Case Report / Olgu Sunumu

Bilateral sudden sensorineural hearing loss caused by leptomeningeal carcinomatosis: case report and review

Leptomeningeal karsinomatozise bağlı iki taraflı ani sensörinöral işitme kaybı: Olgu sunumu ve derleme

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Leptomeningeal carcinomatosis is a rare condition characterized by diffuse infiltration of the meninges after the metastasis of the solid tumors. Bilateral sudden hearing loss is a rare initial symptom. In this article, we report a 44-year-old male patient with bilateral sudden hearing loss and dizziness. Magnetic resonance imaging showed involvement of the bilateral vestibulocochlear nerves. Malignant cells were detected in cerebrospinal fluid cytology. To the best of our knowledge, leptomeningeal carcinomatosis due to duodenum adenocarcinoma has not been reported before in the English literature. Leptomeningeal carcinomatosis should be kept in mind in patients who have bilateral sudden sensorineural hearing loss.
Keywords: Eighth cranial nerve; leptomeningeal carcinomatosis; sudden hearing loss.

Leptomeningeal karsinomatozis, solid tümörlerin metastazı sonucu meninksin diffüz infiltrasyonu ile karakterize, nadir görülen bir durumdur. İki taraflı ani işitme kaybı nadir bir başlangıç semptomudur. Bu yazıda iki taraflı ani işitme kaybı ve baş dönmesi olan 44 yaşında bir erkek hasta sunuldu. Manyetik rezonans görüntülemede iki taraflı vestibülokoklear sinir tutulumu görüldü. Beyin omurilik sıvısı sitolojisinde malign hücreler saptandı. Bildiğimiz kadarıyla, daha önce İngilizce literatürde duodenum adenokarsinomuna bağlı leptomeningeal karsinomatozis bildirilmemiştir. İki taraflı ani işitme kaybı olan hastalarda leptomeningeal karsinomatozis göz önünde bulundurulmalıdır.
Anahtar Sözcükler: Sekizinci kraniyal sinir; leptomeningeal karsinomatosis; ani işitme kaybı.

Leptomeningeal carcinomatosis (LMC) is characterized by diffuse infiltration of the leptomeninges and subarachnoid space by malignant cells and frequently seen in lung cancer, breast cancer and malignant melanoma.[1] A variety of neurologic symptoms can be seen due to involvement of the cerebrum, cerebellum, spinal cord and cranial nerves. Saenger in 1900 was the first to describe hearing loss due to LMC.[2,3] Since then many case reports have been published but bilateral sudden sensorineural hearing loss (SSHL) as an initial symptom has been very rarely

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reported. We present an unusual case of SSHL caused by infiltration of the leptomeninges with tumor cells from a duodenal adenocarcinoma. To the best of our knowledge, the duodenum as a source of leptomeningeal carcinomatosis is reported for the first time.

CASE REPORT

A 44-year-old man developed sudden bilateral hearing loss, dizziness and diplopia six months after a Whipple operation for signet ring cell adenocarcinoma of the duodenum. Two weeks prior to consult, he had right-sided tinnitus and hearing loss which subsequently affected the left side. After a while, dizziness and nausea commenced. Otorhinolaryngeal and neck physical examinations showed no abnormality. On neurologic examination, the Romberg's test was positive, cerebellar tests were normal, nystagmus was negative, and there was no abnormal finding related to the motor and sensorial cortex. The pupils were isochoric, light reflexes were +/+, and eye movements were normal.

Audiometry revealed a 54 dB hearing loss on the right and 116 dB hearing loss on the left, and the patient was hospitalized. Caloric tests and video electronystagmography revealed 60% hypoactivation on the left. Type A tympanograms were found in both ears. All laboratory findings including autoimmune markers, viral and syphilis tests were normal. According to our protocol therapy for SSHL, he underwent a systemic administration of steroids, plasma expanders and intratympanic corticosteroid treatment applied for one dose per day in both ears for a week. During treatment, audiometric tests were performed daily. At the end of the seventh day, hearing levels in the right decreased by 15 dB, whereas those on the left increased by 43 dB. The balance disorder had decreased and the patient started mobilizing. Gadolinium-enhanced magnetic resonance imaging (MRI) showed bilateral vestibulocochlear nerve enhancement on post contrast T_1 weighted images, reported as a finding of LMC. Lumbar puncture showed elevated protein levels (504.2 mg/dL), normal values of lactate dehydrogenase (32 IU/L) and glucose levels (74 mg/dL). Cerebrospinal fluid (CSF) cytology reported pleomorphic malignant cells and atypical lymphoid cells. There were elevated CA 19-9 levels (50.4 U/mL) but the carcinoembryonic antigen (CEA) value was within normal range (1.78 ng/mL). The patient was diagnosed as LMC with these laboratory and MRI findings, and transferred to the Radiation Oncology Department for cranial radiotherapy. During the radiotherapy treatment, the patient had left sided trochlear nerve paralysis and right-sided oculomotor nerve paralysis, and unfortunately died three months after hospitalization.

DISCUSSION

There are many etiologies of SSHL and despite state-of-the-art technology in medicine, diagnosis remains unclear in most cases. The incidence is 5-20 to 100,000 per year.[4] The incidence of bilateral SSHL is low compared to unilateral SSHL, estimated between 0.44% and 4.9% of all patients with SSHL.[5] Viral infections, meningitis,

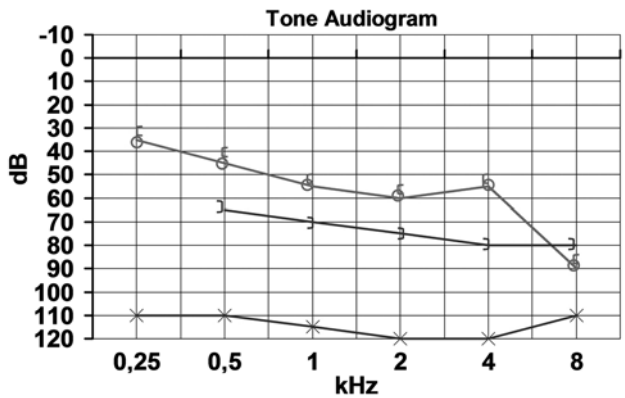

Figure 1. First pure tone audiogram shows hearing loss of 54 dB on the right and 116 dB on the left (before the treatment).

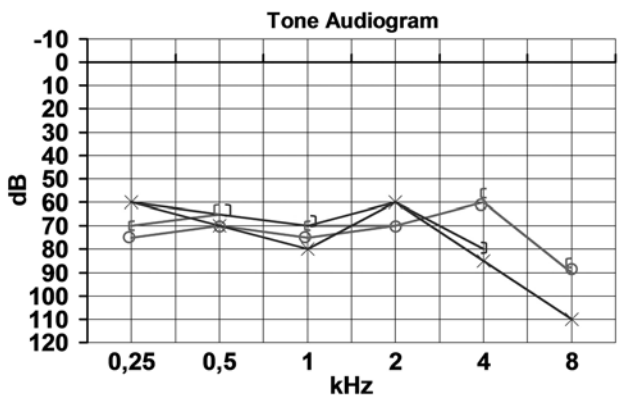

Figure 2. The seventh day pure tone audiogram shows hearing loss of 69 dB on the right and 73 dB on the left (after the treatment).

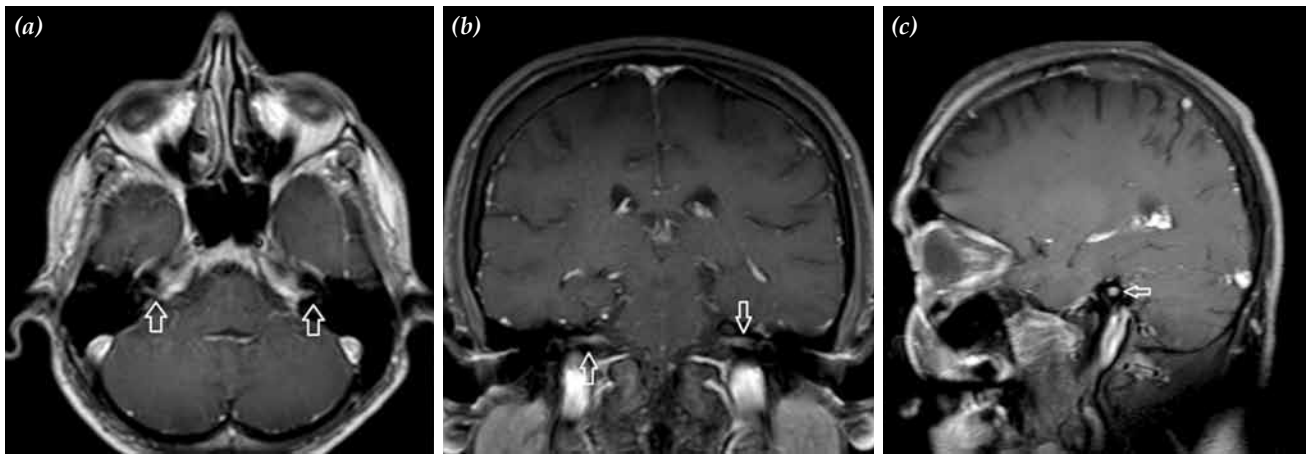

Figure 3. *Gadolinium-enhanced T_1-weighted magnetic resonance imaging shows the thickening and enhancement of both vestibulocochlear nerves.* ***(a)*** *Axial,* ***(b)*** *coronal, and* ***(c)*** *sagittal images.*

cardiovascular diseases, tumor metastasis to the internal acoustic meatus are the main reasons of the SSHL.[6] On the other hand Hashimoto Disease, Cogan's Syndrome, Wegener's Granulomatosis, Multiple Sclerosis and Polychondritis were found in 27% of the patients.[6]

One of the initial symptoms of LMC is bilateral SSHL.[7,8] To the best of our knowledge 22 case reports have been published up to the year 2000, and all case reports after the year 2000 are presented in Table 1. There is no LMC case that we know of that has a duodenum adenocarcinoma as source and has bilateral SSHL as an initial symptom.

Leptomeningeal carcinomatosis is seen in 5% of cancer patients and frequently seen at lung cancer, breast cancer and malignant melanoma.[9] Leptomeningeal carcinomatosis can present with a variety of neurologic symptoms and multiple cranial nerve involvement is common. Third, fifth and seventh cranial nerves are the most affected ones.[10]

Albert and Terence[11] argued that LMC must be considered in patients who have initial bilateral SSHL and they also claimed that 10% of the LMC patients have hearing loss because of eighth nerve involvement. In the same study, it was also shown that during the LMC, tinnitus, one-sided hearing

Table 1. Patients of meningeal carcinomatosis presenting with hearing loss

	Age/sex	Symptoms	Primary tumor
Uppal ve ark.[7]	61/F	Bilateral hearing loss, bilateral facial palsies	Breast adenocarcinoma
Currie ve Tomma[23]	49/F	Bilateral hearing loss, tinnitus	Malignant melanoma
Boukriche ve ark.[24]	59/M	Bilateral hearing loss	Bladder transitional cell carcinoma
Jeffs ve ark.[19]	66/F	Bilateral hearing loss	Metastatic melanoma
Wagemakers ve ark.[18]	52/M	Bilateral hearing loss, tinnitus	Esophagus adenocarcinoma
Jariengprasert ve ark.[16]	64/F	Bilateral hearing loss, vertigo	Unknown/adenocarcinoma
Suzuki ve ark.[15]	60/F	Bilateral hearing loss, left facial palsy	Rectum adenocarcinoma
Lai ve ark.[3]	66/M	Bilateral hearing loss, right facial palsy, vertigo, tinnitus	Lung adenocarcinoma
Baba ve ark.[22]	59/M	Right sided hearing loss, right facial palsy, headache	Stomach adenocarcinoma
Koda ve ark.[17]	63/M	Bilateral hearing loss	Esophagus squamous cell carcinoma
Vitaliani ve ark.[21]	59/F	Right sided hearing loss, vertigo, tinnitus	Over carcinoma
Gu ve ark.[20]	59/M	Left sided hearing loss, vertigo	Lung adenocarcinoma
Mourgela ve ark.[25]	56/M	Bilateral hearing loss, left facial palsy	Lung small cell cancer
Marchese ve ark.[2]			
Case 1	56/M	Right sided hearing loss, vertigo, tinnitus	Pancreas adenocarcinoma
Case 2	64/F	Bilateral hearing loss, vertigo, tinnitus	Unknown/differential carcinoma
Ohno ve ark.[12]	62/M	Bilateral hearing loss, dizziness, headache	Stomach adenocarcinoma
Present study (2011)	44/M	Bilateral hearing loss, vertigo	Duodenum adenocarcinoma

loss and rapid progression to the other side subsequently results in bilateral hearing loss and related facial paralysis is common.

The prognosis in LMC is poor and the course in untreated patients is approximately 4-6 weeks. In some cases survival can be prolonged to 4-6 months after aggressive treatment.[12] Because of the poor prognosis, early diagnosis is very important. The existence of malignant cells in CSF cytology is important for diagnosis, but there may be no malignant cells on first examination, and a lumbar puncture could be repeated. Olson et al.[1] required two or more lumbar punctures in over 90% of his 50 LMC patients, and despite the repetition, only 37 of the 50 patients had positive cytology findings. In our case, positive cytology was reported on first attempt. Other findings of the CSF were increased pressure, elevated protein levels, minimal cellular pleocytosis, predominant lymphocytes, and hypoglycorrhachia.[11] These findings are nonspecific and can be seen in tuberculosis, sarcoidosis and fungal infections.[13]

Myelography, computed tomography, and MRI can be helpful in the diagnosis of LMC. Gadolinium-enhanced MRI is the most sensitive imaging modality for LMC, and enhancement of the leptomeninges, cranial nerves and intraventricular nodules are important for diagnosis. Also MRI has advantages in showing communicating hydrocephaly over other imaging modalities.[3] Despite these advantages of MRI, it must always be kept in mind that findings can be normal at the initial stage. In our case, gadolinium-enhanced MRI showed the thickened and enhanced vestibulocochlear nerves.

Treatment options include intrathecal chemotherapy and whole-brain radiotherapy.[14] Steroids are often employed to reduce edema.[7] Our search of the literature yielded no treated case of hearing loss. In our department the treatment procedure for SSHL includes systemic steroids, plasma expanders and intratympanic corticosteroids. In our case, hearing on the left side improved considerably after treatment whereas it worsened slightly on the right.

Leptomeningeal carcinomatosis should be kept in mind in patients who have bilateral SSHL. The diagnosis of this rare entity can be confusing because of negative cytopathologic findings on first lumbar puncture. With this report, duodenal adenocarcinoma emerges as another source of leptomeningeal carcinomatosis patients with concomitant bilateral sensorineural hearing loss.

Declaration of conflicting interests

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REFERENCES

1. Olson ME, Chernik NL, Posner JB. Infiltration of the leptomeninges by systemic cancer: a clinical and pathologic study. Arch Neurol 1974;30:122-37.
2. Marchese MR, La Greca C, Conti G, Paludetti G. Sudden onset sensorineural hearing loss caused by meningeal carcinomatosis secondary to occult malignancy: report of two cases. Auris Nasus Larynx 2010;37:515-8.
3. Lai TH, Chen C, Yen DJ, Yu HY, Yiu CH, Kwan SY. Isolated acute hearing loss as the presenting symptom of leptomeningeal carcinomatosis. J Chin Med Assoc 2006;69:496-8.
4. Byl FJ. Sudden hearing loss: eight years experience and suggested prognostic table. Laryngoscope 1984;94:647-61.
5. Oh JH, Park K, Lee SJ, Shin YR, Choung YH. Bilateral versus unilateral sudden sensorineural hearing loss. Otolaryngol Head Neck Surg 2007;136:87-91.
6. Xenellis J, Nikolopoulos TP, Stavroulaki P, Marangoudakis P, Androulakis M, Tsangaroulakis M, et al. Simultaneous and sequential bilateral sudden sensorineural hearing loss: are they different from unilateral sudden sensorineural hearing loss? ORL J Otorhinolaryngol Relat Spec 2007;69:306-10.
7. Uppal HS, Ayshford CA, Wilson F. Sudden onset bilateral sensorineural hearing loss: A manifestation of occult breast carcinoma. J Laryngol Otol 2001;115:907-11.
8. Imamura S, Nozawa I, Imamura M, Murakami Y. Clinicopathologic study of leptomeningeal carcinomatosis involving the temporal bone. Ann Otol Rhinol Laryngol 1997;106:674-9.
9. Civantos F, Choi YS, Applebaum EL. Meningeal carcinomatosis producing bilateral sudden hearing loss: a case report. Am J Otol 1992;13:369-371.
10. Morgan MK, Zammit-Maempel I, Hill J. Meningeal carcinomatosis: an unusual cause of deafness. J R Coll Surg Edimb 1998;43:119-21.
11. Alberts MC, Terrence CF. Hearing loss in carcinomatous meningitis. J Laryngol Otol 1978;92:233-41.
12. Ohno T, Yokoyama Y, Aihara R, Mochiki E, Asao T, Kuwano H. Sudden bilateral sensorineural hearing loss as the presenting symptom of meningeal carcinomatosis of gastric cancer: report of a case. Surg Today 2010;40:561-5.
13. Morganroth J, Deisseroth A, Winokur S, Schein P. Differentiation of carcinomatous and bacterial meningitis. Neurology 1972;22:1240-2.

14. Giannone L, Greco FA, Hainsworth JD. Combination intraventricular chemotherapy for meningeal neoplasia. J Clin Oncol 1986;4:68-73.
15. Suzuki T, Sakaguchi H, Yamamoto S, Hisa Y. Sudden hearing loss due to meningeal carcinomatosis from rectal carcinoma. Auris Nasus Larynx 2006;33:315-9.
16. Jariengprasert C, Laothamatas J, Janwityanujit T, Phudhichareonrat S. Bilateral sudden sensorineural hearing loss as a presentation of metastatic adenocarcinoma of unknown primary mimicking cerebellopontine angle tumor on the magnetic resonance image. Am J Otolaryngol 2006;27:143-5.
17. Koda H, Kimura Y, Iino Y, Eishi Y, Murakami Y, Kitamura K. Bilateral sudden deafness caused by diffuse metastatic leptomeningeal carcinomatosis. Otol Neurotol 2008;29:727-9.
18. Wagemakers M, Verhagen W, Borne B, Venderink D, Wauters C, Strobbe L. Bilateral profound hearing loss due to meningeal carcinomatosis. J Clin Neurosci 2005;12:315-8.
19. Jeffs GJ, Lee GY, Wong GT. Leptomeningeal carcinomatosis: an unusual cause of sudden onset bilateral sensorineural hearing loss. J Clin Neurosci 2006;13:116-8.
20. Gu CS, Liu CY, Wang MC. Brain metastasis of non-small cell lung cancer presenting as sensorineural hearing loss and vertigo. J Chin Med Assoc 2009;72:382-4.
21. Vitaliani R, Spinazzi M, Del Mistro AR, Manara R, Tavolato B, Bonifati DM. Subacute onset of deafness and vertigo in a patient with leptomeningeal metastasis from ovarian cancer. Neurol Sci 2009;30:65-7.
22. Baba S, Matsuda H, Gotoh M, Shimada K, Yokoyama Y, Sakanushi A. A case of meningeal carcinomatosis presenting with the primary symptoms of facial palsy and sensorineural deafness. J Nihon Med Sch 2006;73:240-3.
23. Currie L, Tomma A. Malignant melanoma presenting as sudden onset of complete hearing loss. Ann Plast Surg 2001;47:336-7.
24. Boukriche Y, Bouccara D, Cyna-Gorse F, Dehais C, Felce-Dachez M, Masson C. Sudden bilateral hearing loss disclosing meningeal carcinomatosis. Rev Neurol 2002;158:728-30.
25. Mourgela S, Sakellaropoulos A, Ardavanis A. Leptomeningeal carcinomatosis presenting as bilateral sensorineural deafness and unilateral facial palsy. J BUON 2009;14:317-9.