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FEOHROMOCITOM – TUMOR SA MNOGO LICA

PHEOCHORMOCYTOMA – TUMOR WITH A LOT OF FACES

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SAŽETAK

Feohromocitom je redak tumor srži nadbubrežne žlezde, sa incidencom od 0,8 do 2,3 novo-otkrivena slučaja na million stanovnika godišnje, u opštoj populaciji. Može se javiti u svakom uzrastu i okolnostima, uključujući novorođenčad, trudnice i dr.. ali je češći kod žena u životnom dobu između 20 i 50 godina.

Dijagnoza se bazira na tipičnim simptomima, laboratorijskim analizama i lokalizacionim pretragama - ultrazvuk, kompjuterizovana tomografija (CT), nuklearna magnetna rezonanca (NMR), scintigrafija (MIBG). Preporučuje se i genetski skrining kod sumnje na hereditarnu formu. U 0,1-1% slučajeva dijagnozu postavljaju kardiolozi jer podaci ukazuju da svaku novo-nastalu hipertenziju treba evaluirati u pravcu moguće feohromocitomom indukovane hipertenzije. Biohemijska potvrda dijagnoze feohromocitoma se bazira na prisustvu povišenih vrednosti kateholamina i/ili njihovih metabolite u plazmi ili u urinu. Savremene preporuke polaze od stanovišta da bi inicijalni skrining feohromocitoma trebalo da uključuje i merenje frakcioniranog metanefrina u urini i/ili plazmi. Nakon biohemijske potvrde, mora biti sprovedena lokalizacija tumora. U većini slučajeva feohromocitom se može identifikovati pomoću CT-a, ali je NMR bolji izbor za ekstraandrenalne lokalizacije i male tumore. Ako se sumnja na paragangliom, multiple tumore ili metastaze, scintigrafija pomoću I-MIBG daje visoku specifičnost i senzitivnost u njihovoj detekciji.

Hipertenzija se tipično prezentuje kao hipertenzivna kriza, indukovana stresom, fizičkim naporom, naglim pokretima i sl.. Ali, oko 10-20% pacijenata ima umerenu hipertenziju, bez hipertenzivnih kriza. Osim paroksizmalne hipertenzije, klasični simptomi, kao što su glavobolja, bledilo, palpitacije, anksioznost se javljaju samo kod 50% slučajeva. Oko 5% pacijenata sa feohromocitomom su asimptomatski, što može odložiti dijagnoszu. Zato se za ovaj tumor kaže da ima mnogo lica.

U literaturi se za feohromocitom vezuje pravilo 10%, što znači da u 10% slučajeva može biti: ekstraadrenalna lokalizacija, bilateralan, bez hipertenzije, nasledan, maligan, u dečjem uzrastu. Međutim, razvoj sofisticiranih dijagnostičkih metoda dovodi ovo pravilo u pitanje. Hereditarne forme imaju veću incidencu bilateralnih tumora (20-50%), ekstra-adrenalna lokalizacija se javlja u više od 20% slučajeva i rizik od maligniteta raste i do 40%, prema nekim izvorima.

Preoperativna priprema ima dva glavna cilja:kontrolu krvnog pritiska i restauraciju volumena krvi do normalnih vrednosti, primenom alfa-blokatora u inicijalnom tretmanu

Intraoperativno pred anesteziologom su dva najznačajnija problema: hipertenzija (tokom indukcije u anesteziju i resekcije tumora) i hipotenzija nakon devaskularizacije i ekstirpacije tumora. Takođe, mogu se javiti srčane aritmije kao veliki problem, naročito kod epinefrin-sekretujućih tumora. U cilju uspešnog tretiranja ovih hemodinamskih poremećaja, neophodno je preoperativno uspostaviti invazivni monitoring krvnog i centralnog venskog pritiska. Antihipertenzivi i antiaritmici moraju biti momentalno dostupni za primenu. Zbog mogućih dramatičnih hemodinamskih poremećaja, feohromocitom je nazvan još i “anesteziološka noćna mora”

ABSTRACT

Pheochromocytoma is a rare tumor of adrenal medulla, with an incidence between 0.8 – 2.3 per one million per year, in general population. It can present in any age, in any circumstances, including neonates, pregnancy etc, but is more common in females between 20 and 50 years of age.

The diagnosis is based on typical symptoms, laboratory and localization investigations (echo-sonography, CT, NMR and MIBI). Genetic screening is recommended if suspicion of hereditary forms is present. In 0.1-1% cases, the diagnosis is established by cardiologists. This data suggests that every new-onset hypertension must be evaluated in the direction of possibility of pheochromocytoma - induced hypertension. A specific diagnosis of pheochromocytoma is made biochemically, based on the presence of elevated catecholamines or their metabolites in plasma or in urine. The current recommendations state that initial screening for pheochomocytma should include the measurement of fractionated metanephrines in urine and/or plasma. After biochemical diagnosis has been made, the localization of the tumor must be established. CT identifies pheochromocytoma in the majority of cases, but NMR is a better choice for extra adrenal localization and small ones. If there is a suspicion on paragangliomas, multiple sites or metastasis, scintigraphy with I-MIBG has high specificity and sensitivity for their detection. Typical presentations of hypertension implies hypertensive crises in stressful situations, physical effort or sudden movements. But, about 10-20% patients have moderate hypertension, without hypertensive crisis. Except the paroxysmal hypertension, the classic symptoms such are headache, sweating, pallor, anxiety and palpitations occur in only 50% of patients. Approximately 5% of patients with pheochromocytoma are asymptomatic. These circumstances may delay diagnosis. That’s why this tumor is “tumor with a lot of faces”

According to the literature, with the pheochromocytoma patients one can see the rule of 10 %, which means that 10 % of the tumors have extra adrenal localization, 10 % are bilateral, 10 % are malignant,10 % are presented without hypertension, 10 % occur in children and, 10 % are hereditary. However, the development of sophisticated diagnostic abilities put this rule under the question mark. Recent data suggested that up to 30 % of pheochromocytoma may have a hereditary basis. Hereditary forms have a higher incidence of bilateral tumors (20-50 %), extra adrenal presentation (up to 20 %) and risk of malignancy rises up to 40 %, according to some data.

Preoperative preparation has two main goals: to control blood pressure and to restore blood volume to normal ranges, using alpha-adrenergic blockade as a initial treatment. Intraoperatively, there are two major problems in front of anaesthesiologists: hypertension (during induction in anesthesia and during tumor resection) and hypotension following devascularisation or extirpation of the tumor. In addition, cardiac dysrhythmias may be a big problem, especially in the presence of epinephrine-secreting pheochromocytoma. In order to successfully treat these hemodynamic disorders, invasive monitoring of blood pressure and central venous pressure must be established before the surgical procedure. Antihypertensive and antiarrhythmic agents must be available immediately for infusion. Because of possibility of dramatic hemodynamic changes pheochromocytoma is “anaesthesiologists nightmare”.