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Value of CT and NMR Imaging in Diagnosing of Chordomas

Wartość obrazowania TK i NMR w rozpoznawaniu struniaków

Chordomas usually occur in the axial skeleton and they arise from the remnants of notochoridium. Their growth is slow and they often give metastases. Such cases are mainly observed among people above 40 years of age (3). Most frequently chordomas are situated in sacral bone (50%), in spheno-occipital region of the skull base (35%), and in the cervical, dorsal and lumbar spine (15%). Very rarely chordomas are found within sella, paranasal sinuses and nasopharynx (6), and in the forms of ectopic foci in the pons and spinal canal (8).

Such tumour situated within spine destroys vertebral bodies and arches and can bulge into subdural space causing impression of the dural sac (2, 13). Despite osteolytic destruction, chordomas cause visible reactive sclerotization and characteristic granular calcifications or ossifications. Intracranial chordomas destroy bony base of skull, specially clivus, piramides and sphenoid sinuses.

We present our own experience in this topic on the basis of our material containing relatively big number of such cases. There was no sufficient discussion concerning CT and NMR imaging of chordomas in the Polish literature up to now.

MATERIAL AND METHODS

The object of investigations were 11 patients, 5 females and 6 males. Their age ranged between 15 and 65 years of age. The radiologic diagnosis was verified at operation and confirmed by microscopic examination. Ten patients were treated in Neurosurgical Department of the National Hospital for Nervous Diseases, Queen Square, in London in the period of 1966—1987 and one case was examined in Lublin.

Radiologic diagnosis of chordoma was established in 6 cases by means of CT procedure and in the other 5 cases by using of NMR imaging. CT examinations were done using CT 9800 scanner made by General Electric and third generation apparatus Somatom DRH 3 (Siemens). CT scans were performed in axial plane before and after intravenous contrast administration, using narrow and wide window.

NMR studies were done with the equipment made by Picker International (superconductive magnet, 0.26 T). The patients were examined using spin echo sequence (SE — 2000/80) in axial and in sagittal plane, and in inversion recovery sequence (IR 2100/500, 500/40), in sagittal and coronal plane.

RESULTS

Most of all chordomas were placed in the base of the skull in spheno-occipital region (4 cases). Next 3 cases were situated in cervical and dorsal spine. The other tumours (4 cases) were found in the region of sella, paranasal sinuses and sacral bone. Most of patients were in the third and the fourth decade of life (5 cases). The ratio of females to males was 1 : 1. CT scans of chordomas revealed increased density of tumour mass in 6 cases (Fig. 1). The destruction of the bony structures adjacent to the tumour mass was observed in all the examined cases. Minimal contrast enhancement of chordomas was noted in 3 cases. The granular calcifications were seen in 3 cases.

The second group of patients (5 cases) was examined by means of NMR method. In NMR imagining using spin echo sequence, chordomas had a high signal in 3 cases (Fig. 2a, b) and mixed signal with only foci of high signal in 2 cases. In the inversion recovery sequence chordomas had isodense signal (Fig. 3a, b) in 3 cases, and one low and one mixed signal in the other two cases.

DISCUSSION

At present, using CT and NMR imaging one can reveal the pathologic changes with precision unobtainable till now with conventional methods. Anyhow, in our realities, it would be a mistake to resign standard tested X-ray procedures. They will be in use in many hospitals which have not modern CT or NMR equipment. They will be of importance in initial selective radiological diagnosis. The frequency of occurrence of chordomas is rather small. There were found only 600 cases of chordomas in scientific world literature up to 1979 (5).

Chordomas give distant metastases in 10% of all cases. Chordomas are very difficult for treatment and they have tendency for recurrence (9). In CT imaging chordomas show sharply well outlined hyperdense masses (+70 y.H.U.) not seldom with the presence of reticular, nodular or spotted calcifications, sometimes in the periphery of lesion. The frequency of their occurrence according to different authors ranges 34—86% of all cases, which correlates with our observations (60%).

On precontrast scans, the soft part of tumour mass can be of low or intermediate density. The degree of density of chordomas depends on the ratio of cellular and mucous elements. The predominance of cellular elements causes the increase of density of chordomas. After intravenous contrast administration

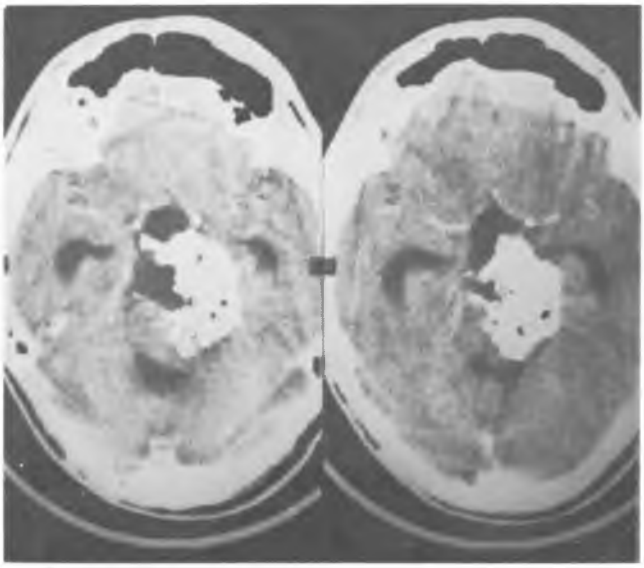
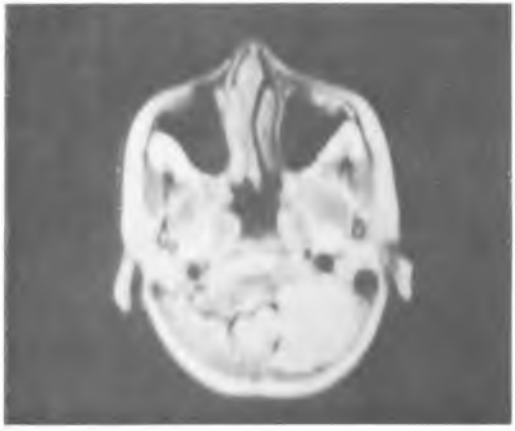


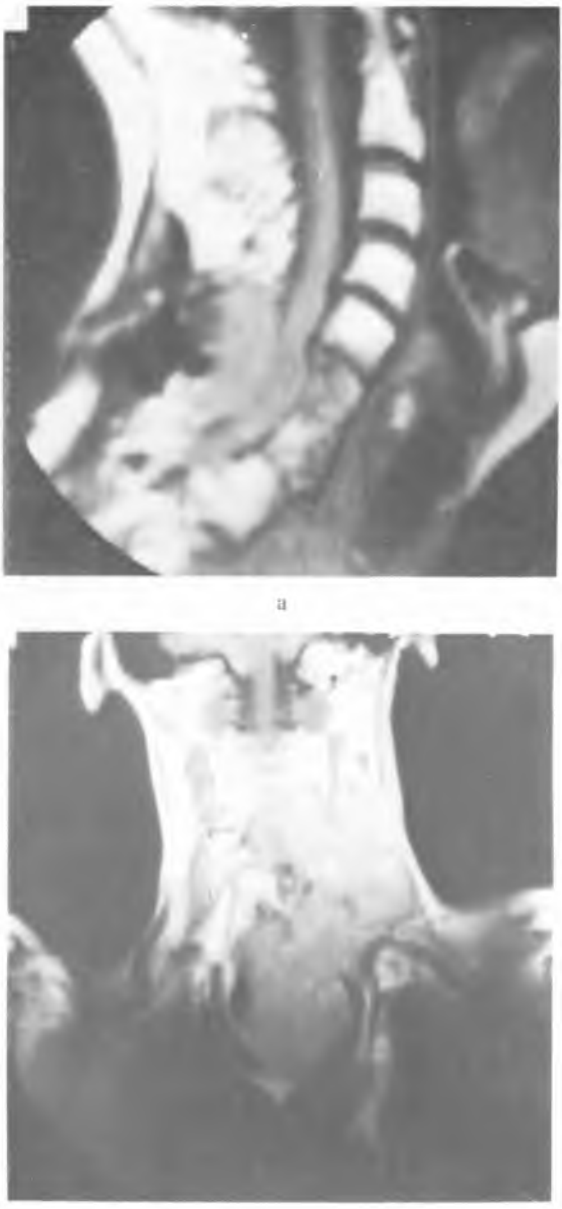
Fig. 1. A man B. D. 45 years. Postcontrast axial CT scan. The tumour mass with irregular outlines and increased density in the region of left pontocerebellar angle is seen. It penetrates into the left middle cranial fossa and infiltrates the region of left thalamus and posterior part of the third ventricle. There is a widening of ventricular system and diplacement of fourth ventricle backwards and to the right side. There is no evident contrast enhancement of tumour mass. Micronodular calcifications within tumour mass are seen. Chordoma was found at operation

a



b

Fig. 2. A woman M. H. 15 years. NMR examination in IR sequence in sagittal plane (a) and in SE sequence in axial plane (b). The tumour mass with lobular outlines in posterior cranial fossa is seen. It presses the left cerebellar hemisphere and penetrates through the big occipital foramen into spinal canal. The tumour mass surrounds the spinal cord till the height of C3. The tumour has a high signal in SE sequence and a low signal in IR sequence. Chordoma was confirmed at operation



a

b

Fig. 3. A woman M. A. 41 years. NMR examination in IR sequence in frontal and sagittal plane. There is an evident bony destruction of C6, C7 and D1 vertebral bodies with visible adherent big tumour mass. The tumour has an isodense signal with tiny foci of high signal (a). It penetrates into soft tissues of neck on the left side and into anterior mediastinum (b). Chordoma was found at operation

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chordomas show the diffuse or spotted contrast enhancement. Big low density chordomas did not show contrast enhancement. Intermediate density chordomas show some contrast enhancement but in lesser degree than meningiomas. Markedly calcified chordomas may not show contrast enhancement, or show only its marginal type.

Chordomas in the region of clivus cause compression and posterior displacement of the brain stem. Grądzki (4) observed this type of chordoma which on CT scans showed destruction of bony structure of sella, penetrated into the sphenoid sinus, with compression of optic chiasma and deformity of the basal cisterns. The occurrence of bony destruction in chordomas is differently mentioned. Chordomas destroy clivus in 53—62% of cases, bony structure of sella in 34—69%, pyramids in 30—62%, orbital fissure in 20—25% and foramen ovale in 23% (5, 7, 12). Sclerotic reactions are seen in 5% of chordomas.

CT imaging of spinal chordomas shows the signs of destruction of vertebral bodies with the presence of tumour mass localized within spinal canal and paravertebrally. Calcifications are seen in 50% of these cases. These tumours have ability of contrast enhancement.

Chordomas of the clivus and the region of sella should be differentiated with meningiomas, chondrosarcomas and benign intra and parasellar tumours. Clivus meningioma has a very similar CT image as chordoma, with a big contrast enhancement and the presence of calcifications. A chordoma usually gives the destruction of clivus, while meningioma causes rather modulation and deformation of the clivus outline. The extent of bony destruction including sella region, clivus, and pyramids, and the presence of calcifications can allow the differential diagnosis between chordoma and pituitary adenoma. The differential diagnosis of chordomas and other malignant tumours of the sphenoid bone may be very difficult because all these tumours can contain calcifications.

Sze et al. (11) showed in his NMR T1 weighted images, the presence of isodence areas in 75% of cases of chordomas, and hypodense areas in 25% of patients. All chordomas had a high signal of different intensity in T2 weighted images. There were low signal septa within high signal tumour masses in 70% of cases. NMR examination in sagittal and coronal planes is very useful for the full estimate of the tumour extent. The marrow cavity of sphenoid clivus in normal conditions is seen as the area of high signal in T1 weighted images, because of big content of fat. The replacement of fat in the marrow cavity by tumour mass gives a low signal, which is characteristic of chordoma (1). Well outlined border between tumour mass and unchanged part of clivus is seen. There is the "drop-out" of signal in case of big calcifications or the presence of sequester, independently of used sequence (1).

Chordomas can infiltrate the cranial nerves, and narrow or occlude the lumen of the carotid artery. These changes can be easily seen in NMR imaging. NMR method allows the exact visualization of epidural space involvement by this kind

of tumour in its spinal localization (10). There are 3 main rules in comparison of diagnostic effectiveness CT and NMR techniques: 1) ability of tumour detection; 2) possibilities of visualization of tumour extent; 3) specific diagnosis. On the basis of our own experience and literature data we can say that both CT and NMR are equal procedures in diagnosis of chordomas (10, 11).

In case of uncertain results of one of these methods, the second procedure allows the settlement of final proper diagnosis. NMR technique can be unreliable in detection of a very small chordoma which is placed completely within clivus. To consider the second criterion, NMR is more efficacious method than CT in definition of the tumour extent (11). NMR technic allows better visualization of the tumour's borders and its relation to blood vessels due to big difference of signal intensity of these structures. Tumour mass has a high signal. There is a lack of signal in projection of vessels due to fast flow of blood within it. Chordomas cause marked displacement of cerebral vesels in 50% of cases.

Both techniques are equally good in visualization of chordomas extent in the skull base, anteriorly towards nasopharynx. NMR is better in visualization of chordomas expansion backwards in the skull base. NMR scans done in sagittal plane are very useful. They allow the precise definition of the epidural and subdural extent of chordoma and possibility of infiltration of the brain stem, structures which are of great importance in planning of operation. NMR technique may show the continuity between foci of chordoma localized in the base of skull and in cervical spinal canal. It helps in better visualization of small metastases of this tumour in lumbar spine. Both metods (CT and NMR) are almost equal in specific diagnosis of chordoma.

Chordomas have characteristic acinar partly cystic structure of tumour mass with the presence of calcifications and destructive changes within. Compared with CT, NMR technique fails in complete visualization of bony destructive changes and small calcifications. Such changes in NMR imaging are seen as areas of low signal. However, CT fails in visualization of early form of intradural chordoma. NMR technique is better than CT in visualization of cystic character of tumour.

There is a good NMR presentation of acinar structure of tumour with septa and the outline of pseudocapsule which give low signal NMR technique allows to conclude with great probability about quantitative and qualitative internal structure of chordomas. It allows to distinguish 2 types of NMR images corresponding with different histologic types of chordomas: real chordomas and chondroid chordomas (1). It is of great importance in prognosis, because patients with chondroid chordoma have a chance of four times longer period of survival (11). The prognosis is bad, because the complete resection of tumour is very rarely possible, due to tumour's infiltration of cerebral structures which are very important for life (9). The optimal procedure is partial resection of tumour and following radiotheraphy and chemiotheraphy.

Conclusions

1. The majority of chordomas in our material were localized in spheno-occipital region (40%) and in cervico-dorsal spine (30%).

2. Half of chordomas concerned patients in the third and fourth decade of life.

3. The ratio of women to men was 1 to 1.

4. CT and NMR are equal to localization of chordoma.

5. The tumour's extent and its sharp borders from surrounding structures were better visualized using NMR technique.

6. Both methods of imaging are almost equal in specific diagnosis of tumour, however CT is better in visualization of bony destruction and the presence of calcifications within tumour mass.

7. Our CT images of chordomas correlated with results obtained by other authors.

8. NMR images of chordomas in IR sequence were similar in studies done by other authors, however, in examinations done in SE sequence beside typical changes with high signal there was a big percentage of cases with mixed signal.

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STRESZCZENIE

Na podstawie materiału, obejmującego 11 przypadków struniaków operacyjnie potwierdzonych i histologicznie zweryfikowanych oraz piśmiennictwa, oceniono retrospektywnie wartość diagnostyczną badania rentgenowskiego ze szczególnym uwzględnieniem obrazowania TK i NMR. Zwrócono uwagę na znaczenie diagnostyczne TK i NMR w korelacji z wykrywalnością i zasięgiem guza oraz z charakterem zmian chorobowych.

Z przeprowadzonej analizy wynika, że obrazowanie przy użyciu technik TK i NMR jest równorzędne, jeśli idzie o wykrywanie i umiejscawianie się guza, natomiast NMR okazała się precyzyjniejsza w wykazywaniu zasięgu struniaka i jego kierunku wzrostu. Rozpatrując możliwości rozpoznawania rodzajowego obu technik wykazano, że zarówno TK, jak i NMR są prawie równorzędne w rodzajowym rozpoznawaniu struniaka, jednak metoda TK wyraźniej uwidacznia destrukcję kostną i obecność zwapnień w masie guza.

W obrazowaniu NMR w sekwencji odrostu namagnesowania (IR) obraz struniaków korespondował z wynikami uzyskiwanymi przez innych badaczy, natomiast w sekwencji echa spinowego (SE) obok zmian typowych, o wysokim sygnale, stwierdzono znaczny odsetek przypadków charakteryzujących się obecnością sygnału mieszanego.