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Cel pracy: Ocena zastosowania badania tomografii komputerowej (*computed tomography* – CT) i tomografii rezonansu magnetycznego (*magnetic resonance imaging* – MRI) w rozpoznawaniu i monitorowaniu leczenia przyzwojaków nienadnerczowych głowy i szyi.
Materiał i metody: Materiał obejmował 20 pacjentów w wieku 24–75 lat leczonych z powodu przyzwojaków nienadnerczowych gowy i szyi. Wszyscy pacjenci byli diagnozowani i monitorowani za pomocą badań CT i MRI. Okres obserwacji wynosił od roku do 7 lat.
Wyniki: Wykryto 29 przyzwojaków (16 tętnicy szyjnej i 13 bębenkowych). Czterech pacjentów miało mnogie guzy. Ośmiu chorych było operowanych, u tych pacjentów ustalono rozpoznanie mikroskopowe. Biopsje guzów nie były diagnostyczne. U pozostałych 12 pacjentów diagnozę postawiono na podstawie objawów klinicznych i badań radiologicznych. Czternastu pacjentów leczono za pomocą radioterapii. W monitorowaniu przebiegu choroby stwierdzono progresję choroby tylko w 1 przypadku.
Wnioski: Autorzy stwierdzili, że rozpoznanie przyzwojaków można ustalić na podstawie oceny klinicznej i wyniku badania radiologicznego. Obie metody – CT i MRI – nadają się do monitorowania leczenia.

Słowa kluczowe: przyzwojaki nienadnerczowe głowy i szyi, tomografia komputerowa, rezonans magnetyczny.

Imaging modalities in diagnosis and treatment monitoring of head and neck non-adrenal paragangliomas

Metody obrazowania w wykrywaniu i monitorowaniu leczenia przyzwojaków nienadnerczowych głowy i szyi

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The goal of the present study was to evaluate computed tomography (CT) and magnetic resonance imaging (MRI) in diagnosis and treatment monitoring of non-adrenal paragangliomas of the head and neck.

Introduction

Non-adrenal paragangliomas, also known as angioneuromas (glomangioneuromas, glomus tumours), are rare neuroendocrine tumours occurring in the head and in the neck. They originate from ganglion cells of the sympathetic and parasympathetic system. However, over 90% of them are hormonally inactive; therefore they do not produce clinical symptoms associated with hormonal dysfunction.

Tumours occurring in the head and in the neck are predominantly paragangliomas of the common carotid artery division (over 60%) and the bulb of the jugular vein. The latter are most frequently localized in the middle ear and in the jugular vein foramen on the cranial base (tympanic paraganglioma).

These tumours occur in middle-aged people (over 40 years old), and are slightly more common in women [1]. They may be multiple. Over 3% of multiple tumours are familial [1].

Their clinical presentation is mainly associated with a characteristic structure (highly vascular) and localization. They are pulsating tumours. In the middle ear they usually present as a pulsatile tinnitus (buzzing in the ear). Tumour extension in this region may lead to ear damage and hearing loss.

These tumours are benign in over 95% of cases, and they are characterized by slow growth [1].

Diagnosis is currently based on characteristic clinical symptoms and typical appearance in CT and MRI studies.

Material and methods

Twenty patients treated in the Centre for Oncology – Maria Curie Memorial Institute in Warsaw between 1997 and 2005 with head and neck non-adrenal paragangliomas (8 men and 12 women), with age ranging from 24 to 75 years, were included in the study.

In our group of patients there are 16 cases with single tumours and 4 cases with multiple lesions (Table 1). Multiple tumours may be familial; however there are no such cases in our group of patients.

The overall number of diagnosed tumours was 29. There were 16 carotid artery paragangliomas, 8 jugular vein paragangliomas and 4 tympanic paragangliomas, and 1 glomus vagale tumour (Table 2).

The tumours had various sizes – from 15 mm to over 5 cm in diameter (Table 3).

The aim of this study was to define the role of imaging techniques in detection of paragangliomas of the head and neck and follow-up of patients treated with radiation therapy.
Material and methods: 20 patients aged between 24 and 75 were diagnosed by CT and MR (both examinations were performed before and after contract administration). Patients had follow-up every 4-6 months for first 2 years and then usually once a year. The follow-up periods was between 1 and 7 years.
Results: we detected 29 paragangliomas, in that 13 jugolothympanic tumors and 16 carotid body tumors, 4 patients have multifocal lesions, 8 lesions were operated, in these cases the diagnosis was established by microscopic examination. Biopsy gave no conclusive results. The remaining 12 patients were diagnosed on the basis of clinical and radiological studies. 14 patients were treated by radiation therapy. In the follow-up of one case we detected progression.
Conclusions: Our study suggested that diagnosis of paragangliomas could be made through clinical examination and radiological imaging. Both methods are equal for monitoring therapy and follow-up.

Key words: head and neck non-adrenal paragangliomas, computed tomography, magnetic resonance.

Table 1. Number of patients – own material
Tabela 1. *Liczba pacjentów – materiał własny*

Number of patients	20
with single tumour	16
with multicentric tumours	4

Table 2. Number of diagnosed tumours
Tabela 2. *Liczba rozpoznanych nowotworów*

Number of diagnosed tumours	29
carotid artery paragangliomas	16
bulb of jugular vein paragangliomas	8
tympanic paragangliomas	4
glomus vagale tumour	1

Table 3. Size of diagnosed tumours
Tabela 3. *Rozmiar rozpoznanych nowotworów*

The large dimension	≤ 15 mm	16-30 mm	31-50 mm	> 50 mm
Number of diagnosed tumours	3	13	7	6

Table 4. Duration of follow-up
Tabela 4. *Okres obserwacji*

Observation period(in months)	≤ 12	12-24	25-36	37-60	> 60
Number of patients	2	2	2	5	9

In the majority of cases the observation period was long: more than 3 years in over 50% of cases (Table 4).

Eight patients were operated on and in these cases the diagnosis was established by microscopic examination. Biopsies performed in 6 patients were not conclusive. The remaining 12 patients were diagnosed on the basis of clinical presentation and imaging studies – CT was performed in 8 cases, MRI in 4 cases.

From the entire group of evaluated patients 14 were treated with radiation therapy, and 6 of them had undergone previous non-radical surgery. All treated patients received a radiation dose of 4500-5000 cGy.

Four patients are being followed up with no treatment applied.

From the group of 20 patients 19 are still in the follow-up. One patient died from brain astrocytoma after 60 months of observation.

Imaging studies, CT and/or MRI, were performed in every patient before commencing treatment and on follow-up visits every 4-6 months in the first two years, and every year subsequently.

CT scans included the cranial base region and the neck to the level of the larynx, and the examinations were performed with single scans of 5 mm slices after intravenous contrast administration.

MRI scans were obtained with T1-weighted and T2-weighted imaging, additionally after intravenous contrast administration with T1-weighted imaging in axial, sagittal and coronal planes. In order to confirm tumour proximity to the vessel, and thus to verify the diagnosis, an angio-MR was performed prior to treatment.

Lesion size was measured in the two largest dimensions. The degree of bone destruction of the cranial base was also evaluated on CT scans.

Results

In every followed-up patient we observed no or only minimal lesion growth.

Only one patient in three years of follow-up after non-radical surgery developed significant tumour growth (over 50%). This was an indication for radiation therapy.

The rest of the patients experienced stabilization of the disease after completion of the radiation therapy.

We did not observe any significant disease progression in 4 patients, who were followed up with no treatment implemented.

Discussion

Head and neck paragangliomas are rare, slow growing tumours. The onset of the disease is asymptomatic. Their structure and localization cause difficulties at the time of diagnosis. Microscopic diagnosis is difficult, since there is no possibility to obtain sufficient material via biopsy. None of the six biopsies we performed was diagnostic.

However, based on our long-term experience and the unanimous opinion of other authors it may be stated that in order to reach a correct diagnosis it is sufficient to closely investigate clinical signs and symptoms and evaluate radiological symptomatology in imaging studies [2-4].

Ultrasound is not specific even in the diagnosis of neck paragangliomas [4].

Therefore head and neck paragangliomas are diagnosed by the clinician in collaboration with a radiologist.

Thus a profound knowledge of radiological signs of these tumours – typical localization and very intense post-contrast enhancement on CT and MRI scans – is essential. On MRI, especially in small tumours, salt and pepper appearance is described. Angio-MR in cases of tumours of the carotid glomus or jugular vein allows demonstration of their proximity to vessels and confirmation of the diagnosis [2-4].

Management of these tumours also presents considerable difficulties. Surgery remains the sole radical treatment [1]. However, localization of these tumours and their proximity to vital nerves and vessels cause the treatment to be associated with a very high percentage of complications, even fatal ones [1, 6, 7].

Radiation therapy has been applied in the management of paragangliomas for over 20 years [6] It is not a radical treatment, and it does not eliminate the tumour, but it arrests tumour growth. From the published studies it may be concluded that the proportion of long-term survivals without disease progression is similar to radical surgery, and the proportion of complications of treatment is minimal [6-8].

For that reason most centres consider radiotherapy a treatment of choice in head and neck paragangliomas.

The use of an imaging method enabling as accurate as possible measurement of tumour dimensions is obligatory for evaluation of treatment efficacy and monitoring of further course of the disease. Our material, as well as the opinion of other authors, indicates that both techniques (CT and MRI) entirely fulfil the requirements. As these tumours are well delineated and intensely enhance after contrast administration, it is easy to accurately determine their margins and measure their size in the two largest dimensions.

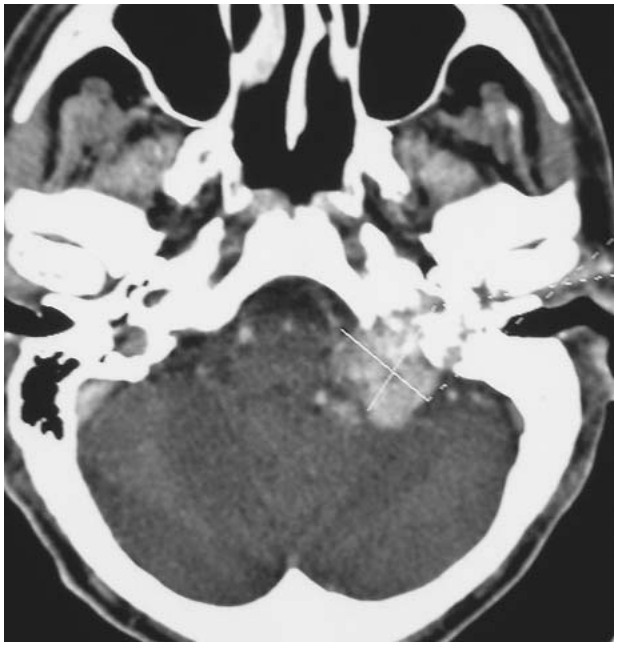

Fig. 1. Glomus jugulare tumour
Ryc. 1. *Przyzwojak opuszki żyły szyjnej*

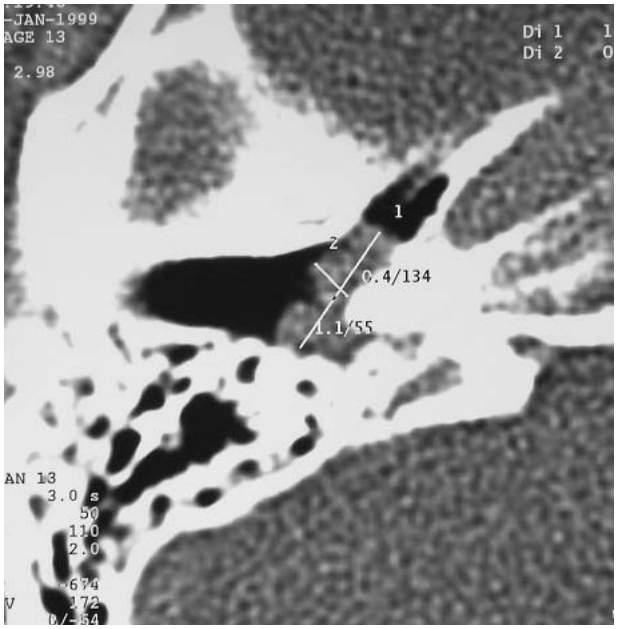

Fig. 2. Glomus tympanicum tumour
Ryc. 2. *Przyzwojak ucha środkowego*

Evaluation of tumours located on the cranial base (middle ear, bulb of jugular vein) present the biggest difficulty. Frequently the first sign of growth of these tumours is simply more extensive bone destruction. In such cases CT scans are more efficient in monitoring of the course of the disease [1, 2]. The purpose of the monitoring is to detect a marked acceleration of tumour growth, which is an indication for more aggressive treatment [6].

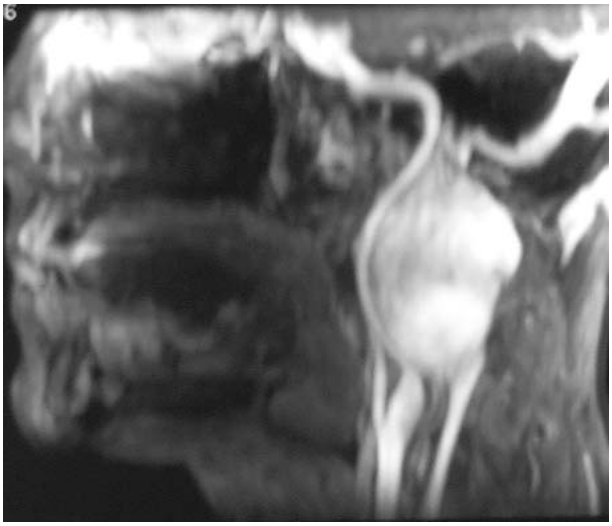

Fig. 3. Vagal paraganglioma
Ryc. 3. *Przyzwojak nerwu błędnego*

In our group of patients there was only one such case, and the progression occurred three years after non-radical operative treatment.

In the presented material there are 4 patients with paragangliomas who, for various reasons (medical contraindication to radiation, lack of consent to treatment), did not undergo any type of treatment and who are only followed up. These tumours are tympanic paragangliomas as well as carotid glomus paragangliomas; one of them is a multiple form. In all evaluated patients the follow-up period was over 5 years, and only in a single case was a very slow tumour enlargement observed.

The presented cases perfectly illustrate the thesis that head and neck paragangliomas are essentially benign tumours characterized by slow growth, and aggressive treatment is not necessary, as it does not influence survival time.

Aggressive treatment should only be applied in cases in which the course of the disease indicates a very dynamic process.

The use of radiation treatment in a dose of 4500-5000 cGy is effective in suppressing tumour growth [6-8].

Every patient with the diagnosis of paraganglioma should be followed for a long-term period. Patients should also have regular follow-up studies for tumour size evaluation.

Both imaging techniques (CT and MRI) are applicable in monitoring of lesions; however, in cases of cranial base paragangliomas CT has a certain advantage.

In conclusion:

1. A clinician and a radiologist collectively establish a diagnosis of non-adrenal head and neck paraganglioma on the basis of clinical picture along with CT and MRI imaging studies.
2. Angio-MR is the most efficient method to verify the diagnosis, demonstrating tumour proximity to vessels.
3. In cases of cranial base paragangliomas CT has an advantage, showing bone erosion.
4. Both methods are appropriate in the monitoring of the course of the disease.
5. Surgery remains radical treatment, but radiotherapy is a treatment of choice in head and neck paragangliomas.

References

1. Pellitteri PK, Rinaldo A, Myssiorek D, et al. Paragangliomas of the head and neck. Oral Oncology 2004; 40: 563-75.
2. Vogl T, Brüning R, Schedel H, Kang K, Grevers G, Hahn D, Lissner J. Paragangliomas of the jugular bulb and carotid body: MR imaging with short sequences and Gd-DTPA enhancement. AJR Am J Roentgenol 1989;153: 583-7.
3. Vogl TJ, Juergens M, Balzer JO, Mack MG, Bergman C, Grevers G, Lissner J, Felix R. Glomus tumors of the skull base: combined use of MR angiography and spin-echo imaging. Radiology 1994; 192: 103-10.
4. van den Berg R, Verbist BM, Mertens BJ, van der Mey AG, van Buchem MA. Head and neck paragangliomas: improved tumor detection using contrast-enhanced 3D time-of-flight MR angiography as compared with fat-suppressed MR imaging techniques. AJNR Am J Neuroradiol 2004; 25: 863-70.
5. Dhiman DS, Sharma YP, Sarin Nk. US and CT in carotid body tumor. Ind J Radiol Imag 2000; 10: 39-40.
6. Larner JM, Hahn SS, Spaulding CA, Constable WC. Glomus jugulare tumors. Long-term control by radiation therapy. Cancer 1992; 69: 1813-9.
7. Hinerman RW, Mendenhall WM, Amdur RJ, Stringer SP, Antonelli PJ, Cassisi NJ. Definitive radiotherapy in the management of chemodectomas arising in the temporal bone, carotid body, and glomus vagale. Head Neck 2001; 22: 363-70.
8. Springate SC, Haraf D, Weichselbaum RR. Temporal bone chemodectomas – comparing surgery and radiation therapy. Oncology (Williston Park) 1991; 5: 131-7.

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