Reviews

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Anesthesia Management in Hereditary Pheochromocytoma and Paraganglioma: Updated Insights into Clinical Features and Perioperative Care

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ABSTRACT

Approximately 40% of pheochromocytoma and paraganglioma (PPGL) cases are familial, typically presenting earlier with more complex symptoms. This paper synthesizes literature and guidelines to inform on clinical characteristics and perioperative care for PPGL. Pheochromocytoma in von Hippel-Lindau (VHL) disease exhibits heightened secretion activity without significant perioperative hemodynamic changes. Tumors in multiple endocrine neoplasia type 2 (MEN2) have a stronger endocrine function, which may induce hemodynamic fluctuations during surgery. Therefore, pheochromocytoma screening is essential at all stages of MEN2. Neurofibromatosis type 1 (NF1) often presents multisystem lesions and can result in difficult airway. Pheochromocytoma should be evaluated when NF1 patients present hypertension. Pheochromocytoma and paraganglioma type 5 may present multiple lesions of pheochromocytoma or paraganglioma. In summary, hereditary PPGLs may present with severe lesions in other systems, beyond tumor function. A multi-disciplinary team (MDT) approach is often invaluable in perioperative management.

Key words: pheochromocytoma; paraganglioma; genetic disease; anesthetic management; hemodynamic; perioperative care

INTRODUCTION

Pheochromocytoma (PCC) is a rare endocrine tumor with an annual incidence of 2–9 per 100,000 person-years. Paraganglioma is another rare neuroendocrine tumor arising from extra-adrenal autonomic paraganglia, predominantly paravertebral. All guidelines recommend that patients scheduled for pheochromocytoma and paraganglioma (PPGL) surgery should receive appropriate drug preparation[1-5]. However, even with adequate preparation, severe hemodynamic fluctuations may still occur during the perioperative period, making PPGL resection a high-risk surgery.

Approximately 40% of PPGL cases are familial, often presenting earlier with complex symptoms and a potentially higher long-term recurrence rate[1, 6, 7]. All inherited PPGLs result from autosomal dominant mutations and can be classified into von Hippel-Lindau (VHL) disease, multiple endocrine neoplasia type 2 (MEN2), neurofibromatosis type 1 (NF1), paraganglioma syndrome types 1–5 (PGL1-5), and conditions caused by other mutations (MAX, TMEM127, and FH)[7-10]. This review synthesizes characteristic lesions and anesthesia management advancements in hereditary PPGL from literature and guidelines, offer-

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ing insights for clinical features and perioperative care.

VON HIPPEL-LINDAU (VHL) DISEASE

VHL disease, caused by mutations in the VHL gene on chromosome 3, is a multisystem tumor syndrome including retinal hemangioblastoma, central system hemangioblastoma, clear cell renal cell carcinoma, and pheochromocytoma[11, 12]. The average annual incidence is about 1 in 40,000[13]. It can be divided into two types, among which type 2 is often accompanied by pheochromocytoma (**Table 1**), which is present in 10%–20% of the patients[14-16]. Interestingly, 30% of these patients exhibit no hypertension or pheochromocytoma symptoms[17]. Patients with VHL disease tend to be younger, frequently present with bilateral tumors, and may have extra-adrenal

Table1. Characteristics of four hereditary pheochromocytoma syndromes

Features	VHL	MEN2		NF	PGLs
Subtype with PPGL	VHL2	MEN2A	MEN2B	NF1	PGL4 (Typical)
Incidence	1/40,000	1/40,000	1/1,000,000	1/3,000	–
Mutation site	VHL	RET	RET	NF1	SDHB
Principal feature	Multisystem tumor	MTC and PCC	Extremely dangerous in MTC	Neurofibroma café-au-lait macules Lisch nodules	Multiple lesions
Feature of PPGL					
Incidence (%)	10 – 20	50	50	0.1 – 5.7	25
Mean age of onset(y)	30	38	25	42	31
Unilateral (%)	55	35	35	85	79
Metastasis rate (%)	3.5	3	3	10	34
Character of tumor	Ongoing secretion of norepinephrine permanent hypertension	Paroxysmal hypertension	Paroxysmal hypertension	Screening PCC when combined with hypertension	Mainly originate from sympathetic paraganglia
Secretion function	Strong	Strong	Strong	–	–
Anesthesia consideration					
Preoperative					
Circulatory system	Myocardial injury	Myocardial injury	Myocardial injury Marfan syndrome	Myocardial injury Cardiovascular anomaly	Myocardial injury
Respiratory system	–	–	Latent anomaly	Impaired	–
Airway	–	MTC involved	MTC involved Skeletal deformity	Occupying lesions	Difficult airway after radiotherapy
Others	Intracranial hemorrhage; maintain stable intracranial pressure; avoid spinal anesthesia	Hyperparathyroid crisis	Gastrointestinal symptoms	Cognitive dysfunction; urinary system obstruction; gastrointestinal bleeding	–
Intraoperative hemodynamic fluctuation*	Similar	Remarkable	Remarkable	–	–

* compared with non-syndromic; VHL: von Hippel-Lindau disease; MEN: multiple endocrine neoplasia; NF-1: neurofibromatosis type 1; PCC: pheochromocytoma; MTC: medullary thyroid cancer; PGLs: paraganglioma syndromes; PPGL: pheochromocytoma and paraganglioma; RET: rearranged during transfection; SDHB: succinate dehydrogenase subu B; –: not available

tumors[6]. All patients exhibit elevated plasma normetanephrine levels while maintaining normal metanephrine levels. Clinically, persistent hypertension is common due to continuous catecholamine secretion [12, 13, 18].

Previous studies have found that, compared with non-syndromic pheochromocytoma patients, those with VHL disease experience earlier pheochromocytoma onset and enhanced secretion function. Patients have elevated urinary norepinephrine levels. It will take longer time to prepare for surgery and the procedure time can also be prolonged. However, there is no significant difference in hemodynamic fluctuations before or after tumor resection. In addition, patients with VHL disease are at risk for medullary hemangioblastoma rupture, necessitating careful attention to the potential for intracranial hemorrhage. Blood pressure can affect intracranial pressure (ICP) and intraocular pressure[19]. Pheochromocytoma resection with epidural analgesia is viable, but preoperative imaging exams are necessary[20-23]. Epidural anesthesia should be considered in priority and spinal anesthesia should be avoided when selecting neuraxial anesthesia for patients with intracranial mass or at risk of elevated ICP. ICP changes cause by dural puncture should be monitored[24, 25].

MULTIPLE ENDOCRINE NEOPLASIA TYPE 2

Multiple endocrine neoplasia syndrome, a genetic disease that manifested as two endocrine gland tumors at least in the RET proto-oncogene, is usually divided into type 1 (MEN1) and type 2 (MEN2) (**Table 1**). Unlike MEN1, 50% of MEN2 patients have pheochromocytoma, which is mostly bilateral but rarely extra-adrenal pheochromocytoma[6]. MEN2 can be further divided into type 2A and type 2B. Pheochromocytoma incidence and MTC risk vary by mutation location. The 634 codon mutation leads to earlier onset, while M918T confers the highest MTC risk[26-28]. Surgery is the preferred treatment for MEN2. Current guidelines recommend prioritizing pheochromocytoma screening in MEN2 patients with a definite diagnosis[28, 29]. Despite MTC commonly being the first sign of MEN2, concurrent diagnosis of MTC and pheochromocytoma occurs in 40% of patients[30]. Some patients undergoing other surgeries may have undetected or surgically untreatable concurrent pheochromocytoma..

In hereditary pheochromocytoma diseases, MEN2 has the highest incidence, warranting pheochromocytoma screening for MEN2 patients at any disease stage. A retrospective study of 61 patients found that MEN2 patients presented with earlier onset and smaller tumor sizes, yet experienced similar intraoperative hypertension severity to non-syndromic pheochromocytoma patients, with a lower incidence and severity of postoperative hypotension[31]. Tumors larger than 5cm are an independent risk factor for hemodynamic instability during operation[32]. A medical center, without prior MEN2A anesthetic experience, found intraoperative hemodynamic instability more severe in MEN2A based on non-syndromic pheochromocytoma experience[33]. Besides, MEN2 patients with inoperable pheochromocytoma but requiring urgent surgery for other lesions may receive non-pheochromocytoma resection following risk-benefit assessment, adequate preparation, and multidisciplinary collaboration. However, even with thorough preoperative preparation, these patients have longer ICU stay and a higher incidence of pheochromocytoma-related complications[34]. Primary hyperparathyroidism is a characteristic lesion of MEN2A, requiring vigilance for hypercalcemia crises despite their low incidence. A recent study found that MEN2B patients may be prone to restrictive, diffuse, and obstructive pulmonary dysfunction for reasons beyond just skeletal deformities[35].

NEUROFIBROMATOSIS TYPE 1

Neurofibromatosis type 1 (NF1), which is caused by mutations in the *NF1* gene on chromosome 17, typically present with skin and nerve tissue lesions as café-au-lait macules, skinfold freckling, Lisch nodules, peripheral nerve tumors, and central nervous system tumors (**Table 1**). Besides, it affects the cardiovascular system, skeletal system, digestive system, and behavioural deficits[36-40]. The incidence of pheochromocytoma in NF1 is about 0.1% to 5.7%. The tumor onsets later and most is unilateral, but more likely to metastasize. Pheochromocytoma should be evaluated when patients present with hypertension[41, 42].

The incidence of pheochromocytoma in NF1 is low, and still lack of comparison with non-syndromic pheochromocytoma. NF1 patients had more intraoperative hemodynamic fluctuations and more complications due to tumor size during open resection[43]. Moreover, multisystem lesions in NF1 are more complex, posing multifaceted challenges for anesthesiologists. NF1 can lead to tumors in head, neck, and pharyngeal cavity, complicating airway management due

to organic obstruction or nerve plexus involvement. Patients with symptoms such as dysphagia, dysarthria, wheezing, or voice changes should be particularly cautious for potential upper airway obstruction. Skeletal deformities, including facial deformities, cervical rigidity, and osteoporosis, contribute to difficult airway. In addition, respiratory system can also be affected, and pulmonary function assessment should be a focus in preoperative examinations. Given potential neurological lesions or tumors, general anesthesia is considered a safer option. Cardiovascular issues, cognitive impairments, and urinary tract obstructions require careful consideration[41, 44-46].

PARAGANGLIOMA SYNDROME TYPES 1 – 5

Most genetic paragangliomas are associated with genes encoding succinate dehydrogenase (SDH) complex subunits, with paragangliomas arising from either parasympathetic or sympathetic paraganglia. Sympathetic paragangliomas can develop anywhere along the sympathetic chain, from the basis cranii to the prostate. Parasympathetic paragangliomas are located in the neck and basis cranii along the glossopharyngeal and vagus nerve branches, with carotid body tumors being the most common manifestation. Parasympathetic paragangliomas grow more slowly than sympathetic paragangliomas and PCCs, with reduced secretory function[3, 14, 47, 48]. Five paraganglioma syndromes (PGL 1–5) have been identified, each with distinct mutation sites and tumor characteristics, potentially associated with pheochromocytomas, renal cell carcinomas, gastrointestinal stromal tumors, pituitary tumors, among others[3, 14, 48].

Patients with SDHx primarily present with PGL, and some may have multiple PGLs, necessitating comprehensive examination during the perioperative period. Surgical risk is associated with the site of tumor invasion[49]. Different from PCC, paragangliomas primarily secrete norepinephrine and/or dopamine. SDHB (PGL4) is the only one principally originates from sympathetic paraganglia (**Table 1**). Dopamine-secreting PGLs may not need preoperative alpha-adrenoceptor blockade, but they carry a higher risk of malignancy. Therefore, clinically and biochemically silent paraganglioma should be considered with suspension [50, 51]. Patients with head and neck paragangliomas, previously irradiated, require heightened awareness for difficult airway management.

Summary

Surgery is the primary treatment for PPGL. Patients with PPGL require thorough preparation, personalized medication to ensure blood pressure control, restoration of blood volume, and improvement in hypermetabolic syndrome and glucose metabolism. Pheochromocytoma in hereditary PPGL is more distinctive, necessitating anesthesiologists to prepare for severe hemodynamic changes. Family history inquiry or genetic testing facilitates earlier diagnosis and treatment of the disease. Clinically, it may also be associated with serious lesions in other systems. Thus, multidisciplinary collaboration is essential perioperatively to devise more comprehensive and individualized preoperative preparation for these patients.

ARTICLE INFORMATION

Conflict of interests

The authors declare no conflict of interest.

Authors' contributions

Li YH collected literature and drafted the manuscript. Shen L designed the study and reviewed the manuscript. Li YH and Shen L revised and approved the final version of this article for publication.

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