Radiological diagnostics in patients with pheochromocytoma – do we need to prepare? Review of the literature

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Abstract:

Introduction: Pheochromocytomas are chromaffin cell tumours derived from the neural crest and they are associated with catecholamine production. Radiological procedures are playing essential role in present diagnostic of adrenal glands. Physicians who send their patients to the radiological examinations should prepare them to have a safe further diagnostic. A well radiologist should be aware of scanning protocols to provide best quality and the safest for the patient radiological examination.

Aim of the study: This article summarizes the current knowledge about radiological imaging of pheochromocytomas and scan procedures. In this paper we also want to answer to the question does a patient with pheochromocytoma need to be specially prepared for radiological procedures.

Description of knowledge: Diagnostic procedures play primary role in present diagnostic and treatment of pheochromocytomas. It is crucial for further diagnostic procedures to locate the tumour and its margins. Ultrasound imaging can be used with success only in big tumours with clinical symptoms. First choice for adrenal gland tumours is always CT. That modality easily shows localisation and tumours smaller than 1 cm. Another method of choice for adrenal imaging is MRI which gives high contrast images between soft tissues. Radiological differentiation of lesions wouldn’t be possible without contrast agents. They are crucial for calculations of washout in CT.

Conclusions: Intravenous administration of non-ionic contrast agent for CT and gadolinium based in MRI is a safe practice for patients with pheos even without α-blocking medication. Only in an intra-arterial iodine-based contrast administration patient should be pharmacologically prepared before examination.

Key words: pheochromocytoma, adrenals glands, contrast agent, incidentalomas, radiology.
Radiological procedures play essential role in present diagnostic. Radiology is a useful tool for further diagnostic and treatment but also, we meet with many difficulties and rare situations such as uncommon syndromes or diseases. That situations require from doctors more work and quick review of their knowledge with the latest guidelines which are frequently changing during clinical practice. New technologies require different approaches and procedures. Nowadays we have fastest scanners, new scanning procedures, better quality of images and safer contrast agents.

In this paper we wanted to answer a question does a patient with pheochromocytoma need to be specially prepared for radiological procedures.

Pheochromocytomas (pheos) are chromaffin cell tumours derived from the neural crest and they are associated with catecholamine production\(^1\). Pheos are rare tumours, with an annual incidence of 2 to 9.1 per 1 million adults and may correspond up to 60% of all adrenal incidentalomas\(^2\). The majority of pheochromocytomas are benign but up to 25% of tumours may be malignant\(^3\). They affects equally males and females and they occur more frequently in the 3rd to 5th decade of life, but in literature there were cases of pheos in any age.\(^3,4\). In children and younger patients, the disease is often hereditary\(^5\), up to 70% of cases\(^6\). Rate of systolic and diastolic hypertensions is 0.2–0.6 in a group of patients with pheos\(^7,8\). Pheochromocytomas may be an accidental finding (incidentalomas) or a a manifestation of hereditary syndromes\(^9\). However 50% of these tumours remain undiagnosed because they are clinically silent during life – they were found post mortem\(^10\).

The primary treatment for a pheochromocytoma is still a surgery. This is why an adequate preoperative evaluation is crucial before surgery for patients\(^11,12\). That evaluation should be very precise and include full patient’s and it’s family history, detailed blood tests, ECG and cardiac ultrasound.

All that preparations and pre-operative medical treatments are made to minimise intra-operative risk by blocking the effects of catecholamines for at least 10–14 days before surgery\(^13\) some authors recommend up to 21 days\(^11\). Pharmacological treatment with \(\alpha\)-blockade has been proven to reduce the number of perioperative complications to less than 3%\(^14\).

A \(\beta\)-adrenoceptor blocker may be used for preoperative control of tachyarrhythmias or angina. \(\beta\)-adrenoceptor blockers should never be employed without first blocking \(\alpha\)-adrenoceptor mediated vasoconstriction\(^13\).

According to polish guidelines the treatment should last 10 - 14 days before surgery and first choice substance is phenoxybenzamine. First dose of phenoxybenzamine 10 mg p.o. 2 × day and slowly increase dose to maximum 1 mg/kg mc./day until achieve BP <140/90 mm Hg or
use doxazosin at first 2 mg p.o. in 1 dose or in 2 divided doses slowly increase the dose till
max. 32 mg/day. In patients with high heart rate it’s wise to add cardio selective β-blockers
after blocking α-adrenoceptor. It is inadvisable to employ labetalol and carvedilol.
Volume contraction associated with chronic vasoconstriction can be seen in patients with
pheos. That’s why proper hydration is very important - pre-operative volume expansion
achieved by saline infusion or increased water intake is recommended to reduce post-
operative hypotension. Prophylaxis from vein thrombosis is mandatory.
In the cases of inoperative and malignant pheos, the chronic medical treatment is the
same as the preoperative treatment. The management of metastatic pheos remains palliative.
In cases that surgical resection is not possible, alternative include external beam radiation,
cryoablation, radiofrequency ablation, transcatheter arterial embolization, chemotherapy, and
radiopharmaceutical therapy.
New molecular targeted therapies that included everolimus, imatinib, sunitinib, had been used
with various results

Diagnostic procedures play primary role in present diagnostic and treatment. Radiological
procedures like CT or MR show anatomical imaging and those images can be
followed by nuclear functional imaging. It is crucial to locate the tumour and its borders also
infiltration to surrounding tissues. Radiological imaging can be also a useful tool in
diagnosing multiple primary tumours or metastatic lesions in patients with various genetic
disorders.
Ultrasound imaging can be used only in tumours already measured in centimetres
because small lesions could not be detected. On ultrasound, pheos have a variable appearance
ranging from solid to mixed cystic and solid to cystic.
Most used method of adrenal imaging is CT scan. That modality easily shows localisation
and tumours, size > 1 cm in size with 87% to 100% sensitivity.
Pheos in CT imaging are often well-defined masses with attenuation values like those of
muscle tissue, measuring approximately 30–40 HU but it can be also less than 10 HU and
the tumour may display more than 60% washout of contrast agents on delayed scanning.
Proper diagnostic CT od adrenals demand administration of a contrast agent -pheos may have
a higher absolute attenuation on the contrast phase and delayed scans allow to calculate the
absolute and relative washout.
The adrenals can be also diagnosed in MRI. An MRI protocol for the adrenals should
consist of both T1- and T2-weighted images. T1-weighted images obtained after intravenous
administration of gadolin contras agents are used to show enhancement patterns of adrenal
lesions. In T2-weighted images we can notice a radiological sign called “light-bulb” because
the tumour is bright and has a signal intensity of CSF. It is recommended to perform a MRI
in large tumors prior to surgery to assess vascular invasion. MRI it is the safest choice for
children and pregnant women because it doesn’t use X-rays.
The main question is can we administer a contrast agent to a patient during CT or MRI after
non-contrast-enhanced phase if we have a doubt, we have found an incidentaloma that could
be a pheo? According to the latest ESUR Guidelines there is no need of special preparation
for intravenous iodine- or gadolinium-based contrast agent in CT or MRI. Otherwise is
recommended in examinations which require intra-arterial iodine-based contrast medium – in
that case we have to orally administer drugs to block α and β-adrenergic receptors.
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