ISSN 2689-8268 | Volume 7

American Journal of Surgery and Clinical Case Reports

Review Article Open Access

Scleroderma A Challenge for the Anesthesiologist

Gurman GM*

Ben Gurion University of the Negev, Beer Sheva, Israel

***Corresponding author:**

Gabriel M Gurman,
Ben Gurion University of the Negev, Beer Sheva, Israel

Received: 02 Feb 2024
Accepted: 23 Apr 2024
Published: 29 Apr 2024
J Short Name: AJSCCR

Citation:

Gurman MG.Scleroderma A Challenge for the Anesthesiologist. Ame J Surg Clin Case Rep. 2024; 7(14): 1-3

1. Introduction

Scleroderma does not appear on the list of the "modern killers" such as: smoking, obesity, alcohol and drug abuse, and diabetes. However, since it affects a series of organs it may represent a real danger, especially in the perioperative period. Scleroderma is a progressive autoimmune vasculopathy, with fibrous deposition throughout the body, with an onset between the ages of 45 and 64 years, predominantly affecting women [three to eight women for each man [1]. This is a disease with a rather rare frequency [2], 38-340 cases/million, and all together some 2.5 million patients all over the world. Today, in the era of advanced possibilities regarding diagnostic investigations and modern treatment the 5-year survival is at around 90%. The main causes of death in patients with systemic scleroderma are pulmonary fibrosis, pulmonary artery hypertension and cardiac involvement [3]. Volkman and Mc Mahan [4] described the gastrointestinal [GI] involvement in scleroderma, which occurs in nearly all patients suffering from the systemic form of the disease. Any part of the upper GI tract may be involved, producing gastro-esophageal reflux, lower esophageal sphincter dysfunction, esophageal dysmotility, dysphagia, and gastroparesis with slow gastric emptying. Overall, patients with scleroderma seem to require more often high-risk surgeries [5]. When compared with the general population, patients with scleroderma undergo twice the number of thoracic, breast and vascular interventions, but fewer urological and gynecological/obstetric procedures.

2. The case

A 59-year-old lady was diagnosed as having a malignant uterine tumor, for which she was planned for a total hysterectomy. She was known for suffering from stabilized arterial hypertension, polymyositis and scleroderma. Five years earlier, she had a cerebral vascular accident, after which she remained with weakness of the left leg muscles. The patient was admitted to a gynecological department, and the preoperative lab investigations showed anemia [Hb 9.1 g/100], slight hypopotassemia [3.8 mEq/l], BUN 50 mg/100, blood sugar 115 mg/100, all the other tests being into normal range. Twenty-four hours before surgery the patient was examined by an anesthesiologist, who classified her as ASA 2. He proposed to the patient general anesthesia, she accepted and signed up the informed consent. Scleroderma was not mentioned in the pre-anesthesia chart. Next morning, during induction of general anesthesia unexpected difficulties in performing tracheal intubation were encountered, as well as difficulty in ventilating the patient. Regurgitated gastric content was noticed in the oral cavity, blood oxygenation saturation decreased to 87%, but returned to normal once the tracheal intubation eventually succeeded and the patient was mechanically ventilated with oxygen [FiO2] at 100%. Because of the need for high inspiratory pressures in order to maintain arterial blood gases within normal limits the surgical procedure was cancelled and the patient transferred to an intensive care unit, where aspiration pneumonia was diagnosed, and antibiotic treatment started. The patient remained sedated and ventilated for the next 10 days. On the 8th day a percutaneous tracheostomy was performed. GI investigations revealed esophageal dysmotility and slow gastric emptying. Two weeks after the first attempt of operating her, the patient was stable from the respiratory point of view, she was weaned off the ventilator, but still needing oxygen through the tracheostomy cannula. A radical abdominal hysterectomy was performed under general anesthesia and the patient returned to the intensive care unit for the continuation of the treatment. Enterobacter was found in the bronchial secretions, while the patient developed bilateral pneumonia. Treatment with cephalosporins i-v was initiated. Other findings were very severe metabolic acidosis [pH 7,15], hypoxia [PaO2 65 mm Hg on FiO2 70%], oliguria, and

sepsis not responding to vasopressors. She died 10 days after surgery, presenting clear signs of adult respiratory distress syndrome [ARDS] and septic shock.

3 Discussion

This case presentation is important for the anesthesiologist who could encounter anytime in his career a patient suffering from scleroderma with a serious GI pathology. The GI symptoms, among other clinical aspects of the disease, demand a special attention in the pre-operative period. Efrimescu et al. [6] emphasize the importance of a correct preoperative assessment: primarily searching for relevant symptoms such as dysphagia, reflux esophagitis, but also for a possibility of limited range of movement or contractures involving relevant joints [neck, mouth opening]. Their recommendation is clear: "An 'organ system' approach should be followed, and careful consideration should be given to the potential anesthetic issues". They also recommend performing gastric scintigraphy, a useful test to confirm abnormal gastric emptying. Carr et al. [7] presented the main concerns for the anesthesiologist regarding scleroderma, among them difficult intubation and danger of aspiration. Marie et al [8] used cutaneous electrogastrography [EGG] as a test for diagnosing disturbances of gastric emptying in patients with scleroderma. The prevalence of EGG disturbances was as high as 81% of the investigated patients. Sallam, McNearney and Chen [9] described the esophageal dysmotility as a most common GI manifestation in scleroderma, resulting in prolonged esophageal transit time and delayed esophageal emptying. Adler et al. [10] mentioned the fact that difficulty in opening the mouth could become a serious impediment in achieving a successful tracheal intubation. The use of rapid sequence induction [11], as a recommended technique in the management of critically unwell patients to address risk of aspiration of gastric content, it is not a panacea. Avery et al. [12] concluded in their report that this technique only reduces, but does not annul, the frequency of failed intubation. It is clear that the first flaw in managing this case was the inadequate preoperative assessment, and this is the reason why the anesthesiologist in charge did not pay attention to the fact that the patient had clear symptoms of GI malfunctioning. This encompassed also the ignorance for anatomical changes which complicated the process of assuring an easy approach to the airways during induction of general anesthesia. Additionally important was the choice for the best anesthesia technique for the proposed surgery, in this case for total hysterectomy. By taking into consideration possible GI related disturbances which accompany the disease in the vast majority of cases, it becomes clear that for this specific surgical intervention a neuroaxial anesthesia would have been the preferred technique. Since anatomical deformities, such as poor mouth opening or temporomandibular fibrosis, can be present in most of the scleroderma patients, mask ventilation and/or tracheal intubation can be difficult to achieve. This is the reason why Dempsey, Rowell and McRobert [12] recommended epidural or continuous spinal-epidural anesthesia for patients with scleroderma, these techniques having the additional benefit of continuous regional analgesia into the postoperative period. They also described other advantages of regional anesthesia in patients with scleroderma, such as: vasodilatation which improves tissue perfusion and promote wound healing, as well as preventing Raynaud's phenomenon, a medical condition in which the spasm of small arteries causes episodes of reduced blood flow to end arterioles. The complications which showed up during the first attempt to anesthetize the patient had all the same explanation: a superficial pre-operative investigation regarding the possible presence of GI symptoms [a situation encountered in the vast majority of patients with scleroderma], and the wrong selection of the most appropriate anesthesia technique for this patient. Selecting neuroaxial anesthesia technique [spinal, epidural, or combined] for this patient would probably have avoided the complications produced by the presence of GI involvement, as well as by the anatomic changes which compromised the chances of a smooth tracheal intubation.

4. Conclusion

Scleroderma is a rare but serious medical condition, since it affects most of organs and organ systems. Among them, the GI involvement produces a series of changes in the functionality of the digestive tract with a direct influence on anesthesia. The anesthesiologist's task includes a correct preoperative assessment, emphasizing those pathological modifications that have direct implications on the subsequent anesthesia technique. The presence of GI changes, especially in the upper part of the tract demands a serious discussion regarding the recommended anesthesia technique, and regional anesthesia is the preferred technique to be used in case it can cover the surgical field of the planned operation.

References

1. Őzcan C, Gőrűr K, Meltem ND. Imaging case study of the month massive bilateral inferior concha bullosa. Ann Otol Rhinol Laryngol. 2002; 111: 100-1.
2. Doğru H, Döner F, Uygur K, Gedikli O, Cetin M. Pneumatized Inferior turbinate. Am J Otolaryngol. 1999; 20: 139-41.
3. Alnatheer AM, Alkholaiwi F. Concha bullosa of the Inferior turbinate. Cureus. 2013; 10: e19089.
4. Pittore B, Safi AL, Jarvis SJ. Concha bullosa of the inferior turbinate:an unusual cause of nasal obstruction. Acta Otorhinolaryngol Ital. 2011; 31: 47-9.
5. Arslan M, Muderris T, Muderris S. Radiological study of the intumescentia septi nasi anterior. J Laryngol Otol. 2004; 118: 199-201.
6. Unlu HH, Altuntas A, Aslan A, Eskiizmir G, Yucel A. Inferior concha bullosa. J Otolaryngol. 2002; 31: 62-4.
7. Alkhaldi AS, Alhedaithy R, Alghonaim Y. Concha bullosa of the inferior turbinate: report of two cases. Cureus. 2021; 13: e15479.
8. Yang BT, Chong VF, Wang ZC, Xian JF, Chen QH. CT appearance of pneumatized inferior turbinate. Clin Radiol. 2008; 63: 901-5.

9. Aydin Ő, Űstundağ E, çiftçi E, Keskin IG. Pneumatization of the inferior turbinate. Auris Nasus Larynx. 2001; 28: 361-3.
10. Cankaya H, Egeli E, Kutluhan A, Kiris M. Pneumatization of the concha inferior as a cause of nasal obstruction. Rhinology. 2001; 39: 109-11.
11. Lei L, Wang R, Han D. Pneumatization of perpendicular plate of the ethmoid bone and nasal septal mucocele. Acta Otolaryngol. 2004; 124: 221-2.
12. Ozcan KM, Gedikli Y, Ozcan I, Pasaoglu L, Dere H. Microdebrider for reduction of inferior turbinate: evaluation of effectiveness by computed tomography. J Otolaryngol Head Neck Surg. 2008; 34: 463-8.
13. Kiroglu AF, çankaya H, Yuca K, Kara T, Kiris M. Isolated turbinitis and pneumatization of the concha inferior in a child. Am J Otolaryngol. 2007; 28: 67-8.
14. Bolger WE, Butzin CA, Parsons DS. Paranasal sinus bony anatomic variations and mucosal abnormalities: CT analysis for endoscopic sinus surgery. Laryngoscope. 1991; 101: 56-64.
15. Cannon CR. Endoscopic management of concha bullosa. Otolaryngol Head Neck Surg.