

Two Primary Intracranial Tumors of Different Histology: Report of a Case with a Choroid Plexus Papilloma and a Concurrent Vestibular Schwannoma in the Cerebellopontine Angle

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A very rare case of multiple primary intracranial tumors is reported. A 41-year-old female patient was referred for surgery with a cerebellopontine angle (CPA) tumor. Medical history and MRI study showed typical findings of a right acoustic neuroma with a hydrocephalus. Neurological, dermatological, and ocular examinations revealed no evidence of neurofibromatosis. During surgery, a red-colored cauliflower like mass was found in the right CPA. The roof of the fourth ventricle could be seen through the lateral recess after removal of the tumor. Another mass, a 1.5-cm sized schwannoma protruding through the right internal auditory meatus, was removed by the transmeatal approach. Although the tumor masses were in contact and compressed against each other, there was a clear demarcation between them. Histological examination confirmed that the first mass was a typical choroid plexus papilloma with fibrovascular core, and that the second was a schwannoma. The patient recovered without any new neurological deficit. Result of a *Medline* search indicated that this rare combination of multiple primary tumors has not been reported previously.

Key Words: Multiple primary neoplasms, brain neoplasms, cerebellopontine angle, choroid plexus papilloma, acoustic neuroma

INTRODUCTION

While multiple primary intracranial neoplasms of the same histological type are not rare in

neurosurgical practice,¹⁻³ those of different histological types are rarely encountered.⁴ It is well known that the phakomatoses, especially neurofibromatosis and tuberous sclerosis, are associated with an increased incidence of multiple tumors of both similar and differing histology.⁵ Robinson has suggested that patients who received irradiation for a single brain tumor are at higher risk of developing a second intracranial neoplasm; in particular a meningioma.⁶ Russell and Rubinstein have stated that when the coexisting tumors are anatomically separate, it is likely to be a chance occurrence.² A discrepancy between clinical and radiographic data may alert the clinician to the possibility of dual tumors. Yet, such a discrepancy was not seen in the case reported here. Acoustic neuroma was suspected clinically, but a second tumor, choroid plexus papilloma, was found unexpectedly during surgery. These tumors were anatomically contiguous. Choroid plexus papilloma of the CPA is by itself a rare entity,⁷⁻⁹ and its coexistence with a vestibular schwannoma has to our knowledge never been reported in the literature.

CASE REPORT

A 41-year-old female patient was referred to our hospital for surgery with a recently diagnosed right CPA tumor. She had experienced progressive hearing loss starting 10 years before admission. Symptoms of headache and unsteady gait had worsened during the last two years. Neuro-

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logical examination revealed cerebellar signs such as dysdiadochokinesia and dysmetria on the right side, as well as ataxic gait. Gag reflex was diminished on the right side and right facial hypesthesia was noted. Funduscopic examination revealed papilledema. Abdominal ultrasound study was normal. No stigmata of neurofibromatosis were found on dermatological and ocular examinations. Preoperative MRI revealed a mass lesion in the right CPA with an obstructive hydrocephalus. The mass showed heterogeneous bright signals on T2 weighted image (Fig. 1.), and was well enhanced after gadolinium injection (Fig. 2.). The broad base of the mass was facing the right petrous bone. The right internal auditory canal, located at the center of the mass, was enlarged by the mass, suggesting a large vestibular schwannoma.

The patient underwent surgery in the park bench position, using the suboccipital transmeatal

approach to remove the tumor. A ventricular catheter was inserted through the posterior parietal route to drain the CSF. As the dura was opened, a reddish-gray colored cauliflower like mass with rough irregular surface was found in the right CPA. Hypertrophied branches of the right PICA fed the tumor. Gross appearance of the tumor suggested that it was not the vestibular schwannoma of the preoperative MRI studies but rather a choroid plexus papilloma with an exophytic growth in the right CPA. As the mass was gently retracted medially, a second separate mass, in contact with the choroid plexus papilloma, was found at the right internal auditory meatus. It was a yellowish, well demarcated, solid mass, suggesting a schwannoma. This second mass was clearly separated from the choroid plexus papilloma by the arachnoid membrane. The choroid plexus papilloma arose at the foramen of Luschka

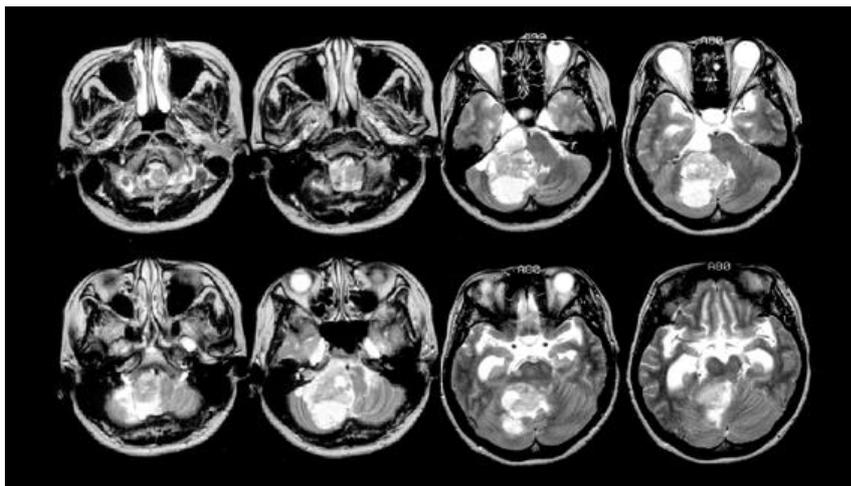


Fig. 1. Preoperative T2 weighted MRI of multiple primary intracranial tumors of different histology. MR images show heterogeneous bright signals with marked cerebellar and brain stem compression by a mass facing the petrous bone in the right CPA angle. The mass has enlarged the right internal auditory canal, suggesting a huge vestibular schwannoma.

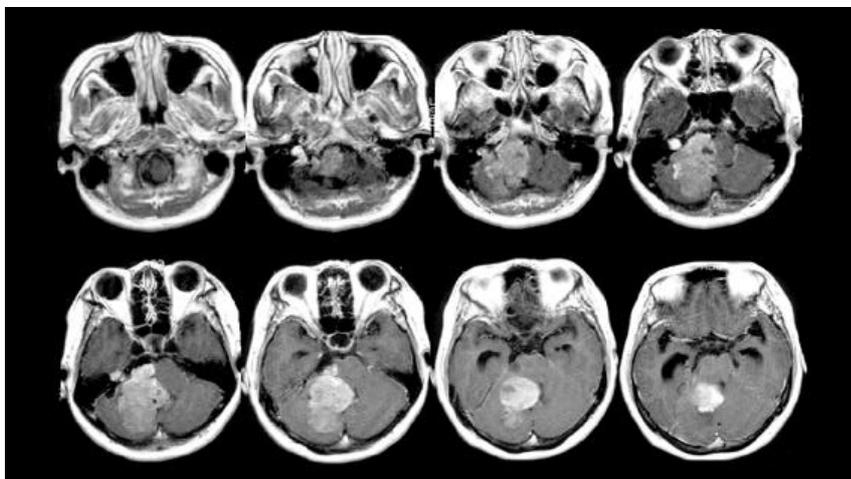


Fig. 2. Preoperative Gd enhanced MRI. Heterogenous enhancement was noted but was less heterogenous than the T2WI. Location and enhancement pattern suggest an intra-axial tumor, which is inconsistent with the mass centered in the enlarged internal auditory canal.

on the right side without an associated intraventricular component. The tumor was adherent to the choroid plexus tuft protruding through the foramen of Luschka. After removal of this choroid plexus papilloma, the roof of the fourth ventricle could be seen through the lateral recess where the tumor had been seated. The second tumor, a 1.5-cm sized schwannoma protruding through the internal auditory meatus was removed by the transmeatal approach. This funnel-shaped, firm, circumscribed tumor, originating from the right inferior vestibular nerve, occupied the auditory canal and enlarged the right internal auditory meatus. Both tumors were removed completely. External ventricular drainage, maintained to monitor the intracranial pressure, was removed

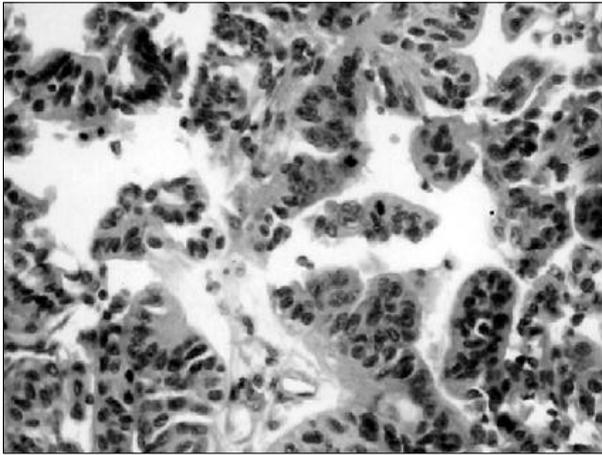


Fig. 3. Microscopic findings of the choroid plexus papilloma composed of a single layer of columnar and cuboidal epithelium supported by a stroma of vascular connective tissue (H&E, $\times 200$).

five days after surgery.

Histological examination of the two tumors disclosed a choroid plexus papilloma (Fig. 3.), and a vestibular schwannoma (Fig. 4.). Recovery was uneventful, and the patient was discharged without any newly developed deficit. A postoperative MRI study (Fig. 5, and Fig. 6.) confirmed complete removal of both tumors. The choroid plexus papilloma tumor, having appeared more like an intra-axial mass on preoperative MRI (Fig. 1 and Fig. 2), was confirmed on postoperative MR images, both T2WI (Fig. 5) and Gd enhanced (Fig. 6), to be mostly an extra-axial one, protruding through the foramen Luschka and the cerebello-medullary fissure.

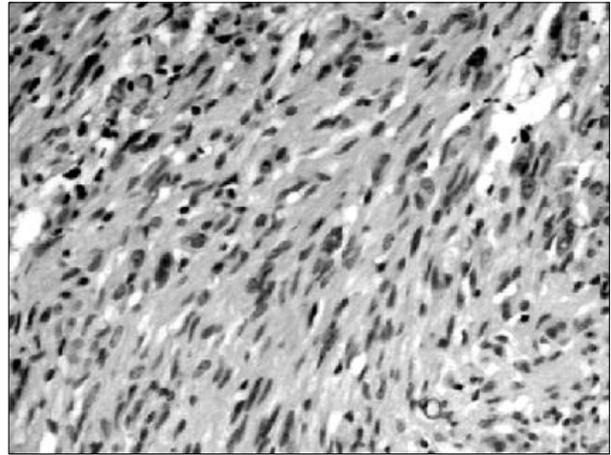


Fig. 4. Microscopic findings of the vestibular schwannoma composed of Antoni type A which have ovoid or rod-shaped central nuclei containing variable amounts of chromatic and inconspicuous nucleoli (H&E, $\times 200$).

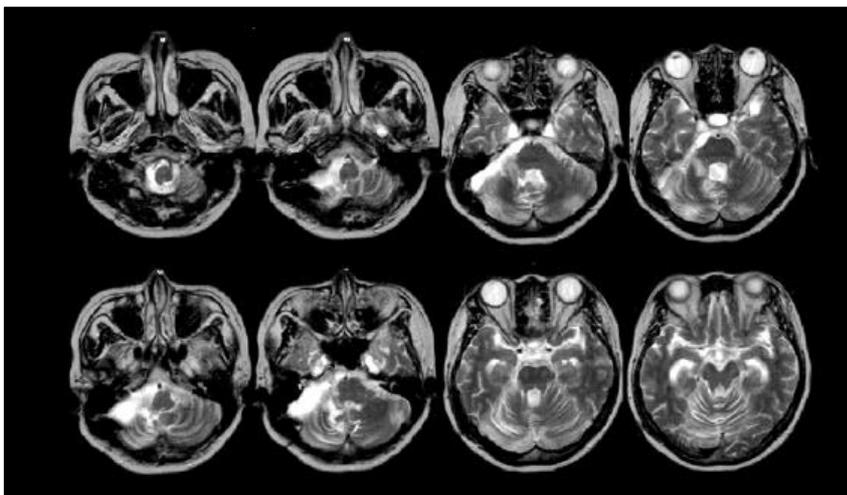


Fig. 5. Postoperative T2 weighted MRI taken three months after surgery. Previously noted tumor mass had disappeared. Widening of the right CP angle cistern can be seen, together with a prominent cerebello-medullary fissure on the right side. A small area of bright signal appears on the right middle cerebellar peduncle. The fourth ventricle is larger than normal but has normal configuration.

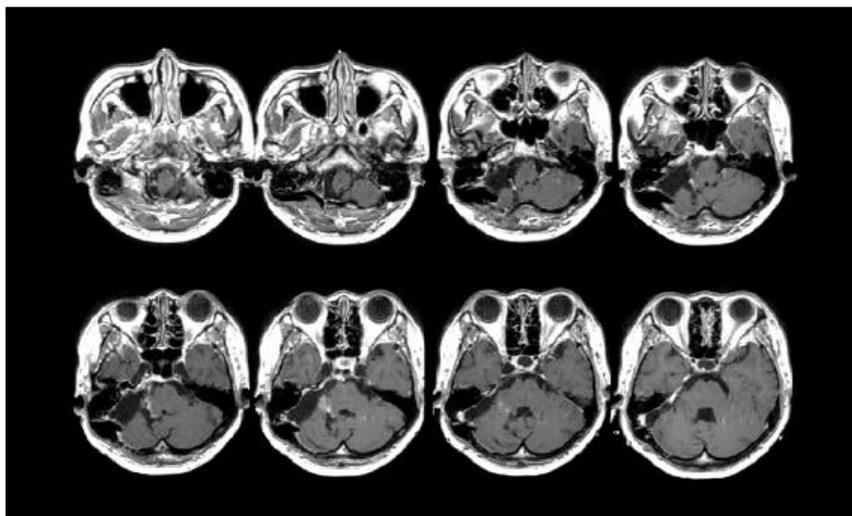


Fig. 6. Postoperative Gd enhanced MRI. Linear contrast enhancement was noted on the right middle cerebellar peduncle. Compared with the preoperative MRI, Gd enhanced images suggest that the mass was mostly an extra-axial one, protruding through the foramen Luschka and the cerebello-medullary fissure.

DISCUSSION

The simultaneous development of histologically different primary brain tumors, aside from phakomatoses or previous irradiation, is rare and its preoperative diagnosis is difficult, especially in contiguous tumors occurring concurrently. Phakomatoses are associated with an increased incidence of multiple tumors, and cranial irradiation for a single brain tumor is associated with a risk of developing secondary intracranial neoplasm.^{1,4} Multiple meningiomas and multiple ependymomas may occur in patients with neurofibromatosis or may occur sporadically. Bilateral acoustic neurofibromatosis, known as neurofibromatosis 2 (NF2), is a disease which predisposes the patient to the formation of multiple tumors in the central and peripheral nervous system.² Sznajder et al. suggested that host factors that increase susceptibility to radiation might be involved in the development of multiple neural tumors.¹⁰ The NF1 and TP53 genes may play a critical role in the progression of a neurofibroma or the development of multiple tumors.⁵ The Carney complex is another rare multisystem tumorous disorder that features myxomas, spotty skin pigmentation, endocrine tumors, and peripheral schwannomas.¹¹

Neither genetic factor nor prior irradiation history was found in our patient who had a rare combination of multiple primary tumors occurring concurrently. Primary manifestation of a

choroid plexus papilloma in the CPA near the opening of the fourth ventricle is rare,⁷⁻⁹ and its coexistence with a vestibular schwannoma has not been reported in the literature. It may be beyond the realm of possibility to theorize the development of such a rare case of concurrent multiple primary brain tumors. Therefore the present case may represent a chance occurrence of two concurrent tumors of different histology.

Although the clinical history of the current case suggested an acoustic neuroma, preoperative MRI findings were unusual for such a tumor. Although most of the tumor seemed to be an intra-axial tumor, the packing of the mass in the widened internal auditory canal together with heterogenous enhancement and multiple cystic changes on T2WI lead to a preoperative diagnosis of a huge acoustic neuroma. The difference in the characteristics of Gd enhancement of the two tumors was barely noticed until tumor exposure in the operating room. Considering the location and size of the mass, it may be unreal to consider the possibility of the coexistence of different tumors in this case by MRI study alone. It is worthwhile to note that postoperative MR images confirmed the mass to be mostly an extra-axial one, protruding through the foramen Luschka and the cerebello-medullary fissure. This case report is of note not only for the rarity that it represents, but also for its highlighting of the diagnostic and therapeutic challenges faced by neurosurgeons.

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