

Background

- Kennedy disease (KD) is an adult-onset, X-linked recessive disorder caused by CAG trinucleotide repeat expansion in the androgen-receptor (AR) gene on chromosome Xq11-12, primarily affecting males.
- KD affects approximately 1 in 40,000 males globally.
- Symptoms typically arise between ages 30-50, with progressive weakness and wasting of facial, bulbar, and proximal limb muscles.
- Progressive symptoms may include dysphagia, dysarthria, respiratory compromise, and, in severe cases, aspiration risk—posing challenges during emergency surgery.
- Disease requiring emergency surgery, focusing on mitigating risks like laryngospasm, hyperkalemia, and respiratory failure.

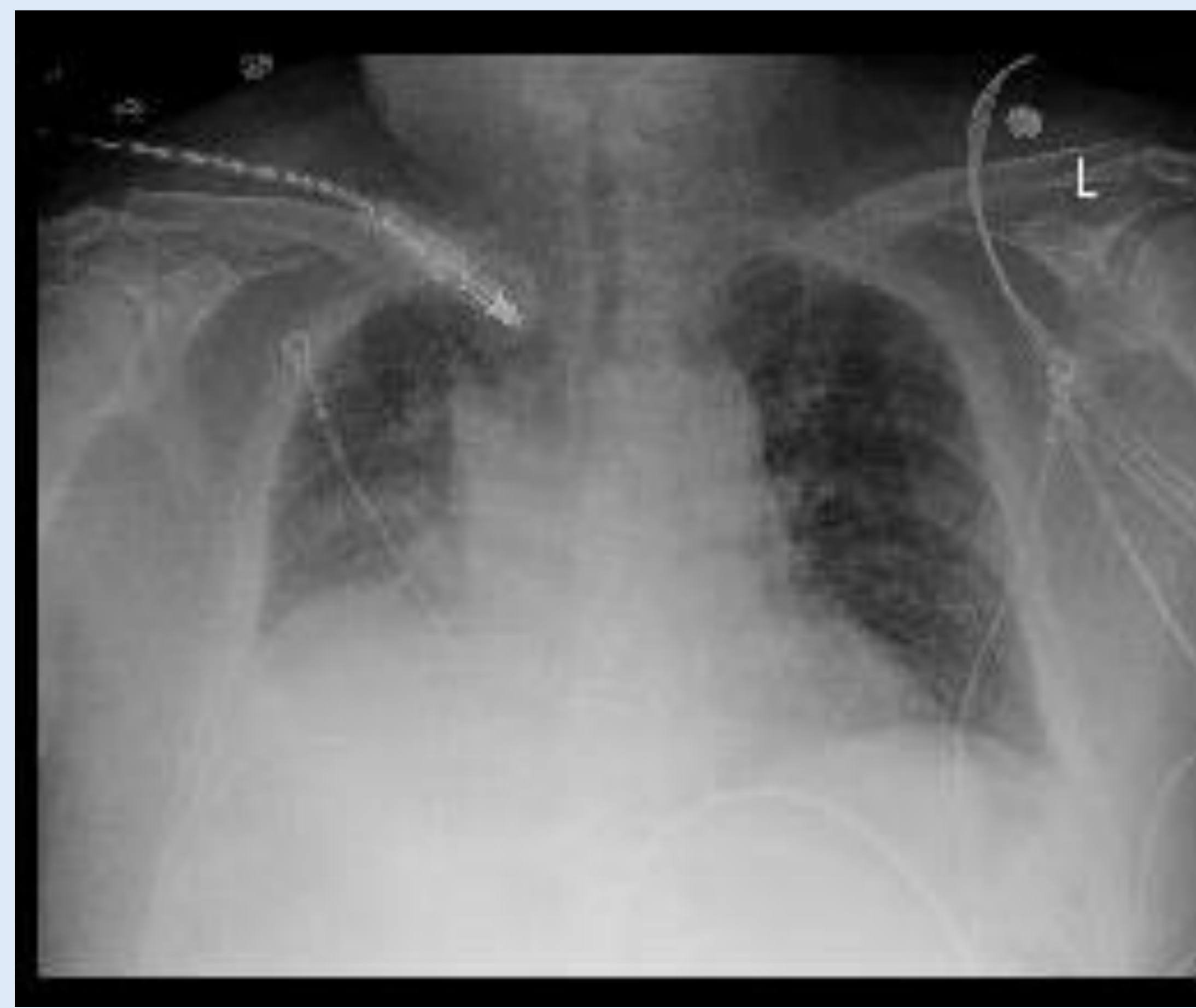
Case Description

A 58-year-old male with Kennedy's disease, hypertension, diabetes mellitus, and hyperlipidemia diagnosed with urosepsis secondary to right pyonephritis with a right ureteral stone, necessitating emergency cystoscopy with right ureteral stenting under general anesthesia.

Postoperatively in the PACU, he could not be safely extubated and was transferred to the MICU.

Extubation occurred on post-op day 1, but he developed altered mental status two days later, requiring re-intubation.

Postoperative chest X-ray showed right-sided atelectasis and elevated right hemidiaphragm.



Case Description (Continued)

Bronchoscopy revealed a mucous plug with thick secretions in the right lower lobe.

He was extubated on post-op day 4, weaned to high-flow nasal cannula, and eventually to room air on post-op day 16.

Discussion

Due to its rarity, there are no established guidelines for safe anesthesia in patients with KD.

Succinylcholine should be avoided due to the risk of hyperkalemia from elevated acetylcholine levels in these patients.

The use of non-depolarizing neuromuscular blockade is acceptable with intraoperative monitoring of the degree of relaxation.

Our case aims to contribute to the existing literature on anesthetic management for KD, emphasizing the unique considerations required for this patient subset.

References

Niesen AD, Sprung J, Prakash YS, Watson JC, Weingarten TN. Case series: anesthetic management of patients with spinal and bulbar muscular atrophy (Kennedy's disease). J Can Anesth 2009;56:136-141

Sperfeld AD, Hanemann CO, Ludolph AC, Kassubek J. Laryngospasm: an underdiagnosed symptom of X-linked spinobulbar muscular atrophy. Neurology 2005;64:753-754