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Case Report

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Perioperative Care of a Patient With Carney Complex

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Abstract

Carney complex is a multiple endocrine neoplasia syndrome, commonly affecting the thyroid, adrenal, and pituitary glands. In addition to endocrine involvement, tumors and myxomas may develop including cardiac myxomas and schwannomas. Approximately 70% of cases result from autosomal dominant germline mutation, with the remaining 30% representing *de novo* spontaneous mutations. Carney complex remains extremely rare, with only approximately 750 cases reported worldwide since 1985. We present an adolescent with Carney complex who underwent left atrial myxoma resection, followed by bilateral adrenalectomy a few years later. The perioperative implications of Carney complex are presented, previous reports of anesthetic care in these patients reviewed, and options for anesthetic management discussed.

Keywords: Carney complex; Atrial myxoma; Endocrine neoplasia; Schwannoma; Pediatric anesthesiology

Introduction

Carney complex is a rare multiple endocrine neoplasia characterized by involvement of multiple end-organs, most commonly the thyroid, adrenal, and pituitary glands. Beyond its endocrine manifestations, Carney complex is associated with a spectrum of tumors and lesions, including cardiac and cutaneous myxomas, testicular and ovarian tumors, melanocytic schwannomas, and breast myxomatosis [1, 2]. Among these, cardiac myxomas and schwannomas are the leading contributors to morbidity and mortality in affected individuals. Cutaneous findings are also frequent, with pigmented lesions such as lentigines, cafe-au-lait macules, and blue nevi, as well as conjunctival pigmentation, commonly observed. Approximately 70% of Carney complex cases are attributed to autosomal dominant germline mutations, while the remaining 30% arise *de novo* from spontaneous mutations. While the exact prevalence remains unknown, Carney complex is extremely rare, with an estimated 750 cases reported worldwide.

Carney complex was first described by Dr. J. Aidan Carney in 1985 as "the complex of myxomas, spotty pigmentation, and endocrine overactivity," and was officially designated as Carney complex in 1986 [3, 4]. Surgical intervention may be required for resection of cardiac myxomas, endocrine tumors, or schwannomas, often necessitating anesthetic management tailored to the specific pathophysiologic features of the syndrome [5]. We present an adolescent with Carney complex who underwent left atrial myxoma resection, followed by bilateral adrenalectomy a few years later. The perioperative implications of Carney complex are presented, previous reports of anesthetic care in these patients reviewed, and options for anesthetic management discussed.

Case Report

Review of this case and presentation followed the guidelines of the Institutional Review Board of Nationwide Children's Hospital (Columbus, OH). Written, informed consent was obtained for use of de-identified information for publication.

A 14-year-old female presented to her primary care physician with a several-month history of persistent cough, exertional dyspnea, fatigue, palpitations, and chest discomfort associated with exertion. On physical examination, she had a split S1, split S2 with a "plop" on cadence more prominent while upright, a II/VI vibratory systolic murmur at the left lower sternal border while supine, and a II/VI diastolic murmur at the apex while upright. Echocardiogram revealed a large obstructive mass within the left atrium obstructing mitral valve inflow with a mean gradient of 15 mm Hg. The tumor measured 7.5 × 5 cm and its echocardiographic appearance was suggestive of a myxoma. She was admitted for surgical resection of a presumed left atrial myxoma. At the time of the preoperative evaluation, she weighed 52.9 kg. Preoperative vitals included a heart rate of 110 beats/min, blood pressure 108/63 mm Hg, and room air oxygen saturation of 100%. Her airway assessment revealed a normal thyromental distance, normal mouth opening, and a Mallampati 1 examination. The remainder of the physical examination was unchanged from her previous examination at her primary care physician (PCP). She denied any drug allergies and was on no medications. She was assigned an American Society of Anesthesiologists (ASA)

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physical status IV. She was held *nil per os* for 8 h and transported to the operating room where routine ASA monitors were placed. Premedication included midazolam (4 mg in divided doses). The induction of general anesthesia was achieved with the intravenous (IV) administration of dexmedetomidine (4 µg), fentanyl (7.5 µg/kg), and midazolam (4 mg). Neuromuscular blockade was achieved with rocuronium (100 mg). After the induction of anesthesia, the patient's trachea was intubated with a 7.0 mm cuffed endotracheal tube. A second peripheral IV cannula, a radial arterial cannula, and a triple lumen central venous catheter were placed. Maintenance of anesthesia was achieved with dexmedetomidine (0.5 µg/kg/h) and isoflurane (exhaled concentration 0.5-1%). Ongoing neuromuscular blockade was provided by intermittent doses of rocuronium. Acute normovolemic hemodilution (ANH) blood volume was achieved by the removal of 450 mL of whole blood. Anticoagulation was achieved with heparin and following median sternotomy, the patient was placed on cardiopulmonary bypass with aortic cross-clamp and the left atrial myxoma was excised. Transesophageal echocardiogram revealed normal left and right ventricular function with no residual tumor. Following rewarming to 37 °C, the aortic clamp was released after 72 min and the patient was separated from cardiopulmonary bypass after 106 min. Residual heparinization was reversed with protamine and the autologous whole blood was reinfused at this time. There were no occurrences of perioperative hypoxia, and there were no intraoperative problems. No allogenic blood products were administered intraoperatively. Residual neuromuscular blockade was reversed with sugammadex (200 mg). When awake, the patient's trachea was extubated in the operating room and she was admitted to the cardiothoracic intensive care unit (CTICU). She was transferred to the inpatient ward on postoperative day (POD) 1. Her postoperative course was unremarkable, and she was discharged home on POD 4.

Following this procedure, the patient underwent genetic testing for Carney complex due to a history of cutaneous lesions and prior removal of an ear mass (trichofolliculoma) at 7 years of age. Results confirmed the diagnosis of Carney complex with a pathological variant in PRKAR1A. She began regular surveillance with cardiology follow-up every 6 months and annually with oncology. Given that she was post-pubertal and had one previous Carney complex-related tumor (atrial myxoma), periodic monitoring for cardiac, dermatologic, endocrine, thyroid, and gonadal tumors was recommended. Surveillance measures included echocardiogram and electrocardiogram (ECG) every 6 months, annual thyroid ultrasound, baseline abdominal ultrasound for evaluation of the ovaries, full body skin exam with dermatology every 1 - 2 years, and annual screening labs including urine free cortisol and serum insulin-like growth factor 1 (IGF-1) levels. If serum cortisol levels became abnormal, the plan included further testing with diurnal urine cortisol studies, a dexamethasone suppression test, and adrenal computed tomography (CT) imaging. Abnormal IGF-1 levels would prompt a pituitary magnetic resonance imaging (MRI), 3-h glucose tolerance test, and 90-min thyrotropin-releasing hormone (TRH) stimulation test.

Over the following 3 years of routine cardiology and oncology follow-up, a thyroid ultrasound revealed a hypoechoic thyroid cyst, although thyroid-stimulating hormone (TSH) and free T_4 levels remained within normal limits. Approximately 36 months after removal of the atrial myxoma, the patient presented to oncology clinic with persistent fatigue, weight gain, acne, mild hirsutism, facial plethora, and irregular menses. On physical examination, she had mild hypertension with blood pressure readings of 130 - 145/70 - 86 mm Hg. Laboratory studies showed elevated urinary free cortisol levels on two occasions (82.9 and 114.9 µg/dL; normal ≤ 56 µg/dL). Further evaluation documented hypercortisolism, confirmed by two elevated salivary midnight cortisol values and an elevated morning cortisol level (dexamethasone suppression test). CT imaging of the abdomen and pelvis with contrast was obtained, showing regions of nodularity bilaterally within the adrenal glands, concerning for primary pigmented nodular adrenal dysplasia (PPNAD), a known endocrine manifestation of Carney complex. Additionally, a Liddle's test, which consists of sequential low-dose and high-dose dexamethasone suppression, confirmed the diagnosis of PPNAD, as evidenced by the characteristic paradoxical rise in urinary cortisol excretion in response to dexamethasone.

With confirmation of Cushing's syndrome secondary due to PPNAD, the patient, now 17 years old, was scheduled for a robotic bilateral adrenalectomy. The preoperative evaluation revealed an 82 kg adolescent in no acute distress. Preoperative vital signs included blood pressure (127/91 mm Hg), SpO_2 (98%), and heart rate (84 beats/min). The patient was held *nil per os* for 8 h. She was premedicated with hydrocortisone (100 mg) and transported to the operating room where routine ASA monitors were placed. Anesthesia was induced by the IV administration of propofol (200 mg), fentanyl (100 µg), and lidocaine (100 mg). Neuromuscular blockade was provided by rocuronium (50) and her trachea was intubated with a 7.0 mm cuffed endotracheal tube. Following the induction of anesthesia, a second peripheral IV cannula and a radial arterial cannula were placed. Maintenance anesthesia included dexmedetomidine (0.5 µg/kg/h), ketamine (0.25 mg/kg/h), sevoflurane (expired concentration 1.5-3%), and intermittent hydromorphone. Ongoing neuromuscular blockade was provided by a rocuronium infusion (0.4 - 0.7 mg/kg/h). There were no significant intraoperative events during the 6-h surgical procedure. Intraoperative fluids included 2,500 mL of isotonic crystalloid. Urine output was 350 mL with an estimated blood loss of 100 mL. After completion of the surgical procedure, residual neuromuscular blockade was reversed with sugammadex and the patient's trachea was extubated in the operating room. She was admitted postoperatively to the pediatric intensive care unit (PICU) for ongoing cardiorespiratory monitoring. Perioperative stress dosing of hydrocortisone was continued (150 mg/day). Postoperative pain was managed with IV acetaminophen and patient-controlled analgesia with hydromorphone. She was transferred to the inpatient ward on POD 2. She developed a left parapneumonic effusion that required tube thoracostomy drainage and the administration of IV antibiotics. On POD 8, she developed diarrhea and was found to be positive for *C. difficile* toxin and was subsequently treated with a 10-day course of oral vancomycin. The remainder of her postoperative course was unremarkable, and she was discharged home on POD 10, receiving maintenance hydrocortisone (10 mg in the morning, 5 mg midday, and 5 mg at bedtime) and daily fludrocortisone

(0.1 mg). Routine cardiac surveillance, thyroid ultrasound, and laboratory studies have been continued on a consistent basis.

Discussion

Carney complex is a genetic disorder with dysregulation of the cyclic AMP (cAMP)-dependent protein kinase A (PKA) signaling pathway. Inactivating mutations in the protein kinase cAMP-dependent type I regulatory subunit alpha (PRKAR1A) gene, which encodes a regulatory subunit of PKA, are the primary cause. Dysregulation of the pathway leads to various tumors (benign and malignant) and other clinical manifestations including skin pigmentation abnormalities including nevi and lentigines.

Patients with Carney complex can present several challenges during anesthetic care. However, given its relative rarity, reports outlining perioperative concerns of these patients are limited [5-7]. As with all anesthetic care, the perioperative plan begins with a thorough preoperative evaluation to identify ongoing medical problems and comorbid end-organ involvement related to Carney complex. The preoperative physical examination follows the general process of airway, respiratory, and cardiac examinations. As noted in our patient, the airway examination in the majority of patients with Carney complex is generally straightforward. However, over-production of growth hormone from a pituitary adenoma may occur in 10-15% of the patients, presenting as acromegaly with its potential impact on airway management [8-11]. Clinical features of acromegaly may include macroglossia, prognathism, enlargement and distortion of the glottis structure, hypertrophy of the soft tissues of the larynx and pharynx, and longer thyromental distance. Mask ventilation in patients with acromegaly can be anticipated to be more difficult and the incidence of difficult endotracheal intubation is higher. When problematic airway management is anticipated, the appropriate equipment, including indirect video laryngoscopy, should be available in the operating room [12, 13].

Cardiac complications, particularly atrial myxomas, represent the most frequent life-threatening manifestations of Carney complex. More than half of affected individuals succumb to cardiac-related causes, with atrial myxomas accounting for approximately 25% of these fatalities. Unlike sporadic (non-familial) cardiac myxomas, which typically present in middle age and have a female predominance, myxomas associated with Carney complex tend to arise earlier, often during the second or third decade of life, and affect both sexes equally. The majority (75%) of myxomas are located in the left atrium, typically near the fossa ovalis. The remainder are found in the right atrium (15-20%), and less commonly, the ventricles (5-10%). Embolism occurs in up to 40% of patients, most frequently involving the central nervous system and presenting as stroke or transient ischemic attacks. Familial myxomas also carry a higher risk of recurrence following surgical resection. Those located within the right atrium pose a particular risk of embolization during placement of central venous devices. Depending on their size and anatomical location, cardiac myxomas may give rise to additional clinical manifestations, including inflow or outflow tract obstruction, arrhythmias, and systemic embolization [14-16]. Myxomas located in the left atrium can impede pulmonary venous return, potentially leading to the development of pulmonary hypertension [17]. The perioperative implications are significant, as highlighted by reports of sudden death following non-cardiac surgery, attributed to unrecognized atrial myxoma embolization [18]. As such, all patients with Carney complex undergoing surgical procedures, regardless of symptomatology, should have a recent transthoracic echocardiogram (within the preceding 12 months) to assess for the presence, size, location, and mobility of cardiac myxomas. If echocardiographic findings are inconclusive, imaging is suboptimal, or if additional anatomic detail is required, cardiac MRI may be beneficial [19]. As noted in our patient, nonspecific symptoms such as fatigue or shortness of breath may be the only presenting sign of an atrial myxoma. Therefore, annual echocardiographic surveillance is recommended for all individuals with Carney complex. Preoperative electrocardiography is indicated to evaluate for arrhythmias with more prolonged monitoring (24-h Holter) in symptomatic patients.

Additional cardiac complications in patients with Carney complex may arise secondary to endocrine abnormalities, including manifestations of acromegaly, Cushing syndrome, and hypertension. In the context of excessive growth hormone secretion (acromegaly), myocardial hypertrophy can develop, leading to diastolic dysfunction and, in more advanced cases, progression to left ventricular systolic dysfunction and congestive heart failure [20, 21]. Hypertension in Carney complex may result from hypercortisolism associated with Cushing syndrome, further contributing to cardiovascular strain. These endocrine-related cardiovascular manifestations underscore the importance of thorough preoperative assessment of cardiac function. A recent echocardiogram should be obtained to evaluate ventricular function, valvular integrity, and signs of hypertrophy. Additionally, serial blood pressure monitoring is essential in the perioperative period, especially for patients with a known history of endocrine dysfunction or hypertension. Intraoperative management may require placement of arterial and central venous catheters to facilitate blood pressure control, guide fluid management, and optimize cardiac contractility during the procedure [22, 23].

Respiratory complications in Carney complex have been less frequently reported in the literature. Perioperative respiratory concerns are primarily related to the end-organ comorbidities of a left atrial myxoma or endocrine manifestations, most prominently acromegaly. As noted previously, left atrial myxomas can cause mitral outflow obstruction while chronic embolization of a right atrial lesion may result in pulmonary hypertension. Obstructive sleep apnea (OSA) is a common comorbid condition in patients with acromegaly related to upper airway involvement. Preoperative screening tools, such as the STOP-BANG questionnaire, should be administered to patients suspected of OSA [24, 25].

Endocrine manifestations must also be evaluated, as they can significantly influence anesthetic planning. In addition to hypertension, Cushing syndrome from PPNAD may result not only in hypertension, but also glucose intolerance. Following surgical resection of adrenocortical tumors, stress dosing of hydrocortisone and continuation of maintenance doses of fludrocortisone are indicated to avoid fluid and elec-

trolyte disturbances or cardiovascular instability. Thyroid involvement may include the development of thyroid nodules, hyperplasia, and malignancies. Periodic imaging (ultrasound) is indicated as part of routine surveillance. Evaluation of thyroid function (TSH, T_4) may be indicated based on clinical symptomatology.

In summary, Carney complex is a rare autosomal dominant multiple endocrine neoplasia syndrome characterized by a constellation of findings, including spotty skin pigmentation (e.g., lentigines), cardiac myxomas, and endocrine hyperactivity involving various organ systems such as the pituitary, adrenal glands, peripheral nerves, testes, and ovaries. Prior to its formal description in 1975 by Dr. Carney, the clinical manifestations were grouped into acronyms such as NAME (nevi, atrial myxoma, myxoid neurofibroma, and ephelides) or LAMB (lentigines, atrial myxoma, and blue nevi) [26]. Perioperative management in patients with Carney complex often involves surgical intervention for cardiac myxomas or endocrine tumors, particularly of pituitary or adrenal origin. Given the rarity of the disorder, published reports on anesthetic and perioperative considerations remain limited and largely anecdotal. However, careful preoperative evaluation is essential, with attention to end-organ involvement, particularly cardiac function in the presence of myxomas and the systemic effects of endocrine hyperactivity such as acromegaly or Cushing syndrome. A multidisciplinary approach, incorporating cardiology, endocrinology, and anesthesiology, is critical to optimize perioperative outcomes in this complex patient population.

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None to declare.

Conflict of Interest

None to declare.

Informed consent

Informed consent was obtained for hospital/anesthetic care and the use of de-identified information for publication.

Author Contributions

DF: preparation of initial, subsequent, and final drafts; KM: direct patient care and review of final document; BLW: review of drafts and final document; JDT: concept, writing, and review of all drafts.

Data Availability

Any inquiries regarding supporting data availability of this study should be directed to the corresponding author.

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