Cerebellopontine angle tumours: radiologic-pathologic correlation and diagnostic difficulties

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Abstract

A group of 119 cases of cerebellopontine angle (CPA) tumours was studied looking at the pathological composition, relative incidence of tumour types, their radiological features and the pathological-radiological correlations. Tumours with preoperative radiological diagnosis and verified pathologically were analyzed. Histopathologically the material consisted of 77 schwannomas and 42 non-acoustic tumours. Radiological retrospective evaluation of CT and/or MRI documentation was performed in 84 patients. The tumours were classified according to Koos’s staging scale. Diagnostic discrepancies (histopathological vs radiological) according to the clinical stage of CPA tumours were analyzed. In our series non-acoustic tumours made up 37% of CPA lesions. Sharp tumour-pyramis angle and intracanalicular fraction are not exclusive features of schwannomas. Tumours in stage IV are the most heterogeneous and diagnostically difficult group.

Key words: acoustic tumours, Koos’s staging scale

Introduction

The cerebellopontine angle (CPA) is an anatomical and radiological term for the space bound anterolaterally by the posterior aspect of the petrous temporal bone and posteromedially by the cerebellum andpons. It contains important vascular structures and cranial nerves. Internal auditory meatus (IAM) is included in CPA structures [4,18,20]. Lesions of CPA, creating about 10% of all intracranial tumours, can be of native origin or they can extend to it from adjacent structures. The tumours can derive from many anatomical structures, including primary origin from IAM, ponto-cerebellar cystern, lateral recessus of the fourth ventricle, temporal bone, brain stem or cerebellar nervous tissue [3,20,23]. The most common CPA tumour is schwannoma, which constitutes about 70–80% of all CPA lesions. The next in order are: meningiomas (5–12%)...
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and epidermoid cysts (2–6%). The other tumours and non-tumourous conditions are rare [5,17].

Surgical approach to CPA tumours is determined by tumour location and preoperative prediction of neurovascular bundle and brain stem displacement based on radiological diagnosis [19–21]. There are several basic neurosurgical approaches to CPA: retromastoid, translabyrinthine, petrosal, suboccipital, transcoclear and middle fossa [2,12,30].

CT and MRI are widely used radiological methods for CPA imaging. The main radiological diagnostic goal is the description of the relation of the tumour to IAM, the brain stem and cerebellar hemispheres. The second line basic information is if the lesion is extra- or intracerebral [4,6,9,31].

The aim of the present study was the retrospective analysis of material diagnosed in the Department of Pathomorphology of the Medical University in Gdańsk in the last 12 years according to the histological diagnosis and diagnostic radiological difficulties, as well as pathological-radiological correlations.

Material and methods

119 cases of CPA tumours were diagnosed in the Department of Pathomorphology of the University of Gdańsk in the period June 1993 – April 2005. The patients were operated on in two neurosurgical departments in Gdańsk. The radiological diagnosis was performed in most cases at the Department of Radiology, Medical University of Gdańsk.

The patients were 83 women and 36 men aged 14–77 years. Most of the tumours were sporadic cases and only in three patients did CPA tumour occur in train of genetic diseases as NF I, NF 2 and meningiomatosis. Clinical symptoms included audiovestibular and facial nerve dysfunction, tinnitus, hearing deficits, deafness, hypoacusis, vertigo, unsteadiness, disequilibrium, walking ataxia, intention tremor and nausea. The clinical history ranged from 3 months to 22 years. The average duration of symptoms was about three years.

The authors studied radiological documentation CT and/or MRI available for retrospective analysis in 84 cases, including 75 patients who underwent MRI and/or spiral CT (70 cases). MRI examination consisted of T1-, PD- and T2- weighted images with SE, TSE, FLAIR, FFE sequences in 3 planes, along with study after i.v. administration of Gd-DTPA. CT included non-contrast and enhanced scans. The CT and MR images were independently reviewed by two radiologists (ES, MDW). Each case was reviewed for signal intensity, type of enhancement (homogeneous, heterogeneous, peripheral) and presence of IAM widening and oedema of adjacent neural structures. The tumours were classified according to WT Koos scale for staging acoustic neurinomas (Koos). Stage I included tumours within IAM; stage II – CPA tumours with or without IAM fraction; stage III – CPA mass attached to the brain stem respecting arachnoid; and stage IV – CPA tumours with brain stem compression and arachnoid invasion.

Diagnostic pathological-radiological correlations and discrepancies were further analyzed, searching for pitfalls in imaging of CPA tumours.

Results

Maximal diameter of the tumours ranged from 0.5 cm to 8 cm (median 3 cm).

In the total material of 119 cases of CPA tumours preoperative radiological diagnosis (CT, MRI) comprised: 83 schwannomas and 36 non-acoustic tumours including: 27 meningiomas, 5 epidermal cysts, two gliomas, one arachnoidal cyst and choroid plexus tumour (Fig. 1).

The same material verified histopathologically consisted of: 77 schwannomas and 42 non-acoustic tumours including: 24 meningiomas, 8 epidermal cysts, 2 astrocytomas, 2 metastases and single cases of choroid plexus papilloma, PNET, chondrosarcoma, arachnoidal cyst, cavernoma and haemangioblastoma (Fig. 2).

Retrospective radiological analysis

Among 84 cases analyzed radiologically there were nine tumours in stage I, 33 in stage II, 30 in stage III and 12 in stage IV (Table I).

Radiologically all nine stage I tumours were diagnosed preoperatively as schwannomas. Pathologically the group consisted of five schwannomas, two meningiomas, one cavernoma and one metastatic adenocarcinoma. Mean diameter of intrameatal tumours was 8 mm.

In four cases there were diagnostic discrepancies (4/9; 44%). Retrospective analysis showed angular or infundibular widening of IAM in two schwannomas, one cavernoma and one meningioma (mean width – 6.9 mm, compared to normal 5.2 mm). All tumours were homogeneously contrast enhancing. The tumour contours were regular, without oedema or bone
Histopathological diagnosis of cerebellopontine tumours

Characteristics of tumours

<table>
<thead>
<tr>
<th>Tumour stage</th>
<th>Number of cases</th>
<th>Number of schwannoma (pathological diagnosis)</th>
<th>Number of rare tumours (pathological diagnosis)</th>
<th>Diagnostic discrepancies number of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>9</td>
<td>5</td>
<td>2*</td>
<td>4 (44%)</td>
</tr>
<tr>
<td>II</td>
<td>33</td>
<td>26</td>
<td>–</td>
<td>7 (21%)</td>
</tr>
<tr>
<td>III</td>
<td>30</td>
<td>16</td>
<td>2**</td>
<td>4 (13%)</td>
</tr>
<tr>
<td>IV</td>
<td>12</td>
<td>5</td>
<td>3***</td>
<td>6 (50%)</td>
</tr>
</tbody>
</table>

* cavernoma, metastatic adenocarcinoma; ** PNET, chorioid plexus papilloma; *** 2 astrocytomas, haemangioblastoma.
attachment to the tentorium or petrous dura mater and created an open angle with the pyramis. Most of the schwannomas were characterized by acute angle with pyramis of the temporal bone (21/26). In all cases the continuous presence of the CSF rim between the pons and/or cerebellar hemispheres was observed (Fig. 4A, 4B).

30 cases in stage III were radiologically diagnosed as: 18 schwannomas, 9 meningiomas, two epidermal cysts and one plexus tumour. Mean diameter of tumours was 3.1 cm. Pathologically, 16 schwannomas and 14 non-acoustic tumours, including eight meningiomas, four epidermal cysts, one case of PNET and one plexus papilloma were recognized.

Diagnostic discrepancies concerned five cases (5/30; 17%). Intrameatal fraction with IAM widening was detected in four schwannomas and one meningioma (mean 6.1 versus 5.2 mm). PNET which was homogeneously contrast enhancing and created an open angle with pyramis was recognized as meningioma. Two epidermal cysts and meningioma, which were heterogeneously contrast enhancing and created an acute angle with the pyramis, were classified in CT and MRI as schwannomas. Eight of 15 schwannomas were homogeneously contrast enhancing and all schwannomas created an acute angle with pyramis. In all cases in stage III the segmental presence of the CSF rim between the pons and/or cerebellar hemispheres was observed. In 14 cases the brain stem was compressed by the tumour mass: in five cases brainstem and cerebellar hemisphere and in nine cases one cerebellar hemisphere. In 20 cases oedema of nervous tissue was observed (66%) (Fig. 5).

12 cases from the analyzed group were consistent with Koos stage IV. Mean diameter of tumours was 4.2 cm. Radiological diagnosis of these tumours included: six schwannomas, three meningiomas, two epidermal cysts and one glioma. Pathological verification disclosed: five schwannomas and seven non-acoustic tumours including two astrocytomas, two epidermal cysts, two meningiomas and one haemangioblastoma. Diagnostic discrepancies concerned six cases (6/12; 50%).

All tumours lacked a continuous rim of CSF on their surface. In seven cases the rim was only segmental or punctate. One glioma was diagnosed as meningioma invading arachnoid; two schwannomas creating an open angle with pyramis were recognized as meningiomas. One epidermal cyst...
with heterogeneous contrast enhancement and invading arachnoid was diagnosed as schwannoma. Haemangioblastoma was primarily diagnosed as a haemorrhagic epidermal cyst. Nine cases presented heterogeneous contrast enhancement with usual hyperintense signal in T2 images. One epidermal cyst was peripherally contrast enhancing. All cases presented with oedema of the brainstem and/or cerebellar hemisphere and in six tumours the fourth ventricle was compressed. Three of 12 tumours in stage IV were extracerebellar (Fig. 6).

Discussion

Cerebellopontine angle tumours, although uniform in location, are diverse pathologically and with regard to the site of tumour origin and displacement of the neurovascular structures. In general CPA tumours are divided into acoustic and non-acoustic tumours [3,20,21]. The main factor underscoring the importance of accurate preoperative diagnosis is the different surgical approach for vestibular schwannomas and the other tumours [13,15,19]. There is general agreement that completeness of tumour resection and preservation of the facial nerve are the major neurosurgical goals [2,19,12]. An appreciation of the vascular and cranial nerve microanatomy and the relationships between neurovascular structures and the tumour are essential for achieving optimal surgical results [13,20,30].

CT is a widely used method for lesions of the CPA, but MRI is also the imaging modality of choice for CPA and internal auditory canal masses [9,22,23]. CT is fairly accurate in diagnosis of schwannomas bigger than 10–15 mm and schwannomas with intrameatal fraction because it can demonstrate enlargement of the acoustic canal as well as the mass itself. However on CT axial images small intracanalicular schwannomas can be missed, as four cases in stage I in our group [6,13]. CT and MRI should be used complementarily in CPA diagnostics and have a great impact on therapeutic planning [9,27,29].

There are common criteria for radiological diagnosis of the most frequent CPA masses, and several clinico-radiological staging classifications have been created for neurosurgical purposes [18,20].

We present the pathological and radiological analysis of the biggest series of CPA tumours reported in Poland [2,24,31]. We analyzed this group according to its pathological characteristics and radiological picture. In our series the CPA tumours were more frequent in women. The most common CPA tumour was typically vestibular schwannoma. It represented 67% of all tumours, so this frequency was lower than in the other described groups (72–80%) [3,5,17,23]. We used schwannoma Koos’s scale for staging of all CPA tumours [12]. Schwannomas in the radiologically reevaluated group were the most numerous group in stage II-26/33 (80%) but in stage IV made up only 41% of cases.

A characteristic radiological feature of schwannomas is the presence of intrameatal fraction (“ice cream on cone” sign) and IAM widening, as well as contrast enhancement and the sharp angle between tumour and temporal bone pyramid [7,19,23]. In our group there were however several cases without intrameatal fraction, which caused diagnostic difficulties. Schwannoma can originate from any place between the gilal-Schwann cell junction and IAM fund. It arises from the Schwannian cells of the superior division of the vestibular nerve [18,25]. Yasargil’s theory explains that schwannomas
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A large meningioma in stage IV involving pons and cerebellum is visible on T2-weighted MR axial image. This lesion does not extend to 7th and 8th cranial nerves.

originate epiarachnoidally in the IAM and push the arachnoid membrane of CP cistern medially, causing folding of the arachnoid between the tumour and the brain. Ohata et al. postulate however the concept of subarachnoidal origin of vestibular schwannoma in IAM [18].

Non-acoustic CPA tumours are less common, but they form an interesting and diverse group of pathologies. They make up from 20 to 25 % of masses of this region [5,15,17,20]. In our group the non-acoustic tumours constituted 33% in the whole group and 40% in the series reevaluated radiologically. In our study the proportion of non-acoustic tumours was higher than in other series [5,17,20].

CPA meningiomas (3–13%) are difficult to distinguish from vestibular schwannomas preoperatively. The information about exact type of the neoplasm also has prognostic significance since the recurrence rate is higher for meningiomas [15,17,26]. Clinically schwannoma is characterized by progressive unilateral hearing loss, usually with tinnitus. Tumour progression results in unsteadiness, but facial nerve symptoms are usually late [16,28]. In meningioma early facial nerve involvement is more common, and
Metastatic tumours of the CPA are also very rare and mistaken for schwannoma when it occurs in IAC [1,17]. Meningiomas of CPA are diverse with regard to the site of dural origin. The most common places of origin of CPA meningiomas are: petrous ridge, tentorium and clivus [17,29]. These tumours can occasionally develop from meningeal cell foci within IAM [18,25]. This can explain the presence of intrameatal fraction of some meningiomas in our study, as well as suggesting that they can grow into IAM.

Epidermal cysts are the second most frequent non-acoustic CPA tumour type (4–7%). They have a developmental origin [5,17]. They grow slowly, often fill the meningeal cisterns, causing adhesions with arachnoid, and they rarely dislocate the vascular-neural structures [15,23]. Epidermal cysts are characterized as cystic non-contrast-enhancing lesions located outside IAM, with signal closed to CSF in 75% of cases [7,9,31]. In our material all these tumours were recognized in stage III and IV. Six epidermal cysts were typically non-enhancing, but heterogeneous contrast enhancement was present in two cases, causing diagnostic discrepancies.

In our series we found several very rare cases of non-acoustic tumours comprising gliomas, PNET, intrameatal haemangioma, metastasis, arachnoid cyst, plexus papilloma, haemangioblastoma and chondrosarcoma. CPA gliomas, as in our cases, are usually exophytic gliomas of the brain stem or cerebellum. PNETs in the posterior fossa are almost always medulloblastomas; however, several extracerebellar cases, including CPA location, have been reported [10]. Cavernous haemangioma is an extremely rare tumour in this region and can be easily mistaken for schwannoma when it occurs in IAC [1,17]. Metastatic tumours of the CPA are also very rare and their differential diagnosis without prior knowledge of the primary tumour is practically impossible [5,8]. Arachnoid cysts are collections of CSF contained within the arachnoidal membrane and subarachnoid space and less than 30 adult cases in CPA have been reported [11]. Choroid plexus tumours and haemangioblastoma develop in the fourth ventricle and superficially in the cerebellum or the brain stem so they involve CPA as their primary site very rarely [14].

Our case of chondrosarcoma developed from the temporal bone structures and invaded CPA secondarily.

The radiological-pathological discrepancies reaching 41%, 21%, 17%, 50% in consecutive CPA tumour stages derived from the high heterogeneity of the analyzed series of tumours.

One of the main causes of radiological misinterpretation was widened IAM in non-acoustic tumours. However, we encountered IAM widening in only 50% of schwannomas in stage I and 62% in stage II, making this proportion much lower than in previous reports [17,22]. The most monotonous were schwannomas in stage II and the basic difficulties in correct radiological diagnosis in this group were intrameatal fraction of meningiomas. We did not notice like Schuibiger et al. [22] differences in contrast enhancement between schwannomas and meningiomas, since Moffat [17] proved that schwannomas are more contrast enhancing than meningiomas. Moreover, big schwannomas, less frequently meningiomas in stage III and IV, presented mostly heterogeneous enhancement caused by cystic degeneration, necrosis and haemorrhage [7,31]. The most variable group pathologically was that of tumours in stage IV – big masses invading arachnoid and compressing the brainstem. In this stage the radiological-pathological diagnostic concordance was rather low and it was also a problem with characterization of intraaxial or extraaxial tumour localization.

Analysis of the tumour-pyramis angle showed that 92% of schwannomas and only two meningiomas created an acute angle. We observed wide dura attachment with meningeal/meningeal enhancement in some non-meningeal tumours, corresponding probably to inflammatory reaction of dura. In our material the oedema of posterior fossa structures, related to large tumour size, was found more often than in the other studies [4,19,27]. We observed it in 60% of cases in stage III and in all tumours in stage IV.

In conclusion in our series of CPA tumours, the non-acoustic tumours were more frequent than in previous reports. Gadolinium-enhanced MRI is more sensitive than CT in detection of tumours in stage I. The presence of intrameatal fraction is not exclusive only for schwannomas. The correct radiological differential diagnosis of tumours in stage IV is very difficult and the final pathological verification gives in half of the cases unexpected results.
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References


