Bronchogenic Cyst of the Craniocervical Junction: A Case Report

Kraniyoservikal Bileşkenin Bronkojenik Kisti: Olgu Sunumu

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ABSTRACT

Intracranial bronchogenic cysts (BCs) are rare benign lesions of the central nervous system, lined with a mucous-secreting epithelium that is presumed to be of endodermal derivation (7, 14). These lesions are most frequently encountered in the cervico-thoracic region of the spine. Their pathogenesis is still poorly understood. Regardless of the surgical approach, the aim of surgery should be total removal of the cyst and its content, whenever feasible. In this case report, a 50-year-old patient with a BC of the craniocervical junction is presented. The patient was operated on through a right-sided suboccipital retrosigmoid approach. The uniform layer of pseudostratified, ciliated and mucus-secreting columnar cells was seen on histological examination. The clinical manifestations, diagnosis, and treatment of this unusual condition are discussed.

KEYWORDS: Bronchogenic cyst, Cervical, Cranial, Surgery

INTRODUCTION

Bronchogenic cysts (BCs) are rare benign lesions of the central nervous system, lined with a mucous-secreting epithelium that is presumed to be of endodermal derivation (7, 14). These lesions are most frequently encountered in the cervico-thoracic region of the spine (2, 3). Although intracranial BCs are rare and primarily found in the posterior fossa (8, 13), a few cases have been reported in various locations such as the third ventricle (6), frontal lobe (15), fourth ventricle (1), and the suprasellar area (10). BCs of the craniocervical junction are even less common and their optimal treatment is still controversial.

In this report, a case of bronchogenic cyst that was located in the anterior craniocervical junction is presented. The clinical course, surgical approach and radiological findings are discussed.

CASE REPORT

A 50-year-old woman presented with a 6-month history of intermittent occipital headaches, neck pain, syncope attacks, and sensory disturbances in her extremities. Neurological examination was normal. Magnetic resonance imaging (MRI) scans showed an extramedullary, well-delineated cystic mass located anterior to the lower pons and upper medulla, with no contrast enhancement (Figure 1A-F). Following admission, the patient was operated on through a right-sided suboccipital retrosigmoid approach. After aspiration of greenish mucinous fluid content, the thin cyst wall was dissected from the surrounding structures and excised gross totally (Figure 2A-F). Histopathological examination revealed a bronchogenic cyst (Figure 3). The patient had an uneventful postoperative course.

DISCUSSION

BCs are considered to be rare developmental malformations and their pathogenesis is still not fully understood. Symptoms associated with BCs of the craniocervical junction derive mainly from inflammatory reaction or mass effect. The most frequent complaint of patients with BCs is headaches, followed by gait disturbance, and, weakness or sensory complaints of the upper extremities (4, 11). Involvement of
cranial nerves may result in trigeminal neuralgia, hearing loss, hemifacial spasm, or facial anesthesia (4, 13).

The differential diagnosis of intracranial BCs includes epidermoid cyst, dermoid cyst, Rathke's cleft cyst, and arachnoid cyst (5, 14). The MRI characteristics of BCs can be quite variable based on the protein content of the cyst fluid. BCs are hyperintense on FLAIR and may appear isointense to slightly hyperintense compared with cerebrospinal fluid on T1WI (14), as in our case. Partial rim enhancement may be present due to reactive changes where the cyst wall is attached to brain parenchyma (14). Although MRI has been reported to be instrumental at initial diagnosis, definitive diagnosis could be made only by histological findings (9).

BCs have a variety of appearances on light microscopy. BCs may be composed of pseudostratified, ciliated, columnar or cuboidal, mucin-producing epithelium with poor cellularity. In the case presented in this study, the uniform layer of pseudostratified, ciliated and mucus-secreting columnar cells was seen on histological examination.

BCs of the craniocervical junction can be removed via a suboccipital retrosigmoid approach, as performed in our case. However, to achieve total removal of cysts located anterior to the brainstem, alternative skull base approaches were recommended (4). Ventrally and ventrolaterally located BCs can be resected through a far-lateral transcondylar approach (11). Regardless of the surgical approach, the aim of surgery should be total removal of the cyst and its content, whenever feasible (11, 12). If dissection of the cyst wall cannot be achieved due to firm adhesions to the surrounding neurovascular structures, subtotal resection is a reasonable and viable option (12). These patients, however, will require long-term follow up to detect recurrence.

Figure 1: Preoperative MRI images of the patient. A) Sagittal T1W, B) coronal T1W, C) coronal T2W, D) axial T2W, E) axial FLAIR, and F) postcontrast axial T1W images show a large, well-delineated cystic mass (stars) near the lower pons and upper medulla, without contrast enhancement. E) The cyst is hyperintense compared with the cerebrospinal fluid on FLAIR images. These images demonstrate the extra-axial origin of the mass and the brain stem displacement.
REFERENCES


Figure 2: Postoperative images obtained 3 months after operation. A) Axial T1W, B) sagittal T1W, C) axial T2W, D) axial FLAIR, E) sagittal postcontrast T1W, and F) postcontrast axial T1W images show no evidence of recurrence or residue of the cystic mass.

Figure 3: The uniform layer of pseudostratified, ciliated and mucus-secreting columnar cells was characteristic of a bronchogenic cyst (H&E x 40).


