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Case Report

Surgical Repair of Secundum Atrial Septal Defect in a Patient with Addison's Disease and Diabetes Mellitus Type I: A Case Report

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Abstract

Primary adrenal insufficiency, most commonly due to autoimmune destruction of the adrenal cortex (Addison's disease), is a rare but life-threatening condition requiring lifelong glucocorticoid replacement. Surgical stress markedly increases cortisol demand and may precipitate adrenal crisis in inadequately covered patients. Atrial septal defect, especially the secundum type, is among the most common congenital heart defects; however, reports describing surgical atrial septal defect repair in patients with coexisting type I diabetes mellitus and Addison's disease are lacking. The perioperative management of such patients presents unique endocrine and hemodynamic challenges. This case highlights the importance of meticulous multidisciplinary coordination to prevent adrenal crisis and optimize glycemic control during major cardiac surgery. A 22-year-old male with a history of type I diabetes mellitus (diagnosed at age 16) and Addison's disease (diagnosed nine months prior) was incidentally found to have a large secundum atrial septal defect. Despite being asymptomatic, surgical repair was indicated following comprehensive cardiologic assessment, including right heart catheterization and balloon occlusion testing. A multidisciplinary team involving cardiology, cardiothoracic surgery, anesthesia, and endocrinology planned perioperative care. Stress-dose steroids were administered intraoperatively, and adequate glycemic control was achieved. The defect was successfully closed. Postoperatively, the patient remained hemodynamically stable without evidence of adrenal crisis. Glycemic fluctuations and reduced oral intake were managed with endocrine optimization. This case demonstrates that successful surgical atrial septal defect repair in patients with concomitant Addison's disease and type I diabetes mellitus is achievable through proactive endocrine optimization and close interdisciplinary collaboration. Early planning, appropriate stress-dose steroid coverage, and vigilant perioperative monitoring are essential to prevent adrenal crisis and improve outcomes in complex comorbid patients undergoing major cardiac surgery.

Keywords

Addison's Disease, Adrenal Crisis, Cardiac Surgery, Perioperative Management

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1. Introduction

Primary adrenal insufficiency (PAI) is defined as the impaired functioning of the bilateral adrenal cortex to produce adrenocortical hormones including cortisol, aldosterone, and androgens due to an underlying disease process. Whilst there can be multiple underlying causes of PAI such as infectious (tuberculosis, HIV), hemorrhagic (precipitated by coagulopathy, trauma), infiltrative (sarcoidosis, amyloidosis) or pharmacologic (by drugs that block cortisol synthesis), the most common cause of primary adrenal insufficiency is autoimmune destruction of the adrenal gland, also termed as, Addison's Disease [1]. The occurrence of Addison's disease is rare with an incidence of 0.6 per 100,000 people annually and its presentation depends on the extent of damage to the cortex [2]. The clinical presentation includes hypotension, altered mental status, anorexia, vomiting, weight loss, fatigue and abdominal pain. Serum cortisol, Adrenocorticotropic hormone (ACTH), renin, aldosterone and chemistry panel should be obtained in those suspected to have PAI [3].

Atrial septal defect (ASD) is one of the most common types of congenital heart defects accounting to about 10 to 15 percent congenital heart diseases. This defect arises from the incomplete closure of the communication between the left and right atria. Secundum defects account for approximately 70 percent of all ASDs and mostly occur as sporadic, isolated defects [4]. Although ASD is known to be reported with many syndromes but its surgical management along with medical optimization of co-existing multiple endocrine disorders including Diabetes Mellitus (DM) type I and Addison's Disease have not been reported before.

In this article, we report the case of a 22-year-old male, known case of DM type I and Addison's Disease, who was incidentally diagnosed with secundum atrial septal defect and underwent a successful surgical ASD closure. This work is compliant with the 2023 Surgical CAse REport (SCARE) guidelines [5].

The surgical repair of the large ASD with deficient rims necessitated interdisciplinary collaboration among anesthesiologists, endocrinologists, cardiologists and cardiac surgeons for successful management to prevent any adrenal crisis intra or post operatively. By detailing the contributions of each specialty to the patient's care, this case report highlights the importance of interdisciplinary collaboration in achieving optimal patient outcomes. By sharing our experience in managing this complex case, we aim to inform clinical practice and facilitate better decision making in similar clinical scenarios.

2. Case Presentation

Our patient was a 22-year-old male, who presented to us for the first time in the outpatient clinic on 6/03/2024 after being referred for cardiac evaluation from his primary physician. He was diagnosed with DM Type I at the age of sixteen and with Addison's Disease nine months ago but currently was asymptomatic and had no active complaints. The patient was non-smoker non-alcoholic with no family history of autoimmune disorders, diabetes mellitus or any cardiac conditions. On examination, he had a Basal Metabolic Index (BMI) of 16.25 kg/m^2 and was vitally stable. The rest of the systemic examination was unremarkable except for a systolic ejection murmur in left upper sternal border graded as 2/6.

His most recent lab results done on 7/02/2024 showed an ACTH of 115 pg/ml, cortisol of 13.87 µg/dl, creatinine of 0.9 and an elevated Hemoglobin A1c (HBA1c) of 8.94%. Serum 17-Hydroxyprogesterone (17 OH) and Anti-glutamic acid decarboxylase antibody (GAD-65) assay were done on 9/02/2024 which were 1.05 and 0.54 IU/ml respectively.

Echocardiogram was done in our centre on 28/03/2024 that showed large sized secundum ASD measuring 30-32 mm with left to right flow. Total septal length was 50 mm. The posterior rim was 4-5 mm (thin and floppy) and the inferior vena cava rim was very thin and deficient. Biventricular systolic function was normal. Left ventricular ejection fraction (LVEF) was 69%. After discussing with the cardiology team, a Right Heart Catheterization (RHC), fluid challenge and balloon occlusion test of ASD was planned and performed on 16/4/2024 under conscious sedation. Patient remained hemodynamically stable. The procedure had no complications.

A multi-disciplinary team meeting was held, and cardiothoracic surgery was taken on-board. After a thorough discussion between cardiology, cardiothoracic surgery, and cardiac anesthesia, surgical repair was planned. The department of endocrinology was consulted, and perioperative recommendations were obtained regarding adjustment of insulin and steroid doses.

Monitoring in the operating room included noninvasive blood pressure (BP), pulse oximetry, and ECG. Intravenous (IV) cannula (20G) was placed. The patient was then preoxygenated. 100 mg propofol and 50 mg atracurium were then administered intravenously. Endotracheal intubation was performed. Anesthesia was maintained with isoflurane at a minimum anesthetic concentration (MAC) of 1.0 in an oxygen and air mixture.

Meanwhile the cardiothoracic surgery team prepped and draped the chest as the patient was ready. Once the patient was on complete cardiopulmonary bypass (CPB), 8 mg of dexamethasone was administered by the anesthetist and 1g of methylprednisolone was administered by the perfusionist. Blood glucose was targeted to be kept between 150-200 mg/dL using variable rate insulin infusion. Right atrium (RA) was opened parallel to the atrioventricular groove and ASD was identified. It was closed primarily in a continuous manner in two layers. Mean arterial pressure (MAP) fluctuated throughout the surgery lowest being 25 mmHg which was responsive on boluses of 100 micrograms of phenylephrine. Norepinephrine was started at 0.05 micrograms /kilograms/

minute (μg /kg/min) to maintain a MAP between 50-55 mmHg. Rewarming was started and the patient gradually weaned off from the CPB machine on a support of 0.05 μg /kg/min of epinephrine, 0.3 μg /kg/min of milrinone and 0.05 μg /kg/min of norepinephrine. Our hemodynamic monitor had the option to estimate cardiac output and index, systemic vascular resistance (SVR) and index and stroke volume. It was observed that the patient was vasodilated owing to SVR, therefore norepinephrine was continued till SVR improved. Since the blood pressure was in the desired range norepinephrine was discontinued towards the end of surgery.

Paracetamol and ondansetron were administered by the anesthetist before extubation. The total bypass time was 48 minutes and cross-clamp time was 27 minutes. Patient was shifted to ICU extubated with inotropic milrinone support at 0.3 μg /kg/min.

Postoperatively, patient remained hemodynamically stable. Initially he had uncontrolled sugars, poor mobilization and inadequate oral intake. It was managed through active mobilization, and encouragement of feed. Oral intake improved after optimization of glucocorticoid therapy. Endocrinology was taken on board. A diabetic diet plan was advised for the patient and his medications were optimized. Patient was kept on insulin for two days. Once the oral intake improved basal bolus regime was started.

Before discharge, he was switched back to pre-operative routine after adjustment of medications. The patient was discharged on post operative day four with no active complaints.

3. Discussion

Cortisol is a steroid hormone, synthesized from cholesterol and its release occurs in a pulsatile manner. It has various functions in the body including regulating stress response and body's metabolism and maintaining immune function. Amongst various stressors such as infection, dehydration, trauma etc. surgery is a significant stressor inducing the release of cortisol [6]. The plasma concentration after a stressful situation remains at 40-50 g/dl, instead of the normal concentration which varies between 51.75 – 250.1 ng/ml depending on the time of the day [7, 8]. Any surgery including cardiac surgeries along with the CPB as in this case, the repair of the ASD secundum using CPB, are situations of acute stress for human organs and tissues [9]. The use of CPB acts as a major stimulus that causes endogenous release of catecholamines i.e. epinephrine and norepinephrine and stress hormones. It triggers activation of the hypothalamic-pituitary-adrenal axis (HPA) which induces an increase in cortisol secretion causing anti-inflammatory effects and inhibiting pro-inflammatory cytokine effects [10-12]. Literature suggests that the cortisol concentration is maintained at an increased level until the end of the surgery, reaching a maximum value within 4–6 hours after the operation. Then, the concentration decreases to the pre-operative values within 24 hours [13].

Patients who are suffering from PAI have impaired functioning of the bilateral adrenal cortex to produce adrenocortical hormones including cortisol, aldosterone, and androgens due to an underlying disease process [1]. In our case, the patient had an autoimmune cause i.e. Addison's Disease of PAI. Such patients may be receiving long-term oral corticosteroid therapy which maintains their cortisol levels. Patients receiving corticosteroid therapy have suppressed functioning of the HPA axis leading to an impaired stress response and immune response [14]. A daily glucocorticoid dose equivalent to prednisolone ≥ 5 mg, for longer than 1 month represents an adrenal suppressive dose in a proportion of adults (7). It is known from previous literature that all routes of administration i.e. oral, inhaled, topical, intranasal or intra articular have the tendency to suppress the functioning of the HPA axis [15]. Any interruptions in their regime accompanied with increased demand during surgical stress can increase the risk of adrenal crisis.

Adrenal crisis is defined as acute, life-threatening condition triggered by various factors with either known or unknown adrenal insufficiency. Adrenal crisis clinically manifests non-specifically. On clinical examination the patient may present with fever, tachycardia, and orthostatic hypotension, and may look visibly unwell [16]. Individuals with primary adrenal insufficiency might display signs such as skin and buccal mucosa hyperpigmentation and scarring [17]. Classic laboratory features include hyponatremia, resulting from mineralocorticoid deficiency, hyperkalaemia, resulting from mineralocorticoid deficiency, hypoglycaemia, stemming from decreased gluconeogenesis and glycogenolysis, and/ or high or high normal ACTH levels, as observed in primary adrenal insufficiency.

Intra- and post- operative complications presenting with non-specific symptoms can be challenging to identify. In the case of our patient, the main challenge was to prevent any adrenal crisis intra-and post-operatively. If the diagnosis is made early, it can prevent serious morbidity and on the other hand, any delay in the initiation of treatment can lead to serious mortality. Previous literature reports 5-10 cases of adrenal crisis per 100 patient-years in patients suffering from chronic adrenal insufficiency. The mortality rate from an adrenal crisis is reported to be 0.5/100 patient-years [17].

A case series published in 1979 highlighted that 5 out of 4364 adult patients who underwent cardiac surgical procedure i.e. coronary bypass grafting on CPB machine experienced non-specific abdominal and neurological symptoms ultimately establishing acute adrenal insufficiency. None of them had previous underlying Addison's disease or endocrine hypofunction. This correct diagnosis was suspected on post operative day 4 to 10 (average day 19) and was proved on post operative day 14 to 42 (average day 21) [18]. This suggests that post cardiac surgery development of adrenal crisis although being a rare complication should be considered with a non-specific presentation of a patient as early intervention can improve patient outcomes. Another case report documented adrenal insufficiency after cardiac surgery even after the patient

was given sufficient pre-operative dose [19].

Patients with comorbidities are more vulnerable to adrenal crisis, notably those with asthma and diabetes. In our case, the patient suffered from DM type I, and we used variable rate insulin infusion to correct blood sugars and to keep them between 150-200 mg/dL. Volume status and urine output should be used to guide resuscitation.

According to recent guidelines, the recommended doses for intra- and post-operative steroid cover in adults receiving adreno-suppressive doses of steroids are as follows. Intraoperatively, for a major surgery, hydrocortisone of 100mg IV at induction, followed by immediate initiation of a continuous infusion of hydrocortisone at 200mg/24h. Alternatively, dexamethasone 6–8 mg intravenously, if used, will suffice for 24h. Post operative steroid replacement includes hydrocortisone 200 mg/24h. by IV infusion while nil by mouth (alternatively, hydrocortisone 50 mg every 6 h by intramuscular injection). It is suggested to resume enteral glucocorticoid at double the pre-surgical therapeutic dose for 48 h if recovery is uncomplicated, otherwise continue double oral dose for up to a week [15].

If there is a doubt about the need for glucocorticoids, they should be given as there are very rarely serious long-term adverse consequences of short-term glucocorticoid administration [20].

4. Conclusion

In conclusion, this case underscores the complexity of managing multiple endocrine disorders in conjunction with a congenital heart defect, necessitating a multidisciplinary approach involving endocrinologists, cardiologists, cardiac surgeons, and anesthesiologists. The successful surgical closure of the large ASD required thorough perioperative management to prevent adrenal crisis and optimize glycemic control. This involved careful adjustment of steroid and insulin therapy, close monitoring of hemodynamic parameters, and collaboration among specialties to ensure optimal patient outcomes.

Abbreviations

PAI	Primary Adrenal Insufficiency
ASD	Atrial Septal Defect
DM	Diabetes Mellitus
SCARE	Surgical CAse Report
BMI	Basal Metabolic Index
ACTH	Adrenocorticotropic Hormone
HBA1c	Hemoglobin A1c
17 OH	Serum 17-Hydroxyprogesterone
GAD-65	Anti-glutamic Acid Decarboxylase Antibody
LVEF	Left Ventricular Ejection Fraction
RHC	Right Heart Catheterization
BP	Blood Pressure
IV	Intravenous
MAC	Minimum Anesthetic Concentration
CPB	Cardiopulmonary Bypass
RA	Right Atrium
MAP	Mean Arterial Pressure
SVR	Systemic Vascular Resistance
HPA	Hypothalamic-Pituitary-Adrenal Axis

Author Contributions

Faiza Qureshi: Conceptualization, Data curation, Investigation, Project administration, Resources, Writing – original draft, Writing – review & editing

Akbar Mistry: Conceptualization, Investigation, Writing – review & editing, Supervision

Syeda Kainat Fatima: Project administration, Writing - review & editing

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Conflicts of Interest

The authors have no conflict of interest to declare.

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