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Case Report

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Sagliker syndrome: a rare manifestation of uncontrolled secondary hyperparathyroidism in chronic renal failure – a comprehensive case report

Abstract

We present a case of a 22-year-old female patient with a history of chronic kidney disease (CKD)and secondary hyperparathyroidism, complicated by pathological fractures of both femurs and subsequent joint replacements. The patient exhibited craniofacial deformities with lytic lesions inthe maxilla and mandible, airway abnormalities and cardiovascular adaptations due to hypertensive heart disease. A subtotal parathyroidectomy was performed, a combination of medications and nerve blocks was used for anesthesia induction and intubation, with subsequentbronchospasm and desaturation necessitating intervention. The surgical procedure was successful, with no further intraoperative issues.

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Introduction

Sagliker syndrome (SS), an intricate medical phenomenon, was introduced in 2004 by Sagliker et al. as an amalgamation of secondary hyperparathyroidism (SHPT) and chronic renal insufficiency.[1] This unique syndrome was brought to light through the observation of two casesof chronic kidney disease (CKD) co-occurring with facial deformity symptoms. Although SS is rare, it demands attention due to its distinct clinical presentation and the potential impact it can have on the lives of affected individuals.

In this case report, we present a detailed account of a patient with Sagliker syndrome, highlighting the distinctive clinical features, diagnostic journey, and therapeutic considerations. Through this case, we contribute to the growing body of knowledge surrounding the clinical understanding andmanagement of this rare syndrome.

Case report

A 22-year-old female, diagnosed with KDIGO stage 5 chronic kidney disease, which has exhibited a progressively worsening course since the age of 17. The underlying etiology remains undetermined. The initial presentation involved the onset of uremic syndrome, requiring the initiation of peritoneal dialysis treatment. Subsequent to this, due to recurrent catheter dysfunction occurring on two occasions, the patient transitioned to hemodialysis. Concurrently, the patient was diagnosed with systemic arterial hypertension and they started her on losartan treatment.

Approximately one year subsequent to the aforementioned events, the patient was diagnosed with secondary hyperparathyroidism. This condition was accompanied by a pathological fracture in the right femur, followed by a similar fracture in the left femur. As a result of these fractures, joint replacement procedures were performed to address the effects on mobility and function.

Clinical course

The patient experienced progressive palatal region volume enlargement, leading to difficulties invoice articulation and chewing, subsequently leading to shortness of breath (Figure 1).

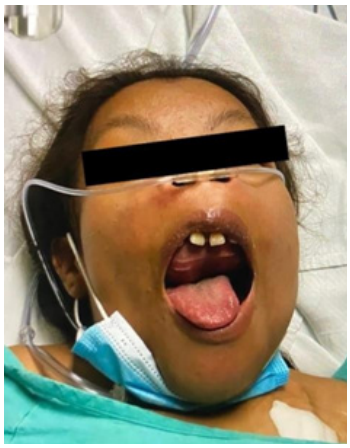

Figure 1

Diagnostic investigations

An assessment of alkaline phosphatase yielded a result of 1734 U/L, accompanied by a serum calcium concentration of 10 mg/dL. Parathyroid hormone (PTH) levels were elevated at 1410 pg/mL. A bone scintigram unveiled hyperfunctioning parathyroid tissue located within the confinesof the lower parathyroid glands. Furthermore, the imaging disclosed the presence of multiple lytic lesions characterized by an expansive morphology and incorporation of soft tissue elements, localized in both the maxilla and mandible. Adjacent bone structures exhibited resorption patternsresembling a "salt and pepper" configuration.

Cardiac evaluation

An echocardiogram was performed, revealing global and segmental mobility of cardiac structures. The left ventricular ejection fraction (LVEF), determined using biplanar Simpson's method, registered at 57%, with preserved biventricular systolic function. There was no data of increasedfilling pressures. Notably, evidence of hypertensive heart disease with mild structural implicationswas identified.

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Physical examination:

Upon physical examination, with a weight of 43 kg, and a height of 130 cm. The patient displayedintegumentary paleness and craniofacial anomalies, characterized by Mallampati classification IV, Patil Aldreti classification III, sternomental distance III, and mandibular subluxation IV (Figure1).

Treatment and surgical plan:

The patient has been scheduled for a subtotal parathyroidectomy to address the hyperparathyroidism.

Intraoperative course:

Given the challenging anatomy, sedation with dexmedetomidine (0.5mcg/kg) infusion along with Ketamine 30 mg IV bolus was performed. Laryngeal nerve block using 2% lidocaine was successfully performed without complications. The patient tolerated the procedure well. Pre- oxygenation was carried out with a face mask delivering oxygen at a rate of 5 liters per minute, coupled with head and neck hyperextension. Intubation was achieved using a Karl Storz C-MACD-blade and was accomplished on the fourth attempt with a POGO score of 70%. The surgery proceeded uneventfully, with the patient remaining hemodynamically stable. Approximately 15 minutes after endotracheal intubation, the patient exhibited bronchospasm, concomitant with a decrease in oxygen saturation to 73%. The bronchospasm was associated with resistance to positive pressure ventilation. Urgent intervention was initiated, including the administration of aminophylline, steroids, and adrenaline. These interventions resulted in a satisfactory response,as evidenced by an increase in oxygen saturation to 92%. The subsequent course of the surgical procedure proceeded without any additional intraoperative complications. The patient remained stable, and the planned surgical steps were executed without issue. Adequate depth of anesthesia was maintained, and no signs of intraoperative awareness were noted. Upon completion of the surgical procedure, the patient's anesthesia was tapered off, and the patient was safely extubated.

Discussion

Sagliker Syndrome, though rare, presents intricate challenges in diagnosis, management, and perioperative care. This case of a 22-year-old female with KDIGO stage 5 chronic kidney disease(CKD) offers insights into the complex interplay between CKD-related complications and associated manifestations.

The progressive worsening of CKD since the age of 17 in this case underscores the need for etiological investigation, considering both genetic and environmental factors. Although several studies have suggested the involvement of different factors in this syndrome, the exact cause is still unknown. Some genetic mutations were detected among 40% of patients with SS, which is possible to be associated with the pathogenesis of the syndrome. Moreover, it is shown that missense mutations on the GNAS1 gene play an important role in SS pathogenesis.[2-4]

The hallmark of SS is the presence of pronounced facial and skeletal alterations, with a preponderance of cases occurring in young women aged between 18 and 39 years.[5] These individuals exhibit severe manifestations, including maxillary or mandibular destruction, short stature, and fingertip deformities. These skeletal changes, coupled with the concomitant secondary hyperparathyroidism, contribute to the unique clinical picture associated with SS. It is shown that high levels of alkaline phosphatase and parathyroid hormone in patients with secondary hyperparathyroidism, who suffered from CKD for a long time, play a major role in SS clinical manifestations (6). The palatal volume enlargement leading to speech and chewing difficulties manifestation is linked to soft tissue changes in CKD. The bone scintigram's findings of hyperfunctioning parathyroid tissue and lytic lesions corroborate the association between CKD,secondary hyperparathyroidism, and skeletal abnormalities. Secondary hyperparathyroidism, particularly when emerging at a young age, has far-reaching consequences that extend beyond the musculoskeletal system. It triggers intricate changes within various organ systems, includingthe immune and cardiovascular systems.[5]

Cardiac evaluation with echocardiogram reveals the coexistence of CKD and hypertensive heart disease, accentuated by structural implications.[6,7]

The planned subtotal parathyroidectomy for hyperparathyroidism management is pivotal. The anesthetic approach using dexmedetomidine, ketamine, and laryngeal nerve block with lidocaine exemplifies careful perioperative planning tailored to the patient's unique condition. However, thebronchospasm incident post-intubation highlights the importance of continuous vigilance and rapid intervention in such intricate cases.[8]

Conclusion

In conclusion the interplay between renal dysfunction, secondary hyperparathyroidism, cardiovascular involvement, skeletal abnormalities, and even unique palatal changes underscores the necessity for a multidisciplinary approach in managing this rare syndrome. Thisreport contributes to the growing body of knowledge, emphasizing the need for a comprehensive perioperative anesthesia care plan to improve the safety of their care, and positively impact theirrecovery and outcomes.

Acknowledgments

None.

Conflicts of interest

None.

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