Tapia's syndrome caused by a neurofibroma of the hypoglossal and vagus nerves

Case report

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Tapia's syndrome is characterized by unilateral paralysis of the tongue and vocal cord, and is caused by a lesion of the 10th and 12th cranial nerves below the nodose ganglion, without involvement of the pharyngeal branches of the 10th nerve. The authors report the case of a 25-year-old man who presented with a 4-year history of progressive glossolaryngeal paralysis. Operation through the right laterocervical region allowed complete removal of a neurofibroma involving the 10th and 12th nerves at their crossing below the nodose ganglion.

KEY WORDS • Tapia's syndrome • neurofibroma • vagus nerve • hypoglossal nerve

Case Report

This 25-year-old man came to our attention in June, 1977, complaining of progressive dysphonia for 4 years. There was no history of trauma to his head or neck. Clinical examination showed palsy of the right vocal cord, and paralysis and atrophy of the right side of the tongue. The movements of the pharynx and soft palate were normal, and no other cranial nerve was involved. Radiography of the base of the skull, angiography, and scintigraphy were all negative.

Based on the clinical picture, surgical exploration of the right laterocervical region was carried out. Through an incision along the anterior edge of the sternocleidomastoid muscle in the upper laterocervical region, a mass was found at a depth of about 2 cm. The tumor, like a small gray olive, wrapped and infiltrated the 10th and 12th cranial nerves at their crossing below the nodose ganglion (Fig. 1). Dissection of the fibroelastic neoplasm showed an anastomotic branch between the hypoglossal and vagus nerves. Further separation of the mass from the nerve fibers was impossible, so a complete removal

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Neurofibroma of hypoglossal and vagus nerves

**TABLE 1**

* Syndromes involving the last four cranial nerves *

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Cranial Nerves Involved</th>
<th>Etiology</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vernet's syndrome</td>
<td>IX, X, &amp; XI</td>
<td>neoplasms: neurinomas, lymphatic metastases, chondromas, tumors of the ear, nose; glomus tumors</td>
</tr>
<tr>
<td>(jugular foramen)</td>
<td></td>
<td>infections: arachnoiditis, osteitis, lymphatic infections, infections from the ear, nose &amp; throat vascular: phlebitis of the jugular vein, arteriovenous aneurysms</td>
</tr>
<tr>
<td>Avellis' syndrome</td>
<td>X (ambiguous nerve) or accessory to the X branch of the XI</td>
<td>injuries: very infrequently other causes: basilar compression, sarcoidosis</td>
</tr>
<tr>
<td>(palatolaryngeal hemiplegia)</td>
<td></td>
<td>injuries, lymph nodes, tumors, vascular (brain stem)</td>
</tr>
<tr>
<td>Schmidt's syndrome</td>
<td>X-XI</td>
<td>injuries, lymph nodes, tumors, vascular (brain stem)</td>
</tr>
<tr>
<td>(scapulolaryngeal hemiplegia)</td>
<td></td>
<td>injuries, lymph nodes, tumors, vascular (brain stem)</td>
</tr>
<tr>
<td>Jackson's syndrome</td>
<td>X (inferior ganglion) (XI-XII)</td>
<td>tumors: of the ear, parotid, of the base of the skull, adenopathies, reticulosis injuries vascular: aneurysms of the carotid artery, phlebitis infections: cellulitis of the neck</td>
</tr>
<tr>
<td>Collet-Sicard syndrome</td>
<td>IX, X, XI, &amp; XII</td>
<td>tumors: parotid, posterior nasal space, lymph nodes, lymphosarcomas infections: pharyngeal abscesses injuries</td>
</tr>
<tr>
<td>Villaret's syndrome</td>
<td>IX, X, XI, &amp; XII (cervical sympathetic)</td>
<td>injuries; tumors of parotid, reticulosis</td>
</tr>
<tr>
<td>Tapia's syndrome</td>
<td>X (below inferior ganglion), XII</td>
<td></td>
</tr>
</tbody>
</table>

was obtained by resection of the two nerves for about 4 cm.

The postoperative course was uncomplicated, and neurological signs remained unchanged. Paraffin sections of the tumor showed a mixed schwannomatous and fibromatous tissue, including fascicles of demyelinated fibers of the two nerves (Fig. 2). The tumor had infiltrated the lower structures of the nodose ganglion. The diagnosis was neurofibroma. No sign of multiple neurofibromatosis was found in the patient.

**Discussion**

True Tapia's syndrome is characterized by hemiplegia and wasting of the tongue and ipsilateral paralysis of the vocal cord. Sometimes there may be associated paralysis of the sternocleidomastoid and trapezius muscles or involvement of the cervical sympathetic nerves (Horner's syndrome).

Tapia first described three cases of glossolaryngeal hemiplegia in 1905. Two of these cases occurred in toreadors who were injured by bulls' hooves behind the angle of the jaw; in the third case, the patient had a tumor of the parotid gland. Other similar cases have

*Fig. 1. The drawing represents the hypoglossal and vagus nerves at their normal crossing in the neck (dotted lines). The neurofibroma can be seen wrapping the two nerves below the nodose ganglion. An anastomotic branch between the 10th and 12th nerves was found at surgery.*
The origin of the neurofibroma from the vagus or the hypoglossal nerve cannot be established. Clinical history favors the vagus nerve, but palsy of the vocal cord is much more evident clinically than palsy of the tongue. On the other hand, at surgery the hypoglossal nerve appeared more directly involved.

Our case illustrates that the association of neurological signs affecting only the 10th and 12th cranial nerves, without involvement of the pharyngeal branches of the 10th nerve, indicates such a precise localization that surgical exploration is advisable, even if clinical or radiological findings of neoplasm are absent.

References


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