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Case Report

Schwannoma with an Incidental Subependymoma in the Cerebellopontine Angle: A Case Report

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Abstract

Cerebellopontine angle (CPA) tumors are the most frequently diagnosed tumors of the posterior fossa, representing 6%–10% of all intracranial tumors [12]. About 5%–10% of all intracranial tumors in adults and 1% of all pediatric intracranial tumors are localized in the CPA [4,8]. Vestibular schwannomas and meningiomas are the two most frequent lesions accounting for approximately 85%–90% of all CPA tumors [12]. Subependymomas are benign tumors generally located in the ventricular system. Herein, we describe a case of a schwannoma accompanied by an incidental subependymoma at the CPA.

A 57-year-old male with a two-year history of right-side hearing loss was admitted to our clinic. Magnetic resonance imaging (MRI) revealed a mass in the right CPA. Complete surgical excision of the tumor was achieved through the retrosigmoid approach. The histopathological diagnosis was schwannoma together with a subependymoma.

Keywords: Cerebellopontine Angle; Schwannoma; Subependymoma

Introduction

CPA tumors are the most frequently diagnosed tumors of the posterior fossa, representing 6%–10% of all intracranial tumors [12]. About 5%–10% of all intracranial tumors in adults and 1% of all pediatric intracranial tumors are localized in the CPA [4,8]. Vestibular schwannomas and meningiomas are the two most frequent lesions accounting for approximately 85%–90% of all CPA tumors [12]. Apart from the poor imaging of soft tissue in computed tomography (CT), MRI has a distinct advantage over CT scans in detecting CPA tumors due to its ability to detect bone structure formations and artifacts. MRI is currently considered the gold standard in CPA tumor diagnosis, and, along with other advanced neuroradiological imaging, can help in the differential pathological diagnosis of CPA tumors.

Subependymomas typically appear to be localized in the subependymal areas of the fourth and lateral ventricles. They are classified as World Health Organization grade 1 lesions, which progress slowly and are generally detected incidentally [11]. The most common symptom of subependymomas is hydrocephalus due to obstruction caused by tumor growth inside the ventricles. These tumors are typically diagnosed in middle-aged people who are in their fifties and sixties [10].

The retrosigmoid approach provides the best wide-field visualization of the posterior fossa and a much clearer view of the inferior portions of the CPA compared to other approaches. This visualization is especially helpful when the displacement of cranial nerves is not predictable, which occurs commonly with meningiomas. The retrosigmoid approach may require cerebellar retraction or resection. However, this method induces postoperative edema, hematoma, infarction, and bleeding. Furthermore, increased incidence of cerebrospinal fluid leak has been observed in other studies, with the leak rate especially higher in cases with tumors extending into the ventricle [5].

In this case report, we describe a 57-year-old male patient diagnosed with a schwannoma with an incidental subependymoma in the right CPA.

Case Report

Clinical history and examination

A 57-year-old male patient with progressive hearing loss for two years was admitted to our clinic. Neurological examination revealed ptosis of the right eye, physiological anisocoria, and sensorineural hearing loss in the right ear. Other systems and findings were normal, and cerebellar functions were intact.

Neuroimaging findings

A contrast MRI examination of the brain demonstrated two radiologically different tumors centered in the right CPA. In the contrast series, the component compatible with acoustic schwannoma held intense contrast, while the subependymoma showed poor contrast. Using three-dimensional (3D) fast imaging employing steady-state acquisition (FIESTA) sequence, the tumor was shown to be slightly heterogeneous. Non-contrast T1A-weighted MRI showed the acoustic schwannoma as being more hypointense than the subependymoma. The subependymomal lesion minimally compressed the brain stem, but no ventricular enlargement was observed.

Surgical procedure

With the patient in a sitting position, the procedure was performed via a right retrosigmoid approach with neuromonitoring. After opening the dura, the cerebellum was gently retracted and cisterns were opened to drain the cerebrospinal fluid. The tumor, identified in an intra-arachnoid location, was hard, viscous, and more difficult to aspirate than the surrounding parenchyma. Lower cranial nerves and the posterior-inferior cerebellar artery were involved with the tumor but were easily dissected away. The tumor was excised with an ultrasonic aspirator. We identified the attachment of the tumor at the seventh cranial nerve, which was recognized as the origin of the tumor. Although the postoperative period was complicated by right-sided central facial paralysis, no electrophysiological signals suggesting facial nerve damage were further detected during intraoperative neuromonitoring. The patient was discharged without any other complications. Postoperative MRI revealed total removal of the tumor.

Histopathological findings

The excised surgical specimen comprised 3 cc of curettage material. It was fixed in formalin and embedded in paraffin. On hematoxylin-eosin examination, two specimens of tumors were detected. One of the tumors was a typical schwannoma with spindle cells presenting a fascicular pattern and obvious palisading in some areas. This tumor had neither increased cellularity nor mitosis. The second tumor specimen consisted of small, round, and uniform cells that were forming clusters in a fibrillary background. No rosette formation or perivascular arrangement was observed in this tumor (Figure 2a). Immunohistochemical examination revealed diffuse S100 staining in the schwannoma component (Figure 2b), while epithelial membrane antigen (EMA) staining revealed intracytoplasmic vacuoles in the second tumor (Figure 2c). Staining also revealed diffuse glial fibrillary acidic protein in the second tumor. Based on these findings, schwannoma with an incidental subependymoma was diagnosed.

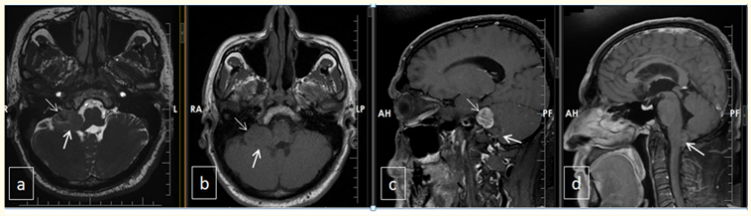

Figure 1: 56-year-old male patient; a) 3D FIESTA, b) T1A, c and d) two tumors observed with different radiological features in sagittal postcontrast T1A sequences (thin arrow: acoustic schwannoma, thick arrow: subependymoma). In contrast-enhanced series (c and d) the component compatible with the acoustic schwannoma holds intense contrast while the subependymal appears to have poor enhancement. In the non-contrast T1A series (b), the acoustic schwannom is more hypointense, whereas in the 3D FIESTA sequence (a) the tumor is more heterogeneous.

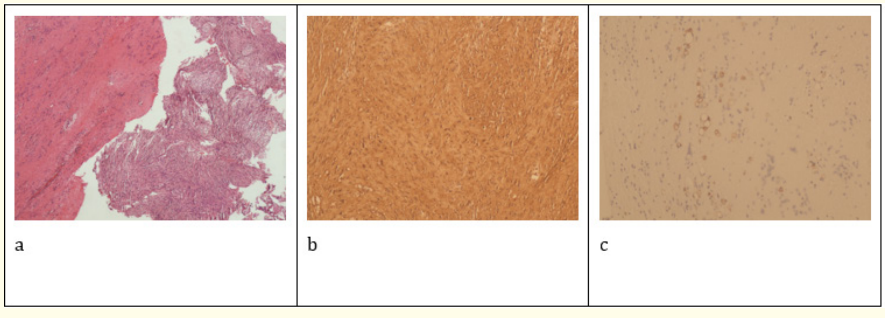

Figure 2: a: (HE x50) Schwannoma forming fascicular bundles on the right side and subependyma in the form of clumps of cells on the fibrillar floor on the left side.
b: (S100 x100) S100 staining in schwannoma.
c: (EMA x100) EMA staining intracytoplasmic lumens in subependymoma.

Discussion

A case of subependymoma detected in addition to a primary pathology and at a rare site is presented here, together with histopathological and radiological findings. The CPA and prepontine cistern involvement without extension into the fourth ventricle is uncharacteristic of subependymomas. The differential diagnosis of CPA lesions includes schwannoma, meningioma, epidermoid cyst, arachnoid cyst, aneurysm, metastasis, medulloblastoma, glioma, and ependymoma [1]. Making the diagnosis preoperatively was difficult, considering the rarity of subependymomas in the CPA and the similarity of MRI findings to other CPA tumors, especially schwannomas.

A review of the English literature on the PubMed database found only five cases of subependymomas exclusive to the CPA. Koral., *et al*. described the case of a 15-year-old boy with a lesion that was centered at the right CPA and did not extend into the fourth ventricle [9]. Cunha., *et al*. reported a 57-year-old man with a large CPA tumor that extended into the jugular foramen, but not into the fourth ventricle [3]. Romoli., *et al*. described a case of a 37-year-old male patient with the lesion arising in the bulbocerebellar angle [13]. One CPA tumor was present among the 24 intracranial subependymomas reported by Chiechi., *et al.* [2]. Jooma., *et al*. published one case of CPA subependymoma in a study involving 12 cases [7]. Hoeffel., *et al*. [11] described four subependymoma cases that extended into the CPA, but in all of them, the tumors arose from the fourth ventricle [6].

Due to the benign course of subependymomas, preoperative suspicion for it is very important in planning the operation. Even if the primary pathology has been identified, a detailed histopathological examination of all sections is important for the diagnosis of other possible existing pathologies.

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