

Pseudo-Ludwig's Phenomenon: A Rare Clinical Manifestation in Liver Cirrhosis

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Abstract

Pseudo-Ludwig's phenomenon, an exceedingly rare clinical entity, is characterized by non-infectious upper airway obstruction resulting from spontaneous hematoma in the sublingual space. The condition is often described in patients on anticoagulants with grossly deranged coagulation profile. We describe a case of pseudo-Ludwig's phenomenon in a cirrhotic patient with significantly deranged hemostasis profile; the first to be reported in medical literature.

Introduction

Liver cirrhosis is often associated with a myriad of hemostatic defects including clotting factor deficiencies, thrombocytopenia, and platelet function abnormalities.¹ Bleeding manifestations are common in cirrhotics and include gastrointestinal bleeds, skin bleeds, and, rarely, hemoperitoneum, intracranial bleeds, and intramuscular hematomas. The sublingual floor of mouth is an uncommon site of hematoma formation; spontaneous hematomas in this region resulting in noninfectious upper airway obstruction are known as pseudo-Ludwig's phenomenon.²⁻⁶ To date, no case of spontaneous sublingual hematoma or pseudo-Ludwig's phenomenon has been described complicating cirrhosis of liver.

Case Report

A 63-year-old male with decompensated alcohol-related cirrhosis of liver presented with a 1-day history of pain and swelling in the floor of his mouth. The patient noticed a few ecchymotic patches over the legs and trunk, and also had associated excessive fatigue, leg edema, abdominal distension, and altered sleep pattern. There was no history of falls, facial trauma, dental procedures, hematemesis, melena, or other orifice bleeds. On examination, the patient was disoriented, had stigmata of chronic liver disease, edema, ascites, and a palpable firm nodular liver in the epigastrium. Examination of the oral cavity revealed a large hematoma in the sublingual region of the floor of mouth (Figure 1). Hemogram revealed hemoglobin 9.8 g/dL and platelet count 28,000 cells/cm². Liver enzymes were bilirubin 5.8 mg/dL, AST 56 IU/mL, ALT 46 IU/mL, total protein 6.2 gm/dL, and albumin 2.4 gm/dL, with INR 6.5.

Serum electrolytes and renal functions were normal at admission. Ascitic fluid examination revealed high serum-ascites albumin gradient (SAAG) ascites with no evidence of spontaneous bacterial peritonitis. The patient was managed with anti-hepatic coma measures, parenteral vitamin K, fresh frozen plasma, and platelet transfusions. The patient had progressive increase in the size of the hematoma causing postero-superior displacement of tongue resulting in airway obstruction. Airway was maintained with nasotracheal intubation. Drainage of the hematoma was performed surgically and hemostasis was achieved with fresh frozen plasma and platelet concentrate transfusions. Unfortunately, the patient developed pneumonia that progressed to septicemia despite aggressive antibiotic therapy, and he died of septic shock and multi-organ failure.

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Figure 1. Clinical photographs revealing sublingual hematoma.

Discussion

Pseudo-Ludwig's phenomenon was first described by Lepore in 1976 as a condition caused by deranged coagulation resulting in spontaneous bleeding into the sublingual and submaxillary spaces and acute upper airway obstruction.³ The floor of the mouth, though highly vascular, is rarely a site of hematoma formation. Floor of mouth hematomas have been described after trauma and dental implant procedures.^{2,6} There are rare reports of hematomas in this region complicating excessive anticoagulation.^{2,4} These hematomas are potentially life-threatening as they cause elevation of tongue

and floor of mouth culminating in upper airway blockade.²⁻⁵ Upper airway obstruction is a well-documented complication of pseudo-Ludwig's phenomenon and it requires urgent intervention for maintenance of the airway.

Cirrhotic patients are prone to spontaneous bleeds due to multiple aberrations in the hemostatic cascades and abnormalities in platelet function and production. Predisposing characteristics for rare bleeds in cirrhosis include Asian ethnic descent and history of alcoholism, both of which were true for our patient. As he had no antecedent trauma to account for his hematoma formation, we hypothesize that hemostatic defects associated with cirrhosis led to the hematoma. Whether the pseudo-Ludwig's predisposed our patient to aspiration pneumonia is unclear though logically possible secondary to obstruction and subsequent impaired normal airway defenses. Spontaneous extra-gastrointestinal hematomas are possible in patients with cirrhosis of liver and are often associated with dismal prognosis.

Disclosures

Author contributions: GS Zacharia drafted the manuscript and is the article guarantor. S. Kandiyil assisted with manuscript preparation. V. Thomas critically reviewed the manuscript and suggested modifications. All authors approved the final manuscript.

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The patient is now deceased, but informed consent for this case report was obtained from the patient's next of kin.

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References

1. Kujovich JL. Hemostatic defects in end-stage liver disease. *Crit Care Clin.* 2005;21:563-587.
2. Puri A, Nusrath MA, Harinathan D, Lyall J. Massive sublingual hematoma secondary to anticoagulant therapy complicated by a traumatic denture: A case report. *J Med Case Rep.* 2012;6:105.
3. Lepore M. Upper airway obstruction induced by warfarin sodium. *Arch Otolaryngol Head Neck Surg.* 1976;102:505-506.
4. Cohen AF, Warman SP. Upper airway obstruction secondary to warfarin-induced sublingual hematoma. *Arch Otolaryngol Head Neck Surg.* 1989;115(6):718-720.
5. Bitar MA, Kamal MA, Mahfoud L. Spontaneous sublingual haematoma: A rare entity. *BMJ Case Rep.* 2012.
6. Smith RG, Parker TJ, Anderson TA. Noninfectious acute upper airway obstruction (pseudo-Ludwig phenomenon): Report of a case. *J Oral Maxillofac Surg.* 1987;45(8):701-704.

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