Phäochromocytom

Hofmann Nikolaus

ANÄSTHESIE FORUM

ALPBACH

REPETITORIUM

Nebennierenmark

- Bildung von sympathomimetischen Hormonen in den chromaffinen Zellen
- Katecholamine: Adrenalin u. Noradrenalin
- Vermehrte Ausschüttung vor allem in Alarmsituationen (körperl.u. psychisch)
- In Ruhe werden nur geringe Mengen freigesetzt

Phäochromozytom

- Tumore, ausgehend von chromaffinen Zellen
- NN – Mark (90 %)
- sympathischen paravertebralen Ganglien
- Leitsymptom:
  - anfallsartige Hypertonie, hypertensive Krise, Kopfschmerz, Herzklopfen, Schwitzen, Angstgefühl, Schwindel, Zittern, Krämpfe
- Attacken dauern ca. 15 Minuten

Phäochromozytom

- Insgesamt sehr selten, 0,1 % der Hochdruckursachen
- DD bei der Tako-Tsubo Cardiomyopathie
- Nachweis: biochemisch, Bestimmung der freien Katecholamine im 24 - Stunden Urin, zusätzlich Best. der Vanilinmandelsäure
- Bildgebende Nachweisverfahren: CT, MRT
- Nuklearmedizinisch: MIBG - Szintigraphie

MIBG - Szintigraphie

- J 131 – Metajodobenzylguanidin
- Molekulare Struktur dem Noradrenalin ähnlich
- MIBG wird aktiv im chromaffinen Gewebe und adrenalen Nervengewebe angereichert
- 85 – 90 % der Phäochromozytome nachweisbar
- Vor allem zur Darstellung ektop gelegener Phäochromozytome und Metastasen
- CT bzw. MRI zur Darstellung der NN – Tumore überlegen

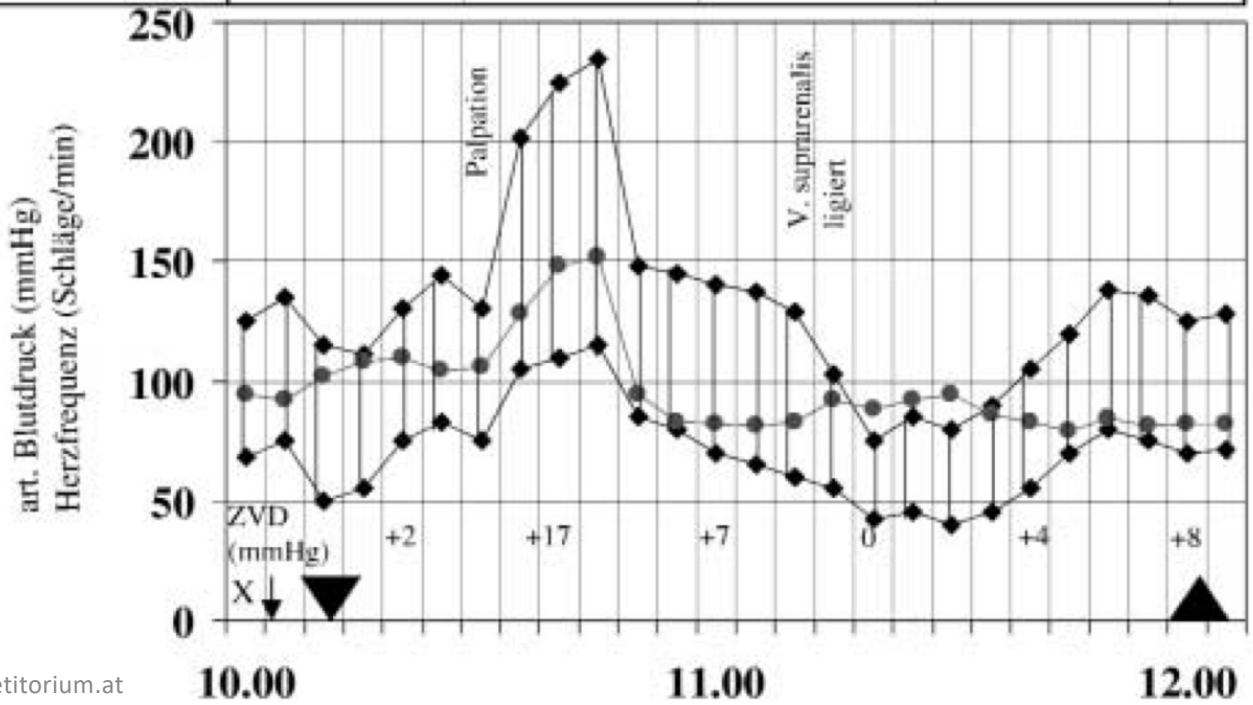
Anästhesie

- Präoperativ
  - Alpha – Blocker
    - Phenoxybenzamin: alpha 1 u. alpha 2 Blocker
    - Reflextachykardie
    - Expansion des Plasmavolumens
- Beta – Blocker
  - Atenolol, Metoprolol
- Kardiale Diagnostik: EKG, Echokardiographie, Szintigraphie
- Blutdruckprofil

Anästhesie

- Intraoperativ
  - Überschießende Reaktion auf schmerzhafte Stimuli
  - Cave okulte Hypovolämie
  - Cave PDA, protrahierte Hypotension
    Katecholaminmangel, Rezeptor - Down – Regulation
  - Hypertensive Episoden: klass. Antihypertensiva
    - Spez.: Magnesiumsulfat 40-60 mg/kgKG als Bolus
- Postoperativ
  - Persistierende Hypertension / Hypotension
  - Hypoglycämie

Sufentanil	(µg)	20	10 10		10
Etomidat	(mg)	12			
Rocuronium	(mg)	25	15	15	10
O_2	(l/min)	6 -1- 0,4----------			----6
N_2O	(l/min)	2- 0,5----------			----|
Isofluran	(Vol.-%)	0,5-- 0,7 --------	1,0 -------0,6 -------0,5		----|
Glyzerolnitrat	(µg/kg x min)		1-0,4----------|		
Esmolol	(µg/kg x min)		500-50- 2 ----------|		
Noradrenalin	(µg/kg x min)			0,03 --0,05------0,04------->	



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Clinical Practice

CLINICAL PRACTICE

Perioperative α-receptor blockade in phaeochromocytoma surgery: an observational case series†

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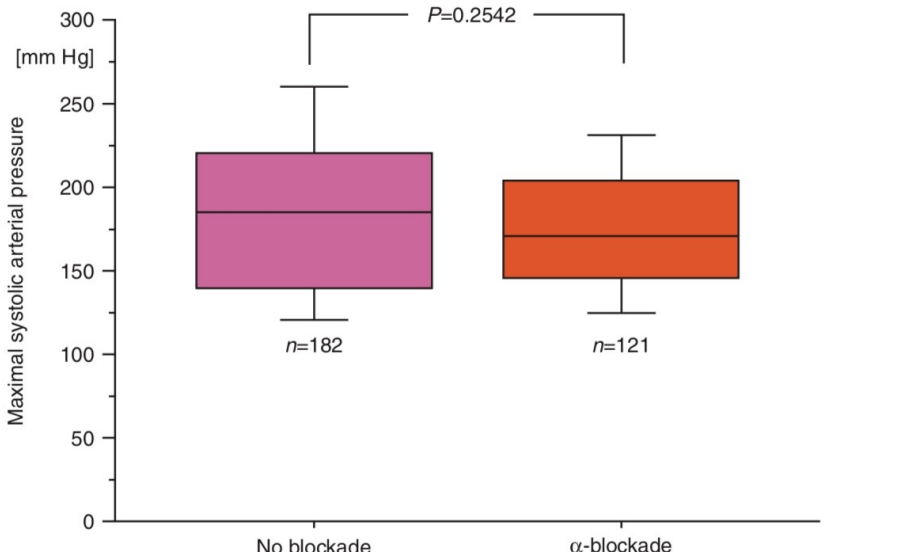

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Mini-Review

Perioperative Management of Pheochromocytomas and Sympathetic Paragangliomas

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Abstract

Pheochromocytomas and paragangliomas (PPGLs) are rare neuroendocrine tumors arising from chromaffin cells of the adrenal medulla or extra-adrenal paraganglia, respectively. PPGLs have the highest degree of heritability among endocrine tumors. Currently, ~40% of individuals with PPGLs have a genetic germline and there are at least 12 different genetic syndromes related to these tumors. Metastatic PPGLs are defined by the presence of distant metastases at sites where chromaffin cells are physiologically absent. Approximately 10% of pheochromocytomas and ~40% of sympathetic paragangliomas are linked to metastases, explaining why complete surgical resection is the first-choice treatment for all PPGL patients. The surgical approach is a high-risk procedure requiring perioperative management by a specialized multidisciplinary team in centers with broad expertise. In this review, we summarize and discuss the most relevant aspects of perioperative management in patients with pheochromocytomas and sympathetic paragangliomas.

Key Words: pheochromocytomas, paragangliomas, perioperative, management

Abbreviations: CT, computed tomography; MIBG, metaiodobenzylguanidine; MRI, magnetic resonance imaging; PPGL, pheochromocytoma/paraganglioma; SDHx, succinate dehydrogenase subunit x; VHL, von Hippel–Lindau.