure}
\centering
\includegraphics[width=\textwidth]{image2.png}
\caption{X-ray film of neck showing retropharyngeal hematoma causing narrowing of the trachea. Tracheostomy tube is in place.}
\end{figure}

In the intensive care unit, physical examination revealed blood pressure 150/100 mm Hg; pulse rate 88; respiratory rate 26; temperature 38.0\textdegree C rectally. The neck was ecchymotic and massively swollen, measuring 47.5 cm in circumference, and a cuffed tracheostomy tube was in place. The rest of the physical examination was noncontributory except for a large spleen that was palpable 2 cm below the left costal margin and signs of peripheral vascular disease of the lower extremities. The hematocrit was 53 percent, hemoglobin 16.3 gm percent, and white blood cell count 27,000 with 95 percent polymorphonuclear leukocytes. The platelet count was 1.15 million. The partial thromboplastin time was 68 seconds with a control of 38 seconds and the prothrombin time was 28 seconds with a control of 11 seconds. The chest x-ray film showed anterior mediastinal widening with the trachea pushed slightly to the right (Fig 1). Anteroposterior and lateral projections of the cervical spine showed a soft tissue mass which extended from the occiput to at least the level of T\textsubscript{2}, and which effaced the hypopharynx, larynx, and trachea and displaced them forward (Fig 2).

The patient was given vitamin K (AquaMEPHYTON), 20 mg IV, ice packs were placed on his neck and he was started on aqueous penicillin. By the next morning, the neck circumference had increased to 52.5 cm. The patient was unable to breathe around the tracheostomy tube when the cuff was deflated. The hematocrit had dropped to 47.3 percent and the hemoglobin to 15.1 gm percent. Repeat x-ray films showed a
right lower lobe infiltrate and a probable increase in the size of the soft tissue mass which had extended further down the mediastinum. Fiberoptic bronchoscopy was performed through the tracheostomy to rule out the possibility of tracheal compression below the tip of the tracheostomy tube. A small non-obstructing bluish bulge was seen in the posterior tracheal wall below the end of the tracheostomy tube.

On the following day, the patient's hematocrit dropped further to 41.7 percent and the hemoglobin to 13.5 gm percent. However, his neck size and x-ray film appearance remained unchanged.

Over the next two weeks, the patient's neck size decreased and he was better able to breathe around the tracheostomy tube with the cuff deflated. Subsequent chest x-ray examinations revealed transient mild bilateral pleural effusions and a right lower lobe infiltrate felt to be secondary to aspiration. The soft tissue densities in the retropharyngeal and mediastinal areas gradually diminished in size. However, the patient continued to have difficulty swallowing and continued to aspirate as evidenced by the suctioning of purple material through the tracheostomy after the patient had taken grape juice by mouth. Because of this, 18 days after admission, the retropharyngeal space was drained through a left lateral neck incision and 150 ml of fluid and clot were evacuated.

The patient did well immediately postoperation, but three days later he passed a large melanotic stool and became hypotensive. At surgery, a posterior penetrating duodenal ulcer eroding the gastrointestinal artery and oozing from a small superficial gastric ulcer were found. Subtotal gastric resection, suture ligation of the bleeding duodenal ulcer and vagotomy were performed. After the operation the patient did well. The tracheostomy tube was removed and he was discharged seven days later with the tracheostomy site closing over and healing.

DISCUSSION

This patient illustrates another complication of polycythemia vera and/or anticoagulant therapy which, to our knowledge, has not been previously reported, i.e., compromise of the upper airway secondary to spontaneous retropharyngeal hemorrhage. It appears likely that the sneezing episode caused rupture of a small retropharyngeal blood vessel which caused hemorrhage that initially progressed slowly. Another possibility is that the bleeding began in the parapharyngeal space or one of the lateral neck spaces and then dissected into the retropharyngeal area. However, three days later when the patient became symptomatic and took some aspirin for relief of his symptoms, the hemorrhage rapidly progressed to a life-threatening event necessitating an emergency tracheostomy.

Retropharyngeal hematomas are known to occur after surgery of the neck and after external trauma. Although small hematomas occasionally occur following carotid angiography, only 1 to 2 percent of these become large enough to compromise the patient's airway; their frequency and location are related to the number and location of unsuccessful puncture attempts.

Retropharyngeal hematomas also occur secondary to infection of the pharyngo-maxillary space with erosion of an artery. Of 237 patients with an abscess of the pharyngo-maxillary space, Baily reported that 76 died from the erosion of a branch of the carotid artery.

There have also been two reported cases of retropharyngeal and mediastinal hemorrhage following transbrachial retrograde catheterization of the left ventricle while the patients were on anticoagulants. Spontaneous retropharyngeal hemorrhage without trauma or surgical intervention has been reported only with hemophilia.

In this case of spontaneous hemorrhage without naso-maxillary infection, there are three factors that may have contributed to the occurrence and/or progression of the hemorrhage to a life-threatening event. These are the presence of polycythemia vera, anticoagulation and aspirin administration. The most common complications of polycythemia vera in its early stage is reported to be hemorrhage and thrombosis. In this patient, thrombosis was manifested by recurrent spontaneous ulcerations of the fingers and toes. Hemorrhage was manifested on two occasions by the retropharyngeal bleeding precipitated by sneezing, requiring his admission to the hospital, and by upper gastrointestinal bleeding which occurred after admission. The most frequent site of hemorrhage in the polycythemic patient is the gastrointestinal tract.

This case adds to the list of causes of compromise of the upper airway and to the list of complications of polycythemia vera.

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