Concerning the Late Diagnosis of Certain Cerebellopontine Angle Tumors

Abstract—The sensitivity of diagnostic methods for cerebellopontine angle lesions (auditory nerve electrophysiology, computed tomography scanning, magnetic resonance imaging) has increased in recent years. Paradoxically, in our experience this has not led to an increased percentage of tumors detected at an early stage. In 1975 20% of tumors were confined to the internal auditory canal, whereas only 6.6% were intercanalicular in 1988. A detailed analysis of 120 recent cases included first symptoms, time between first symptoms and diagnosis, symptoms at diagnosis, and size of tumor. We conclude the low percentage of intracanalicular tumors in the recent series results from: (1) rapid growth in some cases; (2) slow enlargement of the tumor over a long time without compression of adjacent cranial nerves leading to a paucity of symptoms; (3) misleading clinical pictures, including sudden hearing loss and hydrops; and (4) tumor symptoms masked by unrelated pathologic processes (otosclerosis, presbycusis, trauma). (*Skull Base Surgery*, 3(2):74–79, 1993)

Over the past 15 years imaging facilities have developed increased sensitivity (computed tomography [CT] scans, magnetic resonance imaging [MRI]), especially in the diagnosis of tumors of the internal acoustic meatus (IAM) and the cerebellopontine angle (CPA). Before 1975, the only imaging examinations possible were fractional pneumographs for large tumors and Lipiodol meatocisternographies (Pantopaque and similar products) for small tumors. At the end of the 1970s, CT scanning appeared and in the 1980s MRI became operational.

One might have thought that these new techniques would increase the percentage of early detection of tumors of the VIIIth nerve. This is not the case; in fact, our results show that the opposite is true. The otoneurologic teams that one of us helped set up in 1966 in Bordeaux have at present operated on more than 700 tumors of this region.¹ If we examine the statistics for 1975, 1981, and 1988 (Table 1),² we see that the percentage of small tumors detected has decreased, although the means of early diagnosis have improved concomitantly.

It is this apparent paradox that we would like to analyze in order to understand why most of the statistics currently published worldwide^{3–6} show, as ours do, a

relatively low percentage, approximately 6 to 11%, of small intracanalicular tumors.

DATA AND METHODS

Our observations are based on 120 cases of retrolabyrinthine lesions treated in the departments of Portmann, Guérin, and Bébér. The data from these cases have recently been analyzed and documented by our associate Duriez-Auria⁶ and has been the topic of research already published.^{7–9}

In arriving at the correct diagnosis in cases of CPA tumors, one tends to follow a distinct clinical algorithm, and it is the various stages along this path that we have studied.

First the patient complains of a particular symptom and is seen in the clinic. Suspicion is aroused to a greater or lesser extent following standard neurologic, ear, nose, and throat, and cochleovestibular examinations in association with the clinical history. At this stage, the symptoms and signs may be sufficient to justify omitting the electrophysiologic stage and proceed directly to radiologic imag-

Table 1. Neurinoma of the VIIIth Nerve

Stage	1975	1980	1988
I	20%	15%	6.6%
II	25%	35%	23.4%
III	35%	35%	36.6%
IV	20%	15%	33.4%
No. patients	60	200	120

ing. However, in the majority of patients confirmation of a retrolabyrinthine lesion is sought by an electrophysiologic study using brainstem auditory evoked response (BAER) and possibly electrocochleography (ECoChG). The diagnosis is finally provided by imaging with MRI and CT scan.

While analyzing this clinical algorithm, we have paid particular attention to the presenting symptoms, the clinical picture at the time of the first examination, and the time until the diagnosis was finally made. The audiometric tests, stapedia reflex tests, electrophysiologic examination of the VII nerve, and radiologic imaging were also reviewed.

Our classification of acoustic neurinomas includes four stages: stage I: an intracanalicular neurinoma with or without dilation of the IAM; stage II: a neurinoma overflowing from the porus and penetrating into the CPA without any contact with the cranial nerve V and the brainstem (smaller or equal to 1.5 cm in diameter); stage III: neurinoma invading the CPA completely, in contact with the brainstem but without displacing it; stage IV: neurinoma invading the CPA completely, displacing and compressing the brainstem and the other structures of the region to varying degrees).

Finally, the data were analyzed, notably the statistical relationship between the different symptoms and the extension of the tumor.

RESULTS

Presenting Symptom

The initial symptom can be auditory, vestibular, neurologic, or, indeed, none of these. The majority (88%) of cases presented with an auditory symptom, including unilateral hypoacusis (53%), isolated tinnitus (17.5%), sudden deafness (10%), hypoacusis plus tinnitus (4%), and bilateral hypoacusis (3%). Vestibular symptoms accounted for 8.5% of cases, of which most (7.5%) complained of vertigo, with only 1% presenting with loss of balance. The neurologic presenting symptoms were headaches and facial neuralgias and these constituted 3% of cases.

The time lapse between the first symptom and the diagnosis is highly variable; less than 1 year in 38%, 1 to 5 years in 48%, and over 5 years in 16% of cases. The

shortest time lapse was 3 weeks in a patient presenting with sudden deafness and followed by a severe loss of balance and tinnitus. The longest time lapse was 18 years in a case of progressive unilateral deafness not accompanied by any other symptom. At the time of diagnosis, this tumor was a stage IV.

Clinical Picture at Time of Diagnosis

Once again, this is extremely variable. The vast majority (98%) of patients had some degree of auditory impairment. This took the form of a neurosensory hearing loss in 91% of cases, accompanied by tinnitus in 60%, and isolated tinnitus in the remaining 7%. No auditory impairment was detected in 2% of cases. Thus, in 9% of patients there was no subjective hearing loss. Indeed audiometry showed that over a third of patients (34%) had hearing that was under 40 dB HL for conversational frequencies and was still socially useful. The other main symptoms concerned disorders of equilibrium and this problem effected 67% of patients. Along with the disorders already outlined, a variety of neurologic manifestations were often present. Headaches (22% of patients), corneal or facial dysesthesia (13%), facial neuralgias (26%), and damage to the facial nerve (37%) were all noted along with some instances of paresis, dysgeusia, and myoclonus.

In this series of 120, one patient presented with intracranial hypertension and required immediate hospitalization.

In summary, the different clinical pictures can be classified into the following groups: isolated cochlear disorders (26%), cochleovestibular syndrome (41%), and cochleovestibular syndrome with various neurologic signs (24%).

Clinical Picture in Relation to the Stage of the Tumor

It is interesting to study the relationship between the symptoms and signs, using the above classification, and the size of the tumor. This relationship was studied in 112 cases and the results presented in Table 2.

Based on this analysis, it can be seen that 10% of stage IV tumors became apparent only through cochlear symptoms. These symptoms are often not sufficiently specific to alert the patient or the general practitioner to the gravity of the condition.

DISCUSSION

Certain clinical pictures suggest the diagnosis of a CPA tumor and call for functional tests (BAER and ECoChG) and imaging (CT scan and MRI). However several situations may arise that can lengthen the time be

Table 2. Relationship Between Symptoms and Tumor Stage

Stage of Tumor	No.	Symptoms							
		Auditory Only		Cochleo-vestibular		Trigeminal		Central Nervous System	
		No.	%	No.	%	No.	%	No.	%
I	8	2	25	6	75	0		0	
II	26	16	62	6	23	3	12	0	
III	40	17	42.5	15	37.5	5	12.5	0	
IV	40	4	10	7	17.5	11	29	9	24

tween the onset of the first symptoms and the final diagnosis. These situations will be discussed.

The patient may not have sufficient symptoms to alert his doctor. This situation arose in 10% of our stage IV tumors and 42.5% of stage III tumors for a varying length of time. The diagnosis may be delayed by confusion with symptoms of other conditions, for instance pregnancy (cases 1 and 2) or cranial trauma (case 3) which leads to the appearance of the presenting symptom.

The stage III or IV tumors that present suddenly, without any previous symptomatology, generally start in the CPA and not in the IAM. For this reason, their development may have to reach a considerable size before there are clinical symptoms. Even at this stage the symptoms are minor. They may proceed to the stage of intracranial hypertension and severe decompensation without going through a prolonged preliminary otologic phase. In addition the severity of such symptoms may be due to an abnormally rapid development of the tumor.

In some cases the physician is consulted but is not suspicious of a retrocochlear lesion because the symptoms are suggestive of another diagnosis. In this study 10% of patients were diagnosed and treated for sudden deafness, and some cases the hearing recovered, confirming the physician's diagnosis. Four percent of tumors had symptoms that were absolutely identical to that seen in Ménière's disease. In a further 4% of cases the patient presented with a progressive facial palsy on the same side as the lesion and was only correctly diagnosed after several months or even years.

A further difficulty in diagnosis may be caused by the masking of symptoms due to a separate, totally independent, pathologic condition. In our series presbycusis and family deafness occurred in 5% of cases, otosclerosis in 1.8% (see case 4), noise trauma in 3%, cranial traumas in 0.9%, and various nonspecific vascular and nerve disorders in 6.8%.

SOME TYPICAL CASES

Case 1

A 35-year-old woman delivered her third child normally following a problem-free pregnancy. The following week she noted the appearance of a buzzing noise and a

feeling of fullness in her left ear. She did not consult a specialist until 2 months later. Examination of the ear drums was normal. The audiogram showed a slight perception deafness on the left side (tonal threshold at 15, 15, 20, and 40 dB, in the 0.5, 1, 2, and 4 kHz frequencies, respectively). Due to the recent delivery, the origin was thought to be vascular.

A vasodilator treatment was prescribed with another checkup 2 months later. This checkup showed the audiometric thresholds to have worsened slightly, having gone to 15, 20, 25, and 60 dB at 0.5, 1, 2, and 4 kHz. The electrophysiology test that was then carried out was in favor of a retrocochlear origin. The scanner confirmed the existence of a stage IV tumor. A month later, the day before the operation, the audiometric drop was very clearly accentuated (thresholds at 50, 50, 40, and 60 dB). Romberg's sign was unstable toward the right.

There was no clinical impairment of the Vth or VIIth nerve.

Case 2

A 26-year-old woman who, during the 8th month of her first pregnancy, presented with a decrease in hearing on the right side a few days after the appearance of homolateral facial hypoesthesia. During the days following an uneventful delivery, the patient developed a drunken manner of walking as well as frontal and temporal headaches.

Testing showed hypoesthesia of the three branches of the Vth cranial nerve, disappearance of the nasolabial groove at rest, and vestibular and cochlear impairment (Fig. 1). In addition to the headaches, there was paresthesia of the heels and the tendon reflexes were brisker on the right. Electrophysiologic tests confirmed retrocochlear impairment and the MRI showed a large neurinoma pressing against the brainstem with a beginning of ventricular dilation (Fig. 2).

The neurinoma was only slightly extended into the IAM.

Case 3

A 38-year-old woman was the victim of a car accident causing minor cranial trauma without loss of conscious

O. D.

Figure 1. Slight perception of deafness of the right ear (case 2).

ness or fractures. Eight days later, she noticed a decrease in hearing on her left side. Five months later, she consulted a doctor for the first time, because of the appearance of left-sided tinnitus and some disorders of equilibrium. The ear drums were normal, the Babinski-Weil test showed tilt toward the left, and she had a right-sided nystagmus. There was no clinical damage of the Vth or VIIth nerve. The audiogram showed perception deafness with thresholds at 20, 25, 40, and 20 dB for the 0.5, 1, 2, and 4 kHz frequencies, respectively. The auditory potentials suggested retrolabyrinthine origin (space P1P4 = 9 msec). The electromyogram showed an abnormality of the facial and Vth cranial nerves. The MRI confirmed a stage IV tumor without any invasion of the IAM that turned out to be a meningioma (Fig. 3).

It is possible that the symptoms arose when the cranial impact caused elongation of the nerves in contact with the tumor due to the inertia and the severe relative displacement of the latter.

Case 4

A 37-year-old woman was operated on for an otosclerosis on the left side 2 years earlier. A year later, the right side was operated on. The operation did not present any particular problem, but on about the sixth postoperative day, the patient complained of rotatory vertigo. The latter persisted and required antivertigo medical treatment. An improvement in hearing was nevertheless ob-

Figure 2. Magnetic resonance imaging showing a large acoustic neurinoma pressing against the brainstem with overlying ventricular dilation. A: coronal slice; B: axial slice.

Figure 3. Magnetic resonance imaging showing a large meningioma in the posterior fossa with empty inner auditory canal. A: coronal slice; B: axial slice.

tained. Confronted with this symptomatology, a surgical revision of the right side was performed a few months later, which failed to provide any explanation. The piston through the plate was in a correct anatomic position and there was no labyrinthine fistula. In spite of this, the piston was changed, but this did not modify the vertigo, so we therefore decided to request electrophysiologic tests. They confirmed the retrocochlear nature of the left-sided lesion (interval PIP5 = 5.42 msec, interval PIP3 = 3.53). The MRI performed confirmed the existence of a stage II neurinoma. Within 3 months, this was accompanied by deterioration in hearing.

CONCLUSION

Numerous factors explain why the tumors diagnosed early (stage I) do not, in the best statistics, account for more than 10% of cases. These factors are: rapidity of evolution, initial site in the CPA and not in the IAM, absent or insufficient symptoms even at an advanced stage, misleading symptoms suggesting another etiology, or symptoms masked by that of another more apparent disease.

REFERENCES

1. Portmann M, Leman P, Cohadon F, Traissac L: A propos de l'équipe oto-neurochirurgicale. Société Française d'ORL et de Pathologie Cervico-Faciale, Paris, Oct. 1967. CR for Biol (Paris) 161: 1967
2. Duriez-Auria F: Aspects audiolgiques actuels des neurinomes de l'acoustique: À propos de 120 cas. Thèse diplôme Doctorat en Médecine, Bordeaux N° 16, 1988
3. Pech A, Triglia JM, Pellet W: Le neurinome de l'acoustique. *Encycl Med Chir ORL* (Paris) 7, 20250, 1987 (AIO 12)
4. Palva T, Troupp H, Jauhainen T: Team surgery for acoustic neurinomas and the preservation of hearing. *Acta Otolaryngol* (Stockh) 91:37-45, 1981
5. Portmann M, Bébér JP, Dauman R, Duriez F, Portmann D: A propos des premiers symptômes cliniques du neurinome de l'acoustique. *Rev Laryngol* 109:401-404, 1988
6. Portmann M, Bébér JP, Dauman R, Duriez F, Portmann D: L'audiométrie tonale et vocale standard du neurinome de l'acoustique. *Rev Laryngol* 109:405-408, 1988
7. Dauman R, Negrevergne M, Portmann M, Bébér JP, Duriez F, Portmann D: Valeur du réflexe stapédien pour le diagnostic d'une lésion rétrocochléaire. *Rev Laryngol* 109:409-413, 1988
8. Dauman R, Bébér JP, Negrevergne M, Duriez F, Portmann D: L'exploration électrophysiologique des voies auditives dans les neurinomes de l'acoustique. *Rev Laryngol* 109:415-420, 1988
9. Portmann M, Guérin J, Bébér JP, Duriez F, Portmann D: Conservation des fonctions du nerf facial et du nerf cochléaire après exérèse des neurinomes de l'acoustique. *Rev Laryngol* 109:437-441, 1988

REVIEWER'S COMMENTS

The Portmanns' report their somewhat surprising experience that newer imaging techniques, CT and MRI, have not resulted in the detection of acoustic neuromas at an earlier stage. They speculate on the reasons for this.

I thought that their experience must be unique and this stimulated me to review the experience of the physicians of the House Ear Clinic over a similar time period. I reviewed 3 years: 1975, 1980, and 1991; 1975 was pre CT, 1980 was pre MRI.

In 1975, 53 tumors were removed by members of the House Ear Clinic. The average size of these tumors was 1.99 cm. In 1980, 106 tumors were removed with an average size of 2.47 cm. In 1991, 153 tumors were removed. Their average size was 2.24 cm. There is no significant difference in the average size of the tumors for the three time periods.

One can only speculate as to why improved imaging techniques have not resulted in a significant decrease in the size of acoustic neuromas when detected. I agree with the authors that it is most likely that these tumors simply do not manifest clinical symptoms until they are quite large in many cases. Otolaryngologists have been trained to pursue even minor auditory or vestibular symptoms with whatever techniques are available to come to a definitive conclusion that a tumor is not present. Even though imaging techniques have improved, the techniques that were available (Pantopaque myelography and air contrast CT) were as accurate in detecting tumors even though they were more uncomfortable for the patient.

The authors' findings again emphasize the importance of pursuing even relatively minor auditory or vestibular symptoms with appropriate diagnostic and imaging studies.

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REVIEWER'S COMMENTS

The authors observe an interesting paradox obtained from a longitudinal analysis of patients referred to them with acoustic neurinoma. They observe that a larger percentage of recent referrals have larger tumors, an observation that is unexplained by the development of more sensitive and site-specific diagnostic tests in recent years. This is an interesting observation that may be explained perhaps by, among other things, the change in patient referral patterns or a more widely available use of sensitive noninvasive imaging tests. As the authors suggest, a larger number of tumors, recently detected almost incidentally as unrelated sensorimotor disorders, are investigated by brain MRI or CT imaging. In earlier years those patients might very well have been ignored by functional otoneurologic examinations of restricted availability, whereas today such unrelated or comorbid symptoms might prompt immediate MRI or CT.

The authors are to be commended for reporting an interesting observation. Their report might stimulate others to search for explanations of this paradox.

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