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Retropharyngeal Hematoma as an Unusual Presentation of Myelodysplastic Syndrome: A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Male, 90
Final Diagnosis: Spontaneous retropharyngeal hematoma
Symptoms: Dysphagia
Medication: —
Clinical Procedure: —
Specialty: Hematology

Objective: Rare co-existance of disease or pathology

Background: Retropharyngeal hematoma is a relatively rare diagnosis that requires a high clinical suspicion and stabilization of the airway to prevent rapid deterioration. We report a case of a spontaneous retropharyngeal hematoma in an elderly patient with myelodysplastic syndrome and associated thrombocytopenia.

Case Report: A 90-year-old man with myelodysplastic syndrome was brought to the Emergency Department with complaints of difficulty swallowing and muffled voice for 24 hours. Upon arrival, his vital signs and physical exam were unremarkable, except that when he was asked to take a sip of water, he could not swallow it. Complete blood count was remarkable for leukocytosis of 14.3×10^3/mcL, hemoglobin of 9.0 gm/dL, and platelet count of 26×10^3/mcL. Chest X-ray and lateral soft-tissue neck X-rays were grossly unremarkable. The patient was admitted for further evaluation and was scheduled for esophagogastroduodenoscopy. During intubation for esophagogastroduodenoscopy, the patient was noted to have significant airway narrowing. A subsequent CT scan revealed a 3×2×2 cm supraglottic hypodensity, thought to represent a retropharyngeal hematoma. The patient was transferred to the Intensive Care Unit (ICU) and received platelet transfusions. The ICU course was complicated by anemia, which necessitated transfusion of packed red blood cells. On hospital day 7, the patient reported resolution of his symptoms and was discharged home.

Conclusions: This case adds to the growing body of literature on spontaneous retropharyngeal hematomas. High clinical suspicion is warranted in patients who present with acute dysphagia, odynophagia, and dysphonia. Prompt imaging and airway management are vital in managing patients with this condition.

MeSH Keywords: Hematoma • Laryngoscopy • Myelodysplastic-Myeloproliferative Diseases • Thrombocytopenia

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Background

Spontaneous retropharyngeal hematoma is a rare diagnosis that has been linked to numerous coagulopathies [1]. Retropharyngeal hematomas can cause emergent airway compromise and are associated with a mortality rate of approximately 10% [2]. Intubation or cricothyrotomy is often necessary in patients with retropharyngeal hematoma due to rapid progression of the hematoma and blockage of the airway. Thus, initial physical exam and imaging studies are crucial for making a timely diagnosis. We describe a case that appears to be related to underlying myelodysplastic syndrome. Despite the patient's known medical history and clear evidence of thrombocytopenia, the treatment team did not appreciate the hematoma until incidental visualization during intubation. Thus, spontaneous retropharyngeal hematoma should be included in the diagnosis when examining a patient with dysphagia or odynophagia, dysphonia, and dyspnea.

Case Report

A 90-year-old man was brought in to the Emergency Department (ED) from home with a chief concern of difficulty swallowing and a cough. He reported worsening dysphagia over the previous 24 h, with an inability to eat, take pills, or drink during that time. A home health aide noted that his voice had become muffled as well over this time. He had no history of similar symptoms and denied dyspnea and any sensation of throat or mouth swelling. Further review of systems was negative. Past medical history included hypertension, atrial fibrillation, myocardial infarction, prostate cancer, myelodysplastic syndrome, chronic obstructive pulmonary disease and peripheral artery disease. His surgical history included abdominal aortic aneurysm repair, two coronary artery stent placements, laparoscopic cholecystectomy, and tonsillectomy at age 5. Home medications at the time of presentation included metoprolol succinate (50 mg PO once daily), amlodipine (5 mg PO once daily), furosemide (40 mg PO once daily), and tamsulosin (0.4 mg PO once daily).

Upon arrival to the ED, his vitals were as follows: temperature 36.4°C, heart rate 96 bpm, blood pressure 148/60 mmHg, and O_2 saturation 99% on room air. He was not in apparent distress and the physical exam was unremarkable, except when he was asked to take a sip of water, he coughed, sputtered, and could not swallow it.

Complete blood count was remarkable for leukocytosis of 14.3×10^3/mcL (reference range 4.8–10.8), hemoglobin of 9.0 gm/dL (reference range 14.0–18.0), mean corpuscular volume of 98.6 fL (reference range 80.0–94.0), red cell distribution width of 17.1% (reference range 11.5–14.5), and platelet count of 26×10^3/mcL (reference range 140–440). The basic metabolic panel was unremarkable. Coagulation labs showed an international normalized ratio of 1.07 (reference range 0.84–1.12). The chest X-ray was unremarkable and lateral soft tissue X-ray of the neck was read as equivocal, with possible thickening of aryepiglottic folds (Figure 1). The decision was made to admit the patient for esophagogastroduodenoscopy (EGD) and further medical management. During intubation for EGD, the patient was noted via laryngoscope to have a large hematoma posterior to the pharynx, as well as airway narrowing and deviation, so the procedure was aborted. The patient was transferred to the ICU at this time due to new-onset tachypnea and the possibility of airway compromise. A computed tomography (CT) scan was performed, showing an approximately 3×2×2 cm supraglottic hypodensity, thought to represent a spontaneous retropharyngeal hematoma (Figure 2). The patient received 3 units of platelets at this time, with the goal of raising his platelet count above 50×10^3/mcL.

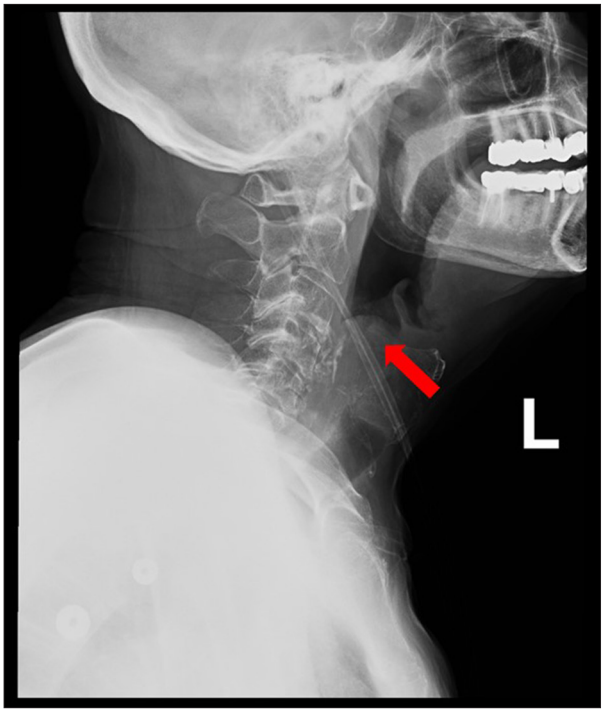

Figure 1. A lateral soft-tissue X-ray of the neck was obtained after initial chest X-ray was negative. The lateral film was read as equivocal, with possible thickening of the arylepiglottic folds (arrow).

The ICU course was complicated by episodes of atrial fibrillation and associated tachycardia, which were controlled initially with metoprolol (5 mg IV every 6 hours) and after three days with his home dose of metoprolol succinate (50 mg PO once daily). He continued to be NPO (*nil per oris* [nothing by mouth]) during the stay and intermittently coughed up bloody sputum. The patient's hemoglobin slowly dropped over the ICU

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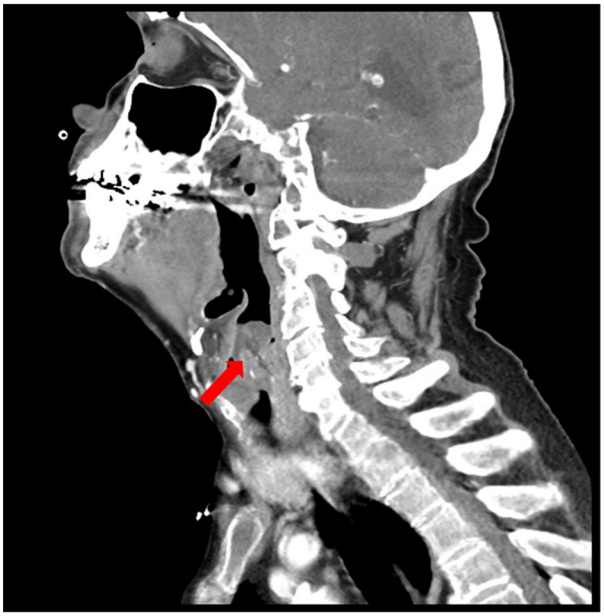

Figure 2. Sagittal CT obtained after abnormal findings noted on direct laryngoscopy showed a 3×2×2 cm supraglottic hypodensity (arrow), thought to represent a spontaneous retropharyngeal hematoma.

course to a nadir of 6.9 gm/dL on hospital day 6, which necessitated a transfusion of 1 unit of packed red blood cells. On hospital day 7, he reported that his swallowing had improved and his voice was sounding less muffled. He was discharged home with the hematoma improved at the time of discharge.

Discussion

While there are sporadic case reports of retropharyngeal hematomas, this case is the first to our knowledge of a spontaneous retropharyngeal hematoma in a patient with myelodysplastic syndrome and thrombocytopenia [1–9]. Until the hematoma was incidentally visualized, neither history, physical exam, labs, nor initial imaging led anyone to suspect this pathology. Thus, this diagnosis requires a high clinical suspicion, particularly in the context of a coagulopathy such as myelodysplastic syndrome.

Myelodysplastic syndrome has varying presentations. In this patient, thrombocytopenia was the dominant manifestation and likely was the main precipitating factor for the development of the hematoma. Other case reports have found spontaneous retropharyngeal hematomas in patients on anticoagulation with warfarin and novel oral anticoagulants, but thrombocytopenia appears to be an independent risk factor as well [1,2,7]. One previous report of spontaneous retropharyngeal hematoma in a patient with Epstein-Barr virus was attributed to a combination of acute tonsillitis, bone marrow suppression and altered clotting profile due to the virus [10]. Spontaneous retropharyngeal hematomas have occurred after minor trauma such as sneezing or coughing, but can also happen in the absence of such trauma [8]. The specific precipitating factor for this case remains a mystery, but it is likely that minor trauma in the setting of thrombocytopenia played a role in the development of the hematoma.

The main lesson that can be taken from this and similar cases is that airway management is the most important consideration. The decision not to intubate in this patient was in part due to the patient's wishes. However, a tracheostomy kit was placed at the bedside and left there for the entire clinical course in anticipation of potentially rapid and devastating airway closure. It has been reported that 33% of patients require emergency surgical airway access within 2 hours of ED arrival and up to 10% of all cases end in death [3]. From the ED to the ICU, plans must be in place for a surgical airway, in addition to rapidly enlarging hematomas and other hematologic complications, in anticoagulated or coagulopathic patients.

Conclusions

This case emphasizes a rare but life-threatening condition that has the potential for rapid clinical deterioration. Clinicians should be aware of spontaneous retropharyngeal hematomas when examining patients with unexplained symptoms similar to those above.

Acknowledgements

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Conflict of interest

None.

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