Coexistence of Cerebellopontine Angle Meningioma and Pituitary Microadenoma Presenting with Sudden Hearing Loss: Case Report

Ani İşitme Kaybıyla Ortaya Çıkan Serebellopontin Köşe Menenjiyomu ve Hipofizer Mikroadenom Birlikteliği

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ABSTRACT The clinic features of the cerebellopontine angle tumors and their management are well known, however there is limited knowledge regarding coexistence of other primary brain tumors which can be incidentally detected. A 50-year-old woman was referred with sudden hearing loss in her right ear of 85 dB. Gadolinium-enhanced MRI showed a mass originating from the internal acoustic meatus with an intracanalicular dural tail sign, which was interpreted as meningioma. Additionally, an incidental microadenoma was detected in the pituitary gland. The meningioma was treated using the CyberKnife robotic radiosurgery system. The tumor volume was reduced by 50% and an audiometric evaluation showed a gain of approximately 50 dB after 3 months of follow-up. To draw attention to multipl primary brain tumors and to familiarize clinicians with its clinical features and management, we report a case of cerebellopontine angle meningioma and pituitary adenoma that presented with sudden sensorineural hearing loss.

Key Words: Hearing loss, sudden; cerebellopontine angle; meningioma; pituitary neoplasms; radiosurgery

ÖZET Serebellopontin köşe tümörlerinin klinik özellikleri ve tedavi yaklaşımları iyi bilinmekle birlikte, rastlantısal olarak saptanan diğer primer beyin tümörleri ile ilgili sınırlı bilgi vardır. Elli yaşında kadın hasta sağ kulakta 85 dB ani işitme kaybı şikayeti ile kliniğimize başvurdu. Kontrastlı MRG'de internal akustik kanaldan kaynaklanan ve intrakanalikuler uzanım gösteren kitle menenjiyom olarak rapor edildi. Bunun yanısıra, rastlantısal olarak hipofiz bezinde mikroadenom farkedildi. Menenjiyom CyberKnife robotik radyocerrahi sistem ile tedavi edildi. Üç aylık takip sonrası tümör volümünün %50 azaldığı ve hastada yaklaşık 50 dB işitme kazancı elde edildiği gözlendi. Multipl primer beyin tümörlerinin klinik özellikleri ve tedavisine dikkat çekmek için ani işitme kaybıyla ortaya çıkan serebellopontin köşe menenjiyomu ve hipofizer adenom olgusu sunulmuştur.

Anahtar Kelimeler: İşitme kaybı, ani; serebellopontin köşe; menenjiyom; hipofiz neoplazileri; radyocerrahi

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Meningiomas, which arise from arachnoid cap cells, are the most common nonglial primary brain tumor, accounting for approximately 15% of all intracranial tumors.[1] These neoplasms occur more frequently in women than men, with a 2:1 ratio, and the incidence reveals increasing risk with age. Ninety percent of all meningiomas are located supratentorially, including the cerebral convexity, falx, parasagittal area, and sphenoid wing regions.[2] However, meningiomas may also be found in unusual locations, such as the cerebellopontine angle (CPA).

Simultaneously occurring multiple intracranial primary tumors are uncommon, and the coexistence of a CPA tumor and a pituitary adenoma is extremely rare. There are only four cases reported in the literature, but all of those were vestibular schwannomas with pituitary adenomas.[3-6]

Here, we report a new case of coexistent CPA meningioma and pituitary microadenoma to familiarize clinicians with its clinical features and management; the patient presented with sudden sensorineural hearing loss (SSNHL) as the first manifestation of the meningioma. To our knowledge, this is the first report of this unusual coexistence.

CASE REPORT

A 50-year-old female presented with sudden hearing loss and tinnitus in the right ear. She denied headache, facial numbness, vertigo, nausea, or vomitting. The patient did not report previous radiation therapy. Her physical examination was normal except for the tuning fork test; the Weber test lateralized to the left. There were no neurological or cutaneous signs. Pure-tone audiometry showed right sensorineural hearing loss, averaging 85 dB, and normal hearing on the left side (Figure 1). The patient was unresponsive to speech discrimination test on the right side. On the audi-

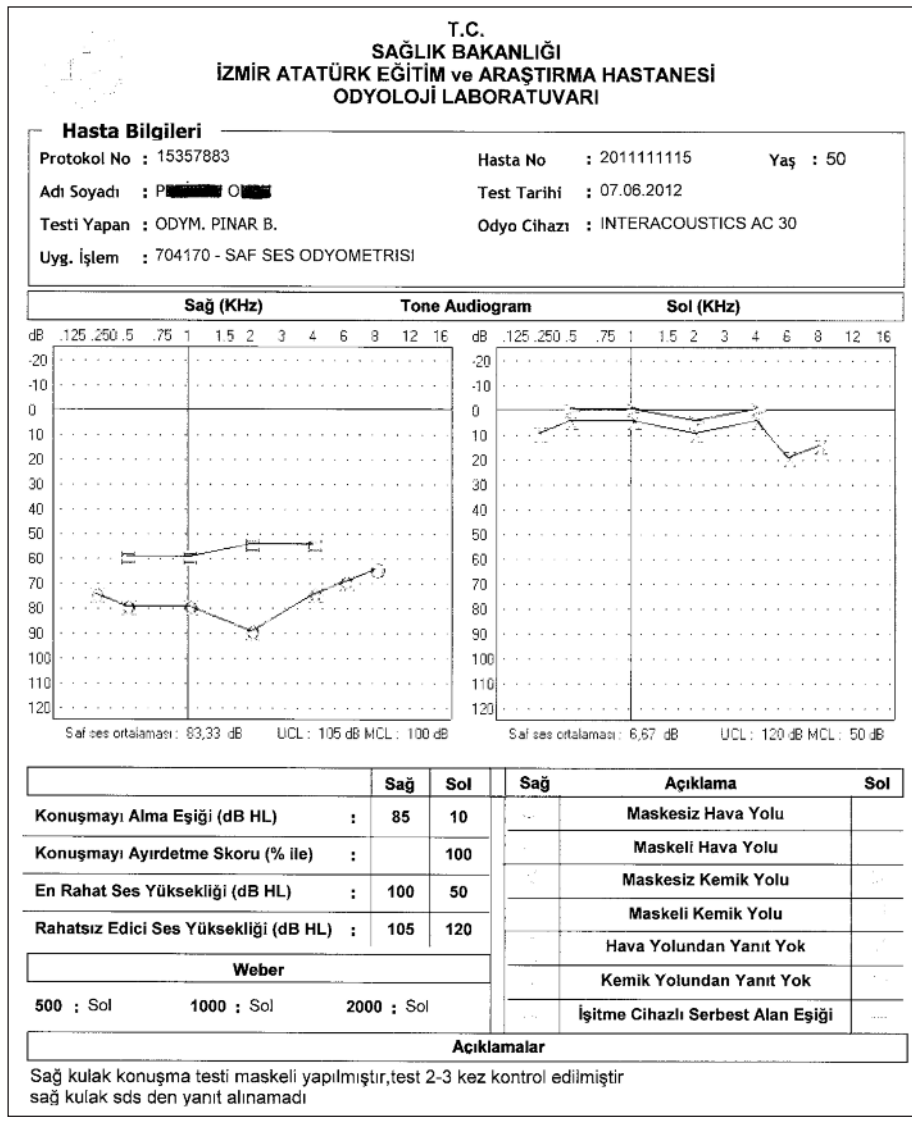


FIGURE 1: Pre-treatment audiometric evaluation: Pure-tone audiometry showed right sensorineural hearing loss, averaging 85 dB.

tory brainstem response test, the latency of wave V was delayed on the right side.

Prednisolone was administered initially at 1 mg/kg/day before the radiologic evaluation. However, the patient did not respond to this medical therapy. The patient was evaluated with gadolinium- enhanced magnetic resonance imaging (MRI) of the brain and the internal auditory canal with cranial and temporal bone protocols. MRI of the internal auditory canal revealed a lobulated, well-defined mass lesion of 17x11x16 mm with contrast enhancement on the right CPA. Additionally, it was shown that the mass originated from the internal acoustic meatus with an intracanalicular dural tail sign (Figure 2). Besides this, MRI showed an incidental mass lesion without contrast enhancement in the pituitary gland, which was 10x8x10 mm in size on T2-weighted images; this was interpreted as a microadenoma (Figure 3). Temporal bone computed tomography (CT) was performed to confirm the diagnosis of meningioma. Axial section temporal bone CT showed focal hyperostosis at the anterior wall of the right internal acoustic canal (Figure 4). After the radiological examination, an endocrinological workup revealed no hormonal discharge. The patient was referred to the Department of Radiation Oncology. The patient was treated with hypofractionated stereotactic radiation therapy. Therapy was given in three fractions over 6 days using the CyberKnife robotic radiosurgery system (Accuray, Sunnyvale, CA, USA). We prescribed 2100 cGy to the 91% isodose line and used different collimators and 240 beams. The conformality index was 1.33 for the planning target volume. The maximum doses were 244 cGy for the left eye and 2044 cGy for the brainstem, with mean doses to the left eye of 75 cGy and the brainstem of 254 cGy. The critical structure doses were within these limits. The patient tolerated the therapy well, with no acute adverse effects.

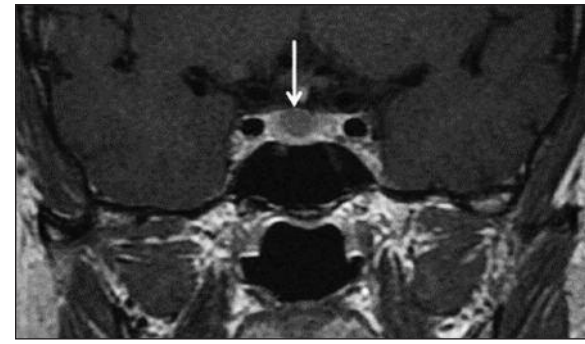


FIGURE 3: T2W MRI: Pituitary microadenoma is shown.

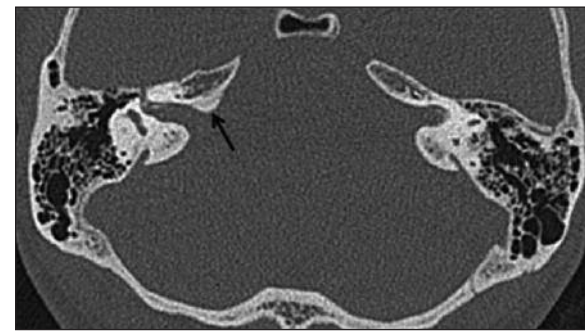


FIGURE 4: Temporal bone CT (axial section): Focal hyperostosis at the anterior wall of the right internal acoustic canal is shown.

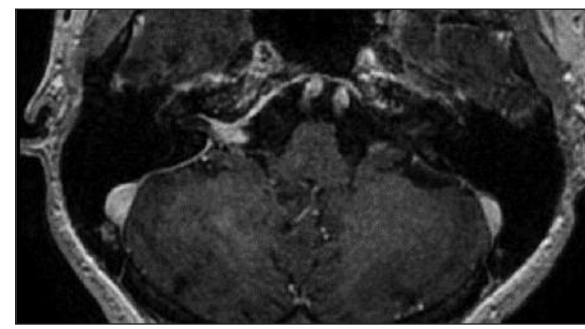


FIGURE 5: Post gadolinium MRI: Decrement of the size of the mass after CyberKnife therapy is shown.

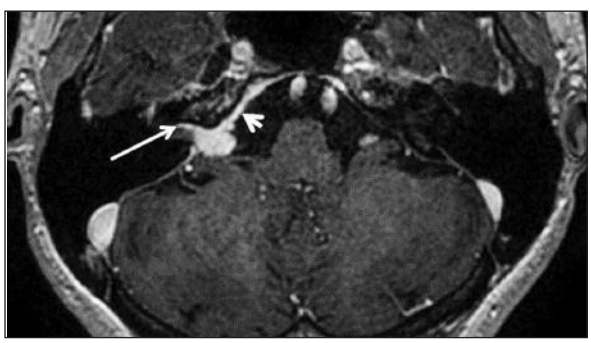


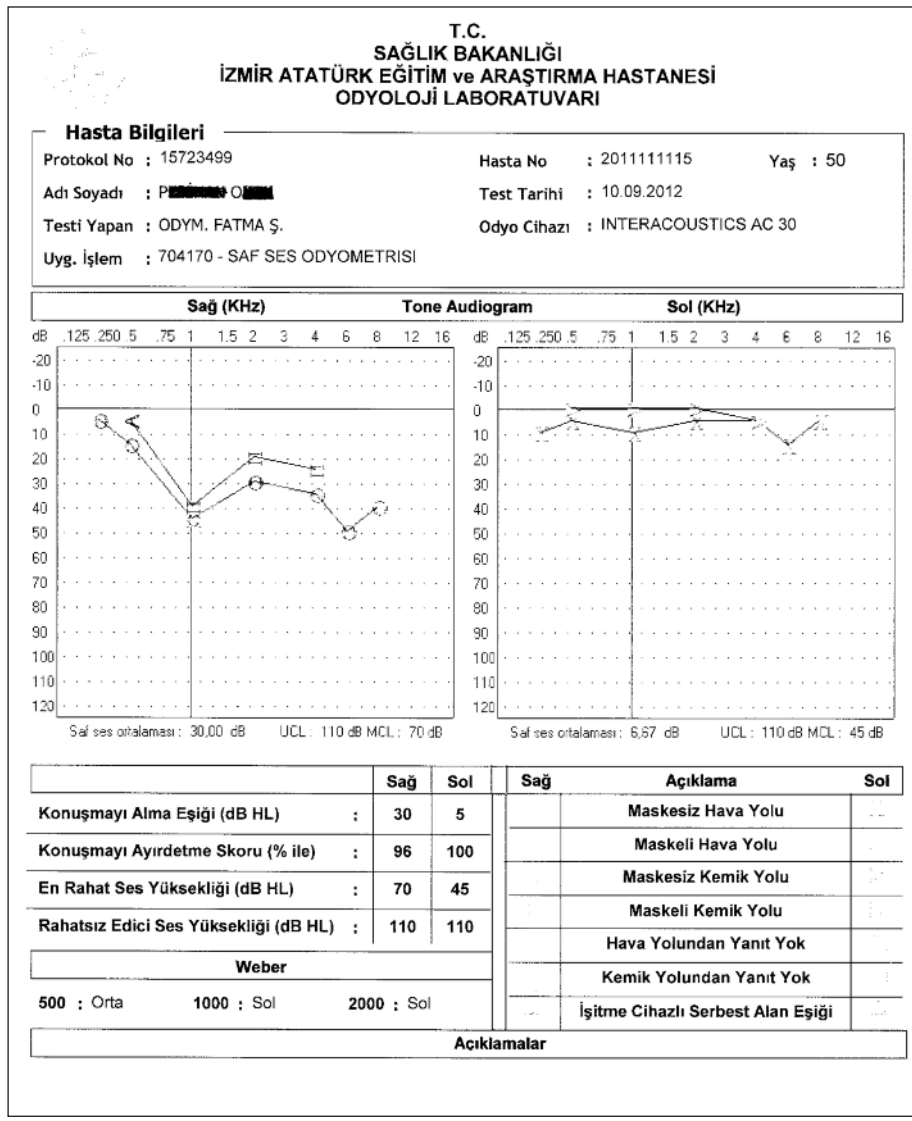
FIGURE 2: Post gadolinium MRI: The mass originated from the internal acoustic meatus (white arrow) with an intracanalicular dural tail sign (arrow head) is shown.

MRI performed 3 months after CyberKnife therapy and revealed that the size of the mass had decreased to 13x10x11 mm, and that the tumor volume was reduced by approximately 50% (Figure 5). An audiometric examination was also performed 3

months after the treatment and showed a gain of 50 dB with 96% speech discrimination score (Figure 6).

DISCUSSION

A CPA tumor should be suspected in patients with SSNHL who do not respond to medical therapy.[7] CPA tumors generally show slow growth patterns, and they typically present with slowly progressive asymmetric hearing loss. However, they may sometimes present with SSNHL. Gadolinium-enhanced MRI is sensitive for the diagnosis of retrocochlear pathologies. High-resolution CT scanning may be used if MRI cannot be performed (e.g., claustrophobia, metallic implants, and pacemakers), but, in fact, it does not show the fine details of a CPA as well as MRI. MRI is also effective in identifying other specific etiologies of SSNHL, including cerebral ischemia, multiple sclerosis, and inflamation. However, despite its utility, a significant percentage of MRI scans show incidental findings that are unrelated to SSNHL. Indeed, 54 patients with sudden deafness were investigated using brain MRI in a previous study. Thirty-one cases (57%) had MRI abnormalities, but only in six cases (11%) were the abnormalities directly related to SSNHL.[8] In this case, a pituitary adenoma, which


T.C.
SAĞLIK BAKANLIĞI
İZMİR ATATÜRK EĞİTİM ve ARAŞTIRMA HASTANESİ
ODYOLOJİ LABORATUVARI

Hasta Bilgileri

Protokol No : 15723499 Hasta No : 2011111115 Yaş : 50
Adı Soyadı : P[redacted] O[redacted] Test Tarihi : 10.09.2012
Testi Yapan : ODYM. FATMA Ş. Odyo Cihazı : INTERACOUSTICS AC 30
Uyg. İşlem : 704170 - SAF SES ODYOMETRISI

Sağ (KHz) Tone Audiogram Sol (KHz)

Saf ses ortalaması : 30,00 dB UCL : 110 dB MCL : 70 dB
Saf ses ortalaması : 6,67 dB UCL : 110 dB MCL : 45 dB

	Sağ	Sol
Konuşmayı Alma Eşiği (dB HL) :	30	5
Konuşmayı Ayırdetme Skoru (% ile) :	96	100
En Rahat Ses Yüksekliği (dB HL) :	70	45
Rahatsız Edici Ses Yüksekliği (dB HL) :	110	110

Weber
500 : Orta 1000 : Sol 2000 : Sol

Sağ	Açıklama	Sol
	Maskesiz Hava Yolu	
	Maskeli Hava Yolu	
	Maskesiz Kemik Yolu	
	Maskeli Kemik Yolu	
	Hava Yolundan Yanıt Yok	
	Kemik Yolundan Yanıt Yok	
	İşitme Cihazlı Serbest Alan Eşiği	

Açıklamalar

FIGURE 6: Post-treatment audiometric evaluation: Pure-tone audiometry showed approximately gain of 50 dB with 96% speech discrimination score.

was unrelated to the sudden hearing loss, was detected in addition to the CPA meningioma and the patient was investigated for adenoma.

Meningiomas represent only 3-12% of CPA tumors and must be distinguished from vestibular schwannomas, which are, by far, the most common tumors in this location.[9] Histopathological analysis is important for the differentiation of meningioma and vestibular schwannoma. Histopathological analysis requires a surgical approach, so it is not applicable to every patient. Although over 95% of meningiomas are benign (WHO grade I or II), only half of the diagnoses of benign meningiomas are confirmed histologically.[10] In this case, although the diagnosis of meningioma was not confirmed pathologically, the radiological findings showed no suspicion of meningioma.

The radiological distinction of meningiomas and schwannomas has been clearly defined. On MRI, meningiomas are often excentric to the internal auditory canal and have adjacent dural enhancement, referred to as a "dural tail," reflecting dural infiltration and/or increased vascularity. Vestibular schwannomas are more spherical than meningiomas, and usually involve the internal auditory canal or have an intracanalicular placement. CT demonstrates the chronic effects of meningioma on the adjacent skull very well, especially osseous destruction in malignant meningiomas or hyperostosis associated with benign meningiomas. Hyperostosis of the surrounding skull area is seen 70% of benign meningiomas, whereas it rarely occurs with vestibular schwannomas.[11] In our case, focal hyperostosis at the adjacent skull and intracanalicular dural tail sign were observed.

The treatment of meningiomas remains controversial because there is a lack of knowledge regarding the natural history of meningiomas and the growth potential of these tumors. They usually have a slow growth rate, and the clinical manifestations depend on the location of the tumor. Niiro et al. noted that 35% of their patients showed tumor growth but only 12% of patients became symptomatic during the 36-month follow-up period.[12] For this reason, a 'wait and scan' policy may be appropriate in selected patients. Nevertheless, many tumors grow rapidly with high proliferative activity and can become life-threatening. Moreover, Oya et al. reported that volumetric growth was seen in 74% of 154 intracranial meningiomas.[13]

The CyberKnife is an image-guided robotic radiosurgery system that has been used in the treatment of CPA tumors in recent years. Initial reports demonstrated high rates of tumor control and a favorable side-effect profile for small meningiomas. Peritumoral edema, hearing impairment, vertigo, and injury to the facial or trigeminal nerve are possible adverse effects that have been reported. In a group of 199 benign intracranial meningiomas, Colombo et al. reported 18% tumor reduction and 74% tumor stabilization.[14] Surgical resection is still the preferred option for larger tumors, cystic lesions, and meningiomas with brainstem compression. CyberKnife treatment was preferred due to the size and placement of the our patient's tumor.

Although the incidence of pituitary masses has increased in the last decade because of advances in diagnostic techniques, many of these tumors have not the capacity to synthesize hormones. Fewer than 50% of symptomatic pituitary tumors were microadenomas, and prolactinomas are the most common hormone-secreting tumors of the pituitary.[15] In a study of 2598 patients, 282 incidental pituitary masses were discovered and 118 of them were identified as non-functional pituitary adenomas.[16] Growth of nonfunctioning pituitary adenomas without treatment occurs in about 10% of microadenomas and 24% of macroadenomas.[17] The evaluation and treatment of small, asymptomatic pituitary incidentalomas may not be cost-effective.[18] We measured the hormone levels; they were within normal ranges, and we did not need to do any further investigation of a non-secreting pituitary adenoma.

The coexistence of multiple primary brain tumors with different histological characteristics is unusual. The simultaneous occurence of meningioma and pituitary adenoma in the absence of neurofibromatosis type 2 or a history of irradiation is very rare. To our knowledge, the coincidence of CPA meningioma and pituitary adenoma has not been reported previously.

The patients presented with SSNHL should be initially evaluated for CPA tumors using MRI. MRI can also reveal incidental findings unrelated to SSNHL. These findings may include the concurrence of multiple primary brain tumors. Additional studies are required to improve our understanding of the mechanisms leading to multiple primary brain tumor growth.

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