Basilar Invagination Associated with an Intercarotid Paraganglioma
—Case Report—

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Abstract

A 24-year-old female presented with basilar invagination and kyphoscoliosis of the cervical spine associated with a large intercarotid paraganglioma. She had suffered from pharyngeal discomfort from the age of 9 years due to the tumor. The tumor had originated from the right carotid body and extended in the parapharyngeal space compressing the upper cervical spine. Presumably the slowly growing tumor had caused the kyphoscoliosis and disturbed osseous development of the occipito-atlanto-axial complex, resulting in anterior basilar invagination, hypoplasia of the clivus, and aplasia of the posterior arch of the atlas.

Key words: basilar invagination, atlanto-axial joint, intercarotid paraganglioma, kyphosis, scoliosis, parapharyngeal tumor

Introduction

Paragangliomas are rare tumors of the extra-adrenal paraganglion system, which consists of numerous nests of neural crest-derived cells, scattered in the head, neck, thorax, and abdomen, in association with the autonomic ganglia. Paragangliomas are classified according to the anatomic position and further divided into functional or non-functional, depending on the ability to produce catecholamines.

Intercarotid paraganglioma is a well-known but rare neoplasm in the neck. Most intercarotid paragangliomas occur in adults, but may appear at any age. They are generally benign, and the clinical course is slow. A painless palpable neck mass may be the only sign, but some patients present with local discomfort, pain, hoarseness, or dysphagia. The interval between onset of symptoms and diagnosis is long; 4 to 8 years on average, and sometimes decades.

We describe a 24-year-old patient who presented with a large intercarotid paraganglioma, associated with craniovertebral maldevelopment and cervical kyphoscoliosis.

Case Report

A 24-year-old female sought medical attention for a headache. She had a 15-year history of pharyngeal discomfort and difficulty in swallowing beginning at the age of 9 years. Physical examination found a soft, large (6 x 7 cm) mass in the right upper cervical region. Laryngoscopic examination revealed a right posterolateral parapharyngeal mass causing stenosis of the laryngeal pharynx. Her blood pressure was normal. Laboratory examination showed no abnormalities, and the levels of catecholamines and their metabolites were normal (urinary vanillylmandelic acid 7.3 mg/24 hr, urinary homovanillic acid 4.9 mg/24 hr, urinary epinephrine 10.2 MIC/24 hr, urinary norepinephrine 48.8 MIC/24 hr, urinary dopamine 500 MIC/24 hr, serum epinephrine <0.01 ng/ml, serum norepinephrine 0.05 ng/ml, and serum dopamine <0.02 ng/ml).

Radiography of the cervical spine demonstrated...
marked kyphosis and scoliosis to the left, in addition
to a soft tissue mass (Fig. 1). Lateral craniovertebral
tomography showed anterior basilar invagination
with platybasia and short clivus. The entire anterior arch of the atlas is above Chamberlain's line. Aplasia of the posterior arch of the atlas and the persistent apical ossification center of the odontoid process are also seen.

Fig. 1 upper: Cervical radiograph showing a soft
tissue mass (arrow) and scoliosis to the left.
lower: Lateral craniovertebral tomogram
demonstrating anterior basilar invagination
with platybasia and short clivus. The entire anterior arch of the atlas is above Chamberlain's line. Aplasia of the posterior arch of the atlas and the persistent apical ossification center of the odontoid process are also seen.

located upward, being attached to the posterior edge of the shortened clivus anteriorly, and to the persistent apical ossification center of the odontoid process posteriorly. The posterior arch of the atlas was aplastic. Magnetic resonance imaging disclosed a demarcated tumor attached to and compressing the cervical spine at the C-2 to C-6 levels (Fig. 2). Digital subtraction angiography showed a hypervascular tumor in the right peripharyngeal space, mainly fed by the external carotid artery. The mass was located between the right external and internal carotid arteries, displacing the external carotid artery anteromedially and the internal carotid artery posterolaterally.

She underwent excision of the tumor. The very vascular, well-demarcated tumor was attached to both internal and external carotid arteries, especially at the bifurcation. The tumor extended to the anterior aspect of the C-2 to C-6 vertebral bodies. The tumor was carefully dissected away from the internal carotid artery and totally removed.

Histological examination disclosed the "Zellballen" (cell balls) pattern of epithelioid cells with pleomorphic nuclei embedded in a fibrous stro-
ma and numerous vascular spaces (Fig. 3). The diagnosis was benign non-functioning intercarotid paraganglioma.

She was discharged without neurological abnormality 2 weeks after the operation. Two years later, cervical radiography demonstrated improvement of the kyphosis.

Discussion

Basilar invagination, the most common congenital anomaly of the atlanto-occipital articulation, usually results from hypoplasia of the occipital bone and/or upward bulging of the margins of the foramen magnum, but may occur secondary to trauma or malacic change of the skull base bones, such as Paget’s disease, fibrous dysplasia, or osteomalacia. Primary basilar invagination may be associated with various vertebral anomalies. The pathogenetic mechanism of occipital hypoplasia and basilar invagination is complex and probably involves more than one pathological event interfering with the osseous development. The occipital bone consists of more than 10 embryological parts, and three to five vertebral segments also participate in this process. The osseous development of the cervico-occipital region is not completed in the neonate but continues for several years after birth, and sometimes up to the middle of the second decade. In the atlas, two ossification nuclei of the anterior arch appear at about 1 year of age, and these nuclei fuse with the lateral masses by 6 to 9 years of age. The ossification centers of the posterior arch fuse much later, sometimes later than the age of 10 years. The odontoid process develops the apical epiphysis after the age of 2 years, and this fuses with the main portion of the odontoid by 12 years. The apical center of the odontoid process may be observed up to the age of 12 years in normal children.

A previous case of basilar invagination evolved over 18 years (from 2 to 20 years of age). In the present patient, the large tumor mass had existed from the age of 9 years, so the compressive force by the mass probably only interfered with the later

Fig. 3 Photomicrograph of the tumor tissue showing the typical “Zellballen” pattern. HE stain, original magnification ×100.

Fig. 4 Scheme showing the proposed pathogenesis in the present patient. left: Normal anatomy. center: The first step is dislocation of the odontoid process from the articular facet during flexion. The anterior arch of the atlas then compressed the basilar occiput upward, resulting in disturbance of the osseous development of the basilar occiput (double arrow), as well as narrowing of the posterior atlanto-occipital space, causing aplasia of the posterior arch of the atlas (arrowheads). right: Eventually a new atlanto-axial joint was reconstructed with the pseudo-tip of the odontoid process, i.e., the persistent apical ossification center of the odontoid process (arrow).
processes, i.e. formation of the posterior arch and the apical epiphysis of the atlas. The mechanical compression could also have caused the kyphoscoliosis. The following mechanism may have caused the complex anomalies of the present case (Fig. 4). The first step involved the displacement of the atlas and axis during the neck movement. Fixation of the upper cervical vertebrae could accentuate the upward sliding movement of the atlas relative to the axis which is observed in both the pediatric loose joint and the tighter adult joint,59 finally resulting in dislocation of the atlanto-odontoid joint. The displacement allowed the tip of the odontoid process to slip out from the articular facet during the neck movement. The anterior arch of the atlas together with the apex of the odontoid process could then have compressed the posterior edge of the clivus upward, causing fusion and invagination of the structure and leaving the ossification center of the apex. Since the posterior atlanto-occipital space became tight before the osseous fusion of the posterior arch of the atlas, the arch could not be formed. Although these steps are entirely speculative, they are consistent with the timing of the osseous development.

References

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