Plasmacytoma of the clivus presenting with an unusual combination of symptoms

Case report

CRISTIAN L. VERÁ, M.D., LUDWIG G. KEMPE, M.D., AND JAMES M. POWERS, M.D.

Departments of Neurosurgery and Pathology, Medical University of South Carolina, and Section of Neurosurgery, Charleston Veterans Administration Hospital, Charleston, South Carolina

A case of plasmacytoma of the clivus is presented. The patient was asymptomatic until a coincidental traumatic lesion produced manifestations of vagus nerve dysfunction. Conduction was blocked in some fibers of the vagus nerve while, at the same time, episodes of activity were triggered in fibers of the same nerve trunk, resulting in an unusual combination of symptoms. The symptomatology of the case is discussed.

KEY WORDS □9 plasmacytoma □9 vagus nerve □9 cardiac arrest □9 clivus □9 skull tumor

We are reporting a case of clivus plasmacytoma in a patient who was asymptomatic until he sustained a coincidental head injury in a car accident. He then developed paralysis of the left vocal cord and episodes of cardiac arrest when he attempted to sit or stand.

Conduction was apparently blocked in one group of fibers of the vagus nerve, while activity was triggered with abnormal timing in another group of fibers of the same nerve. This situation produced an unusual combination of symptoms. The anatomical disposition in the posterior fossa of the rootlets composing the vagus nerve can explain a coexistence of symptoms that might seem mutually exclusive.

Case Report

This 49-year-old, right-handed, normotensive, non-diabetic man was asymptomatic until November 5, 1976, when he backed his truck into a tree. His head was thrown about violently, but no direct impact was reported, and he did not lose consciousness. He was asymptomatic after the accident except for some pain in the back of his neck. During the following 3 days, he developed a rasping throat sensation with coughing, hoarseness, and the production of clear mucous sputum. He also noticed progressive difficulty in swallowing solid food. He had no headaches, diplopia, or vomiting. He fainted several times while trying to stand up or sit up in bed.

He was taken to a hospital where he was found to have a bradycardia of 46 beats/minute. When he sat or stood up, his heart rate fell to 30 beats/minute, and this was followed by several seconds of cardiac standstill and “fainting.” Concomitantly, his blood pressure dropped from 130/80 to 90/60 mm Hg. His condition improved when he lay down. Electrocardiogram (EKG) was reported as showing sinus bradycardia with the patient in the horizontal position. Cervical spine x-ray films were reported as normal. Skull x-ray films were considered unsatisfactory, but no fracture lines were seen over the vault. Laboratory studies showed 3.8 million red blood cells/cu mm, with 8000 white blood cells, and a normal differential. Concentrations of sodium and potassium in the serum were within normal limits.

Examination. The patient was subsequently seen in neurosurgical consultation. The previous findings were confirmed. The patient’s bradycardia was not found to be permanent. His heart rate rose to over 80 beats/minute when he experienced pain or agitation while in the horizontal position. His hearing was slightly impaired on the left side, and a left hemotympanum was seen. The soft palate drooped a little on the left, and the posterior wall of the pharynx bulged slightly. Gag reflex was very active on both
Operation. With the pacemaker in place and with the patient under general endotracheal anesthesia, a low left temporal craniotomy was performed. The left edge of the tentorium cerebelli was divided. The prepontine dura mater on the floor of the posterior fossa was intact, although somewhat tense. This portion of the dura was incised, and a bloody, friable tumor, which had destroyed the clivus, became visible. Microscopic examination showed neoplastic plasma cells.

Postoperative Course. Recovery was uneventful except for a transient paresis of the left side of the tongue and left sternocleidomastoid muscle. Bone marrow aspiration was performed twice within 1 month after the tumor biopsy. The first was cytologically normal; the second demonstrated a mild plasma cell pleocytosis. Three weeks postoperatively, a course of radiotherapy was started with 3500 rads delivered to the tumor area.

The constant rasping sensation of the patient's pharynx, his cough, mucoid sputum, and dysphagia persisted for several months. They cleared completely 1 year after the biopsy. The left vocal cord paralysis cleared only partially after 3 years, but the hoarseness of his voice diminished considerably. The bulging of the posterior wall of the pharynx remained unchanged. No new bone formation or sclerosis was apparent on control x-ray films of this area. The serum protein electrophoresis curve has not shown a monoclonal spike (Table 1). Immunoelectrophoresis of serum proteins and of concentrated urine disclosed a gammopathy of immunoglobulin A and lambda light chains (Fig. 3). A similar study carried out 14 months later gave the same results.

Radiological examination of the rest of the skeleton was negative for other myeloma lesions 2 weeks after the operation, and again 1 year later. A CT scan performed 14 months postoperatively failed to show the retropharyngeal, prepontine tumor.

Discussion

Cranial myelomas, especially of the skull base, occur infrequently. These tumors usually consist of multiple lesions ("multiple myeloma"). The designation of our case as a "solitary myeloma" seems justified at present, since no other skeletal lesions could be detected in repeated skeletal roentgenological surveys. However, those who have studied large series of these tumors believe that "solitary myelomas" will disseminate if given sufficient time. Nevertheless, some well documented cases of single tumors of this kind have been observed for very long periods. Although this tumor had extensively invaded the cranial base, the patient remained asymptomatic until a fracture through the tumor-invaded skull base produced the hemorrhage visible otoscopically and at surgery. Such an event is not surprising in view of the
Plasmacytoma of the clivus

**TABLE 1**

Serum protein values compared with normal values*

<table>
<thead>
<tr>
<th>Factor</th>
<th>Value (gm%)</th>
<th>Normal Value (gm%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>albumin</td>
<td>3.4</td>
<td>3.6-5.3</td>
</tr>
<tr>
<td>A1 globulin</td>
<td>0.3</td>
<td>0.04-0.19</td>
</tr>
<tr>
<td>A2 globulin</td>
<td>1.4</td>
<td>0.3-0.8</td>
</tr>
<tr>
<td>B globulin</td>
<td>1.6</td>
<td>0.61-1.3</td>
</tr>
<tr>
<td>G globulin</td>
<td>1.2</td>
<td>0.5-1.8</td>
</tr>
</tbody>
</table>

*Normal figures based on data obtained in 1972 from blood donors from Charleston, South Carolina.
most often skull base myelomas block conduction through somatomotor axons presenting with cranial nerve palsies.\textsuperscript{7} Occasionally, however, tumors and other lesions can make motor fibers fire bursts of impulses, producing manifestations such as hemifacial spasm.\textsuperscript{10,14,22}

The nerve rootlets forming the vagus trunk enter the jugular foramen at different angles.\textsuperscript{14} Somatomotor laryngeal rootlets\textsuperscript{6} and those to the upper esophagus\textsuperscript{11} originate more caudally and bend sharply over the edge of the foramen. Such angulation offers the geometrical conditions for pressure on them to produce a conduction block. Those vagal rootlets originating more cephalad do not bend or bend little over the edge of the foramen. They could not have been blocked, since no sensory deficit could be demonstrated in their area of distribution (territory of the superior laryngeal nerve).\textsuperscript{20}

We suggest, then, that this patient's lesions produced a paralysis of the left vocal cord and dysphagia by mechanically blocking conduction through the appropriate vagal somatomotor nerve fibers, while the same mechanical factors triggered episodes of cardiac standstill by initiating hyperactivity in thinner cardiac efferent fibers coursing in the same vagus nerve trunk.

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Address reprint requests to: Cristian L. Vera, M.D., Department of Neurosurgery, Medical University of South Carolina, 171 Ashley Avenue, Charleston, South Carolina 29403.