Chronic arachnoiditis in the posterior fossa: a study of 82 cases

ZHANG RONGXUN

From the Department of Neurosurgery, Second Affiliated Hospital of Hebei Medical College, Shijiazhuang, People's Republic of China

SUMMARY Arachnoiditis in the posterior fossa is not a rare disorder in China. Eighty-two cases seen in a three year period were studied with reference to symptomatology, aetiology and treatment. The presentation was contrasted with that of increased intracranial pressure caused by tumour. The efficacy of treatment by direct exploration and shunt surgery was emphasised.

Chronic arachnoiditis in the posterior fossa is not rare, particularly in northern China. Most of the patients are young and very few are over the age of 50 years. The illness is usually sub-acute or chronic with long intermissions between relapses which are finally recognised as due to "increased intracranial pressure". The outcome of correct management is usually satisfactory. Between 1970 and September 1981, 82 patients with chronic arachnoiditis in the posterior fossa were admitted to this department. All the cases were confirmed by operation and, in most cases, by histological examination of removed tissue. These patients constituted 0-94% of 8708 neurosurgical cases admitted during the same period. They also constituted 22.84% of 359 who had posterior fossa operations with increased intracranial pressure.

Patients

There were 50 males and 32 females. Their ages at the time of diagnosis ranged from 5-55 years, with 78% (64) of the patients between 18 and 50 years. Fifty-six patients were peasants, six workers, one cadres, one worker's relative and 18 were children of whom 17 came from rural areas and one from the city. In the whole series 73 patients (89%) came from rural areas and only nine (11%) from the city. Thirty-six cases had previous relevant medical history. Otitis media has occurred in 11, cranio-cerebral trauma in seven, systemic infection in 10, sinusitis in one, pneumonia in one, pleurisy in one and cerebro-vascular accident in three. Intoxication by farm insecticides was recorded in two cases.

Address for reprint requests: Zhang Rongxun, Department of Neurosurgery, Second Affiliated Hospital of Hebei Medical College, Shijiazhuang, People's Republic of China.

Received 10 June 1981 and in revised form 16 November 1981
Accepted 12 December 1981

Table 1 Symptoms in 82 cases with chronic arachnoiditis in the posterior fossa

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>No of cases</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Headache</td>
<td>78</td>
<td>95.1</td>
</tr>
<tr>
<td>Vomiting</td>
<td>46</td>
<td>56.1</td>
</tr>
<tr>
<td>Visual disturbance</td>
<td>32</td>
<td>39.0</td>
</tr>
<tr>
<td>Hearing loss &amp; tinnitus</td>
<td>20</td>
<td>24.4</td>
</tr>
<tr>
<td>Unsteadiness of gait</td>
<td>20</td>
<td>24.4</td>
</tr>
<tr>
<td>Vertigo</td>
<td>10</td>
<td>12.2</td>
</tr>
<tr>
<td>Limb weakness</td>
<td>6</td>
<td>7.3</td>
</tr>
<tr>
<td>Epilepsy</td>
<td>4</td>
<td>4.9</td>
</tr>
<tr>
<td>Intermittent coma</td>
<td>4</td>
<td>4.9</td>
</tr>
<tr>
<td>Limb numbness</td>
<td>2</td>
<td>2.4</td>
</tr>
<tr>
<td>Face numbness</td>
<td>2</td>
<td>2.4</td>
</tr>
<tr>
<td>Obesity</td>
<td>2</td>
<td>2.4</td>
</tr>
</tbody>
</table>

Table 2 Signs in 82 cases with chronic arachnoiditis in the posterior fossa

<table>
<thead>
<tr>
<th>Signs</th>
<th>No of cases</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Papilloedema</td>
<td>78</td>
<td>95.1</td>
</tr>
<tr>
<td>Neck stiffness</td>
<td>10</td>
<td>12.2</td>
</tr>
<tr>
<td>Ataxia</td>
<td>9</td>
<td>11.0</td>
</tr>
<tr>
<td>Nystagmus</td>
<td>9</td>
<td>11.0</td>
</tr>
<tr>
<td>Abducent nerve paralysis</td>
<td>5</td>
<td>6.1</td>
</tr>
<tr>
<td>Auditory nerve disturbance</td>
<td>4</td>
<td>4.9</td>
</tr>
<tr>
<td>Doubtful nystagmus</td>
<td>3</td>
<td>3.7</td>
</tr>
<tr>
<td>Glossopharyngeal nerve paralysis</td>
<td>3</td>
<td>3.7</td>
</tr>
</tbody>
</table>
CSF findings
Before operating, lumbar puncture was performed in 34 cases. The cerebrospinal fluid pressure was under 150 mm H2O in two cases, ranged from 250-300 mm H2O in 20 and from 300-500 mm H2O in 12. In the cerebrospinal fluid, the white blood count was under 10/mm3 in 18 cases. Twenty-one cases had no white blood cells in the CSF. The CSF had a normal protein value (0-3 g/l or less) in six cases. The protein value ranged from 0-5-3-6 g/l in 35 cases. In all the sugar and chloride levels were normal.

Radiography
Skull films showed evidence of increased intracranial pressure in 24 cases, others were negative. Carotid angiography showed hydrocephalus in varying degrees in 59 cases. Ventriculography was performed in all the 82 cases. The manifestations varied with the anatomical types of the lesion: (1) adhesive obstructive arachnoiditis of the cerebellomedullary cistern was shown by the enlargement of the whole ventricular system without displacement together with and filling of the cerebellomedullary cistern and/or the upper portion of the cervical canal. There was no flow of the contrast material over the cerebellar surface. Sometimes the arachnoid in the cerebellomedullary cistern was like a cyst. In some cases the lateral ventricles and the third ventricle were enlarged but with little or no filling of the aqueduct. Fifty-nine cases (71.9%) were of this type. (2) obstruction of the foramen of Magendie caused dilation of all the ventricles, but the cerebellomedullary cistern was not filled. Eighteen cases (21.9%) were of this type. (3) the shape of the cerebellopontine angle showed the presence of a space-occupying lesion in five cases (6.1%).

Operation
Posterior fossa decompression and removal and/or dissection of the thickened membranous arachnoid in the cerebellomedullary cistern were carried out in 59 cases, among which Torkildsen's procedure was carried out in four cases at the same time. Posterior fossa decompression and dissection of the membranous adhesion of the foramen of Magendie were carried out in 12 cases. Posterior fossa decompression and splitting the vermis were carried out in six cases. Removal of the arachnoid cyst wall and dissection of adhesion in the cerebellopontine angle were carried out in five cases. After operation there was recurrence of adhesions in eight cases, among which a ventriculoperitoneal shunt, through a silicone tube no 8 or 10, was employed in five cases.

Pathology
Pathological examination was performed in 61 cases. Macroscopically the arachnoid was thickened, pale and opaque. In the cerebellomedullary cistern the thickened arachnoid resembled a cyst in form. In some serious cases it had a strong resemblance to an opaque plastic membrane in small patches. The adhesive membranes sometimes spread to the bottom of the fourth ventricle and the lower end of the aqueduct. Sometimes, by permitting the ingress of CSF and impeding its egress, an arachnoid cyst formed in the cerebellopontine angle.

Microscopically, sections of arachnoid contained many chronic inflammatory cells, in which the predominant type was lymphocytes. Initially the exudate was characterised by extensive lymphocytic infiltration mostly around small blood vessels. In more advanced cases the exudate became more diffuse and was accompanied by increasing amounts of fibrin. The arachnoid became fibrillary and hyperplastic. Sometimes there were papillary change with hyperplasia (fig 1). In some cases, massive fibrillary hyperplasia and extensive chronic inflammatory infiltration could be found at the same time (fig 2). Later, the arachnoid showed marked hyperplasia but the chronic inflammatory infiltration decreased. Then only massive hyperplastic fibrous tissue could be found (fig 3).

Results
Sixty cases (73.2%) were cured, eight (9.8%) were improved, two (2.4%) were left with severe disability and 12 cases (14.6%) died. Deaths after operation occurred from pneumonia in five cases, acute meningitis in two, heart failure in one and adhesion recurrence in four.

Review of 48 cases followed up from six months
Fig 2  Arachnoid showing a massive fibrillary hyperplasia and an extensive chronic inflammatory infiltration. (Haematoxylin and eosin × 90)

Fig 3  The arachnoid contains massive hyperplastic fibrous tissue and infiltration by a few lymphocytes. (Haematoxylin and eosin × 90)

to seven years, showed that the late result was good in 41 cases who had returned to school or former occupation, and was fair in six cases who were able to perform the tasks of daily living but were no longer able to work or attend school. One patient died at home from general metabolic disturbance and bedsores with infection.

Discussion

In 1889, Maunsell¹ first reported an arachnoid cyst in the posterior fossa. In 1932, Craig² reported chronic cystic arachnoiditis, described the clinical manifestations and ascribed the disease to inflammatory reaction of the arachnoid. In 1958 and 1959, Ogleznev³ and Laponogov⁴ reported their experience with surgical treatment of this disease. Later, the clinical features, pathogenesis and treatment of the condition occasionally were described, but large collections of patients were reported rarely.⁵⁻⁷

In the present series, most of the patients were young or in the prime of life, with 78.1% (64) of them between the ages of 18–50 yr. The incidence in rural areas was much higher than that in the city. Thirty-six cases had a medical history suggestive of pathogenic factors, which included otitis media in 11 cases, craniocerebral trauma in seven, systemic infection in 10, sinusitis in one, pneumonia in one, pleurisy in one and cerebrovascular accident in three. Those factors were approximately similar to other reports in the literature.⁷⁻⁹ Two cases had a history of intoxication by farm insecticide before suffering from this disease. No patient had had tubercular meningitis or systemic tuberculosis.

For a cyst formed by thickened arachnoid in the cerebellomedullary cistern, many different terms have been used, such as “arachnoid cyst of the cerebellomedullary cistern”, “cystic hydrops of the posterior fossa” or “inferior midline cyst”.⁸ Usually, the superior wall, the posterior wall and the lateral walls of the cyst can be clearly seen in the operative field. But where the inferior wall near the cervical spine
ends has not been described previously, or whether the cyst communicates with the ventricular system or the subarachnoid space or both. For this reason, three cases were studied at operation. In these, a preoperative diagnosis of space occupying lesion in the posterior fossa was made by ventriculography and an arachnoid cyst of the cerebellomedullary cistern, which respectively measured $3 \times 3 \times 4$ cm, $3 \times 4 \times 2$ cm and $4 \times 4 \times 2$ cm was found at operation. The superficial arachnoid of the cyst was thickened and without lustre or transparency. There was no adhesion between the arachnoid and the dura mater. The cyst wall was carefully protected from rupture. When 10 ml of dilute methylene blue solution was injected into the lateral ventricle via a Scott cannula, the blue solution rapidly passed into the cyst. Next, lumbar puncture was carried out. The lumbar CSF respectively showed a blue colour 2 minutes, 5 minutes and 6 minutes later. Before the three operations we had examined in one case the CSF obtained from the lateral ventricle, the "cyst" and the spinal subarachnoid space. The three specimens all had normal protein, sugar and chloride and cellular content. Thus it can be seen that the so-called "arachnoid cyst of the cerebellomedullary cistern" seen at operation is only a continuation of the arachnoid elsewhere. Its inferior boundary extended farther into the spinal canal rather than only to the inferior side of the cerebellomedullary cistern. It is clear that the superior boundary communicates with the fourth ventricle and could not obstruct the ventricular system. Therefore, the term "arachnoid cyst of the cerebellomedullary cistern" gives a false impression.

Adhesions around the cerebellomedullary cistern impede the CSF circulation and cause increased intracranial pressure. If there is an absorptive defect of the CSF via arachnoid villi at the same time, the clinical effects will be more severe, but should not be attributed to mechanical compression of the "cyst". We feel that it is better to name this disease "adhesive obstructive arachnoiditis of the cerebellomedullary cistern". In this group of 59 cases, three belonged to this type. In the remaining 56 cases, after the thickened arachnoid was carefully dissected or partially removed, the dye test proved that the ventricular system communicated with the cerebellomedullary cistern.

ANATOMICAL TYPES OF THE LESION
The clinical picture and prognosis of this disease vary with the anatomical type of the lesion. These types may be listed as follows:

1. Adhesive obstructive arachnoiditis of the cerebellomedullary cistern was characterised by a chronic course and the general symptoms and the signs of increased cranial pressure. The other signs are rare. Vertigo, headache and tinnitus developed chronically and occurred for several months or years. Next, the headache became severe; nausea, vomiting and diminution of visual acuity gradually occurred. The common signs, in addition to papilloedema, included paralysis of abducens nerve, occasionally atypical ataxia. Nystagmus was rare. The CSF pressure was usually more than 250-300 mm H₂O. The CSF cellular content, sugar and chloride level were normal. Usually, the protein content was increased. At operation, the arachnoid situated in the cerebellomedullary cistern had become thickened, pale and opaque. It was similar to a cyst in form and was under tension. When the thickened arachnoid, which was usually like a cyst wall, was moved partially or dissected, a dilated foramen of Magendie could be found. Fifty-nine cases were of this type. They constituted 72% of all cases. Six of them died.

2. Obstruction of the foramen of Magendie: The general clinical manifestations of chronic headache, vertigo and tinnitus occurred for several weeks or months. Later, the symptoms were aggravated suddenly and the condition deteriorated rapidly, associated with vomiting, neck rigidity, low fever, papilloedema with haemorrhage and exudates in the fundus. Nystagmus and ataxia were present in some patients. The CSF pressure was usually more than 250-300 mm H₂O. The protein content in CSF was also increased. The cellular content was increased or at least normal. At operation, it was found that the CSF in the cerebellomedullary cistern was diminished and the arachnoid thickened. The foramen of Magendie was obstructed. If the adhesion was dissected carefully or the vermis was split the obstruction was relieved, and CSF flowed from the fourth ventricle. Eighteen cases were part of this type (21.9%). It is noteworthy that 14 of the 18 cases had bilateral herniation of the tonsils. In one case, the tonsils were at the level of C1. Six cases died after operation.

3. Cerebellopontine angle arachnoiditis had a clinical picture quite different from that of tumour in the same site although the ventriculography showed little difference between the two diseases. Long-standing increased intracranial pressure occurred first and the cerebellopontine angle syndrome appeared next; or symptoms from adhesion of the lower four cranial nerves occurred first and the loss of hearing appears next. The hearing loss was either partial or bilateral. Widening of the internal auditory canal was found rarely. The CSF pressure was more than 250 mm H₂O. The protein content was increased but the cellular content, the sugar and the chloride levels were normal. At operation the
arachnoid in the cerebellopontine angle was thickened and adhesive. It had formed an arachnoid cyst, in which the CSF had a light yellow colour. When the cyst wall was removed partially or dissected, some adhesive membranes or cords around the trigeminal, facial, auditory, and the fourth nerves were found. The adhesions were carefully dissected. Postoperatively, the symptoms and signs of increased intracranial pressure and cranial nerves were much improved. Five cases were of this type (6·1%). All recovered and were discharged.

OPERATIVE METHODS
If only cranial nerve symptoms were present the patients could be mainly treated by combined traditional Chinese and Western medicine if increased intracranial pressure and herniation of tonsils could be ruled out. If there had been increased intracranial pressure, operative treatment was undertaken whether there were symptoms of cranial nerves and the herniation of tonsils or not.

The method of therapy varied with the anatomical types of the lesion. The purpose of operative treatment was to relieve the obstruction of the CSF circulation and improve the CSF absorption and to direct the excessive CSF into suitable channels and decrease the intracranial pressure.

1. Adhesive obstructive arachnoiditis of the cerebellomedullary cistern was treated by posterior fossa decompression. The thickened arachnoid in the cerebellomedullary cistern, which was usually like a cyst wall, was removed or dissected as much as possible. Postoperatively, the majority of patients were treated by corticosteroids, usually with combined traditional Chinese and Western medicine. If the patients' symptoms postoperatively had not improved, or if they recurred after improvement, various methods of guiding the CSF through a catheter to the aricrobe, the peritoneum or from the spinal subarachnoid space to the retroperitoneal tissue were used.10-13 In this series, there was adhesion recurrence in eight cases. Five of them had a ventriculoperitoneal shunt through a silicone catheter instead of the Holter valve. Postoperatively four cases recovered and were discharged; one died from pneumonia in hospital. The four cases, followed up from seven months to three years, were all able to perform the tasks of daily living and work. The remaining three cases who refused to accept the shunt procedure all died from recurrent intracranial hypertension. (2) Obstruction of foramen of Magendie: if the foramen had a membranous adhesion which could be dissected easily, it was removed carefully. When the adhesion was too severe to dissect, splitting the vermis was thought to be justified as a proper measure without damage to the medulla oblongata. It is to be noted that the laminae of the cervical vertebrae at the level of the herniation of the tonsils, was removed; the dura and arachnoid above the herniated tonsils were opened sufficiently to relieve compression of the brain stem. (3) Cerebellopontine angle arachnoiditis: if an arachnoid cyst had formed, the cyst wall was removed as much as possible and the adhesions around the cranial nerves dissected carefully. Usually the results in these cases were satisfactory.

References