



# Metastatic Adenocarcinoma of Unknown Primary in the Bilateral Cerebellopontine Angles: Case Report and Review of the Literature

## *Bilateral Serebellopontin Açıda Primeri Bilinmeyen Metastatik Adenokarsinom: Olgu Sunumu ve Literatür Derlemesi*

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### ABSTRACT

Metastatic adenocarcinoma in bilateral cerebellopontine angles (CPA) is rare. We report a case and review the current literature in order to enhance recognition of metastatic adenocarcinoma in the cerebellopontine angle. A 44-year-old man was referred to the hospital with right-sided diminished hearing for 7 weeks, left-sided facial palsy for 2 weeks, and left-sided sensorineural hearing loss for 1 week. On Magnetic Resonance Imaging (MRI) two tumors in bilateral CPAs were detected. The left-sided tumor was resected and histopathological examination revealed an adenocarcinoma. Many investigations could not find the primary tumor. One should be careful with middle-aged or elderly patients with sudden progressive deficits in the VIII<sup>th</sup> or VII<sup>th</sup> cranial nerves, particularly in bilateral CPA.

**KEYWORDS:** Adenocarcinoma, Cerebellopontine angle, Metastasis

### ÖZ

Bilateral serebellopontin açıda (SPA) metastatik adenokarsinom nadirdir. Serebellopontin açıda metastatik adenokarsinom farkındalığını artırmak üzere bir olgu sunuyor ve mevcut literatürü gözden geçiriyoruz. 44 yaşında bir erkek hasta, 7 haftadır sağ tarafta işitme azalması, 2 haftadır sol tarafta fasiyal paralizi ve 1 haftadır sol tarafta sensörinöral işitme kaybı nedeniyle hastaneye sevk edildi. Manyetik Rezonans Görüntüleme (MRG) bilateral SPA'da iki tümör saptandı. Soldaki tümörün rezeksiyonu yapıldı ve histopatolojik inceleme bir adenokarsinom olduğunu gösterdi. Yapılan çok sayıda araştırmaya rağmen primer tümör bulunamadı. VII veya VIII. kranial sinirlerde aniden progresif defisit olduğunda ve özellikle bilateral SPA tutulması durumunda ve orta yaşlı veya yaşlı hastalarda dikkatli olunmalıdır.

**ANAHTAR SÖZCÜKLER:** Adenokarsinoma, Serebellopontin açısı, Metastaz

### INTRODUCTION

Primary tumors predominate in tumors of the CPA; metastasis is rare (0.2% of all lesions). The majority of the tumors in the CPA are vestibular schwannomas, followed by meningiomas, usually resulting in unilateral partial or complete sensorineural hearing loss. Metastasis of adenocarcinoma in bilateral CPAs without a known primary tumor is very rare (1). We report a case of metastatic adenocarcinoma in bilateral cerebellopontine angles.

### CASE REPORT

A 44-year-old man was admitted to our neurosurgical ward because of diminished hearing on the right side for 7 weeks, facial palsy on the left side for 2 weeks, and sensorineural hearing loss on the left side for 1 week. There was no headache, amblyopia, vertigo, tinnitus, nausea or vomiting.

Hearing tests demonstrated that the acoustic sensitivity on the left side was lost completely, with a pure-tone average of 45 dB HL in the right ear. The House-Brackmann scale (HB) for

the left facial paralysis was grade IV. There was no nystagmus or gait ataxia.

Contrast-enhanced MRI of the brain demonstrated the presence of two parenchymatous lesions with a diameter of 0.8 cm in the right CPA and 1.0 cm in the left (Figure 1). Bilateral internal auditory canals were dilated on computed tomography (CT) scan (Figure 2A, B). There was no obvious edema around the tumours. The left-sided tumor was resected completely. The tumor was found to be attached to the facial nerve, as well as to the acoustic nerve. Pathology evaluation revealed the definite diagnosis to be an adenocarcinoma (Figure 3A,B). CK7 and CDX-2 were positive, while CK20, PSA and TTF-1 remained negative.

Additional physical examination and investigations (including contrast-enhanced CT scan of the chest and abdomen, ultrasound examination of the urinary system and isotope bone scan) failed to find a primary tumor. The carcinoembryonic antigen (CEA, 0.94 ng/ml, normal range 0-3.4ng/ml),

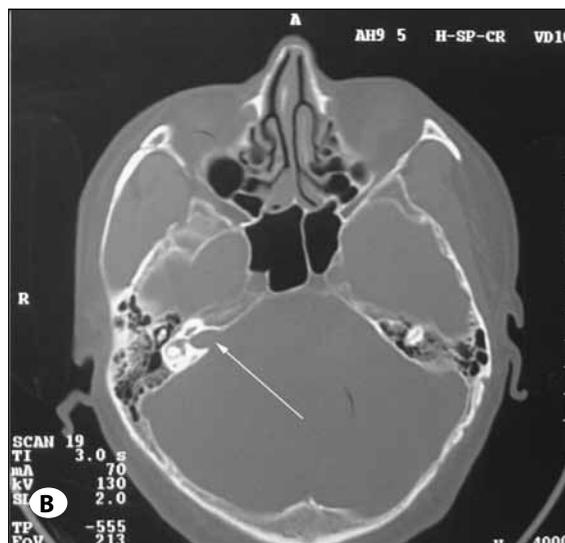
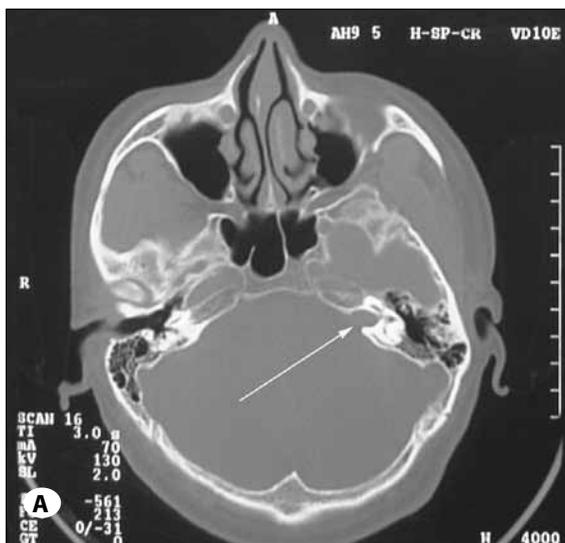


**Figure 1:** Axial Gadolinium-enhanced T1-weighted MRI-scans, showing two parenchymatous space occupying lesions with a diameter of 0.8 cm in the right CPA and 1.0 cm in the left.

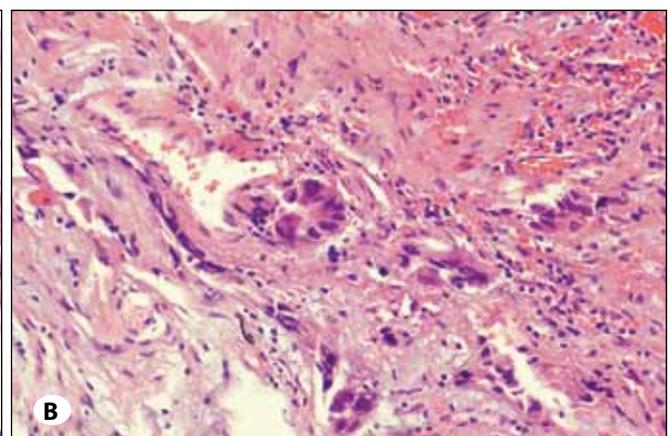
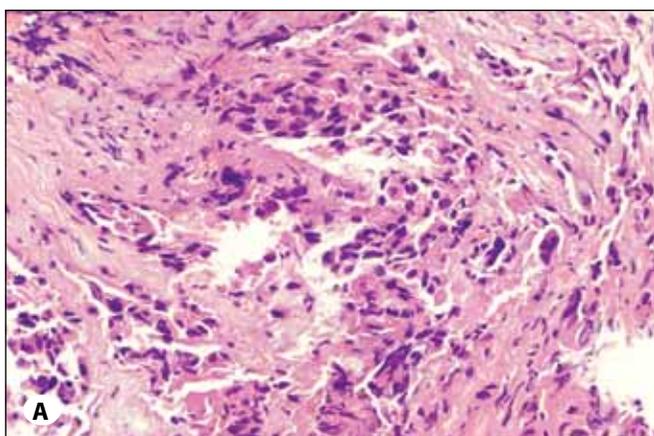
Total prostate-specific antigen (T-PSA, 0.584 ng/ml, normal range 0-3.0 ng/ml), squamous cell carcinoma antigen (SCCAg, 0.6 ng/ml, normal range 0-1.5 ng/ml), and Serum CA-125 (12.71 u/ml, normal range 0-35 u/ml) levels were normal. A Gamma Knife was used for the right tumor as the patient refused surgery for the second time. After 6 months, the patient was referred to our neurosurgical ward again for bilateral sensorineural hearing loss, left-sided facial palsy and gait ataxia. On contrast-enhanced MRI of the brain, two tumors in the midbrain were found as the tumor in the right CPA had not disappeared and there was a new tumor on the left side (Figure 4A,B). Additional examination did not reveal a primary tumor. The tumors in the midbrain were treated with stereotactic radiosurgery (Gamma knife). He died 2 months later.

### DISCUSSION

Most of the tumors in the CPA are benign and metastases are rare. Brackmann and Bartels reported only 3 metastases in a series of 1354 CPA lesions (1). To our knowledge, metastatic adenocarcinoma in the CPA has been reported only in 15



**Figure 2A,B:** Axial computed tomography (CT) showing that bilateral internal auditory canals were dilated.



**Figure 3A,B:** A minor part of the CPA lesion consists of polymorphous epithelial cells partly arranged in glandular structures with pleomorphic nuclei (H-E, x200).

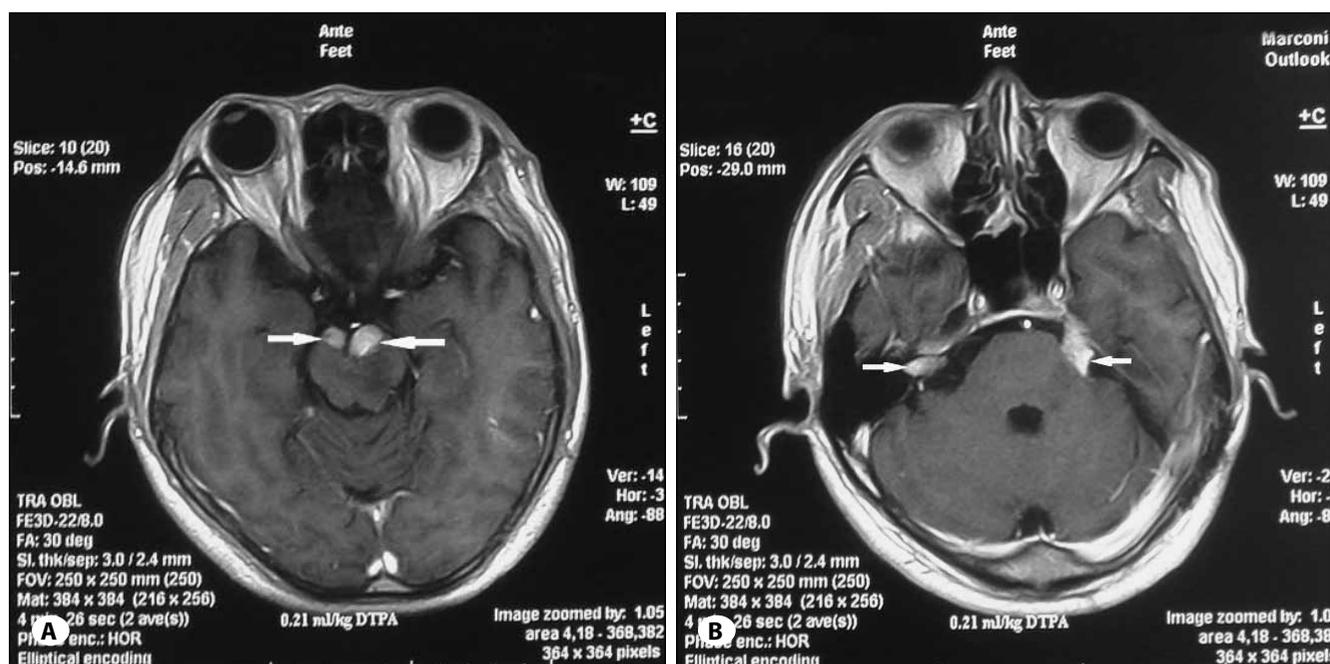
patients in English literature, and both cerebellopontine angles were involved in only 5 patients, including ours. The age of onset is from 36 years to 69 years (mean 57 years) (Table I). Benign tumors in the CPA usually result in unilateral partial or complete sensorineural hearing loss. The process of hearing loss is slow until the tumors become large.

Sudden sensorineural hearing loss may be a characteristic manifestation of metastatic adenocarcinomas in the CPA. Table I shows that 9 patients had rapidly progressive hearing loss. Primary severe involvement of the facial nerve is rare in benign tumors and appears especially in the late stage but it often occurs in metastatic adenocarcinomas. Of the

**Table I:** Summary of Metastatic Adenocarcinomas in the Cerebellopontine Angle (CPA) (8,3-16)

Patient #	Sex Age (y)	Primary cancer	Progressive facial palsy	Progressive hearing loss	Bilateral involvement of the VIIth or VIIIth cranial nerve	Cranial nerve disorder as the initial sign
1	M 68	Colon	+ (rapid)	+ (rapid)	+	-
2	M 58	Lung	+ (rapid)	+ (rapid)	-	+
3	F 57	Lung	+ (slow)	+ (slow)	-	+
4	F 36	Unknown	-	+ (rapid)	-	-
5	M 67	Lung	-	+ (rapid)	+	+
6	F 69	Parotid	+ (rapid)	+ (rapid)	-	-
7	M 44	Lung	+ (rapid)	+ (rapid)	-	-
8	M 53	Unknown	+ (slow)	+ (slow)	+	+
9	F 66	Breast	+ (slow)	+ (slow)	-	-
10	M 44	Unknown	+ (rapid)	+ (rapid)	+	+
11	M 67	Renal	-	+ (slow)	-	-
12	M 46	Gallbladder	+ (slow)	-	-	+
13	M 46	Ileum	+ (rapid)	+ (rapid)	-	+
14	F 64	Unknown	+ (rapid)	+ (rapid)	+	+
15	M 68	prostatic	+ (rapid)	-	-	+

"rapid" means that the symptoms developed quickly. "slow" means that the symptom developed slowly.



**Figure 4:** A) Axial Gadolinium-enhanced T1-weighted MRI scans, showing two parenchymatous space-occupying lesions in the midbrain. B) Axial Gadolinium-enhanced T1-weighted MRI scans, showing the tumor in the right cerebellopontine angle and a new tumor on the left side operated 6 months ago.

15 patients, 12 were found to have a facial palsy and 8 had experienced a rapidly progressive facial palsy. Cranial signs presented as an initial symptom in the course of illness in 9 patients. One should therefore be careful with patients suffering from rapidly progressive hearing loss, particularly with facial palsy, regardless of whether it is an initial symptom or not.

Large areas of edema may be associated with metastatic tumors (15). However, there was no obvious edema in the Magnetic Resonance Imaging (MRI) of our patient and the intracranial pressure was normal. The acutely diminished hearing and rapidly progressive facial palsy are thought to have been caused by direct tumor invasion and ischemia. Metastatic cancer should be suspected when there is a symptom of acutely diminished hearing and rapidly progressive facial palsy, even if there is no obvious edema surrounding the tumor. Metastatic adenocarcinomas in bilateral CPAs are very rare. Among the 5 cases, 4 had a history of progressive hearing loss and/or facial palsy and many investigations could not find the primary tumor. It is very important to discriminate metastatic and benign CPA lesions as further treatment is very different. Benign CPA tumors usually result in symptoms of VIIIth cranial nerve dysfunction such as unilateral partial or complete sensorineural hearing loss and tinnitus, and rarely other cranial nerve dysfunctions. In contrast, metastatic adenocarcinomas in the CPA mainly present with rapidly progressive hearing loss and particularly with facial palsy in middle-aged or elderly patients. One should be careful with middle-aged or elderly patients with sudden progressive deficits in the VIIIth or VIIth cranial nerves particularly in bilateral CPAs.

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