RAPID ONSET SENSORINEURAL HEARING LOSS SECONDARY TO BILATERAL CEREBRAL MELANOMA METASTASES TO THE CEREBELLOPONTINE ANGLE

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Abstract

Background: We describe a case of bilateral cerebellopontine angle melanoma metastases in the context of a literature review. The case is a rare presentation of sensorineural hearing loss and an unusual site of metastasis for malignant melanoma.

Methods and results: A 54-year-old man presented with sudden deafness. Bilateral melanoma metastases were confirmed with MRI and lumbar puncture. He died 3 months after treatment.

Conclusions: Melanoma metastasis should be considered as a differential for sensorineural hearing loss. A review of the literature found that the median survival of patients with bilateral cerebellopontine angle metastatic melanoma is 6 months post-treatment. At surgery these tumours are largely unresectable from cranial nerves VII and VIII. Surgical intervention is unlikely to result in survival or symptomatic benefit and the focus of treatment should be on maintaining quality of life. Hearing rehabilitation and gamma knife surgery may have a role.

Keywords: hearing • melanoma • cerebellopontine

LA SORDERA SÚBITA A CONSECUENCIA DE METÁSTASIS BILATERAL DEL MELANOMA EN EL ÁNGULO PONTO-CEREBELOSO

Resumen

Introducción: Se describe el caso de la metástasis bilateral del melanoma en el ángulo ponto-cerebeloso. Este punto puede ser una fuente de la pérdida auditiva de tipo de recepción y un punto raro de la presencia de la metástasis del melanoma maligno.

Métodos y resultados: Estudio del caso de un hombre de 54 años de edad, que había experimentado la sordera súbita. En base a la prueba MRI (de resonancia magnética) y de la punción lumbar, se ha encontrado la metástasis bilateral del melanoma. El paciente falleció a los tres meses después de la finalización del tratamiento médico.

Resultados: La metástasis del melanoma se debe tratar como una causa más de la pérdida auditiva de tipo de recepción. Una revisión de la literatura especializada muestra que el tiempo medio de supervivencia de los pacientes con metástasis bilateral del melanoma en el ángulo ponto-cerebeloso es de 6 meses después del tratamiento médico. Estos tumores son en gran parte no operacionales e imposibles de separar a través de la cirugía de los nervios craneales del grupo VII y VIII. La intervención cirúrgica no aumenta la probabilidad de supervivencia o beneficio sintomático. El tratamiento debe centrarse en mantener la calidad de vida. La rehabilitación de la audición y la radiocirugía pueden traer resultados positivos.

Palabras clave: audición • melanoma • ángulo ponto-cerebeloso
Hearing loss of rapid onset is not an uncommon otolaryngological presentation, occurring at an annual incidence of around 5–20 per 100,000 population [1]. Over two-thirds is idiopathic in origin, other major causes including infection, vascular events, and trauma [2]. Only about 2% of cases are of neoplastic origin, the majority of these comprising vestibular schwannomas [2]. We describe a rare case of bilateral cerebellopontine angle melanoma causing bilateral sensorineural hearing loss, and present a review of the literature regarding survival and potential treatment.

A 54-year-old man presented to our clinic with a 6-week history of progressive bilateral hearing loss and a feeling of imbalance. On otoscopy, both tympanic membranes and middle ears looked normal. His facial nerve function was unaffected. His pure tone audiogram demonstrated bilateral profound sensorineural hearing loss (Figure 1), with significant progression from the moderate hearing loss described at referral only 2 weeks previously. No baseline pure tone audiogram was available on record for this patient, as hearing had not been a significant symptom previously to cause him to seek medical attention.
He had a past medical history of a 1.3 mm Clark Level 4 melanoma, which had been excised from behind his left ear some 12 years ago, and a radical neck dissection secondary to a recurrence of the melanoma on his left neck 6 years earlier. A chest CT at this time demonstrated multiple pulmonary nodules, which were resected via a median sternotomy. A right parietal metastasis diagnosed 4 years before presentation had been treated with stereotactic radiotherapy. This had shown no progression on repeat MRI scans, the latest of these 3 months before recent symptoms began.

Three days before his appointment was due, he developed severe headaches. An urgent CT head with contrast displayed enhancement within both internal auditory meati (IAM) was done. An MRI head was arranged, which confirmed bilateral gadolinium enhancing lesions within the IAM, extending to the cerebellopontine angles (CPA) (Figure 2). Atypical cells found in CSF from a lumbar puncture confirmed the diagnosis of malignant meningitis secondary to metastatic melanoma.

The patient began a course of oral steroids and he was referred for behind-the-ear hearing aids. However, 1 month later he reported complete deafness, and progression of pure tone audiograms was recorded over this time (Figure 1). No further audiology was undertaken as diagnosis of pathology was confirmed by MRI scanning. At diagnosis, he was referred urgently to medical oncology for further treatment. He underwent gamma knife surgery (GKS) at another hospital. Two days after this treatment, he was admitted to hospital acutely confused. Repeat MRI demonstrated response of both IAM lesions to GKS, but also displayed enhancement of cerebellar folia, consistent with meningeal spread of metastases (Figure 3). He was discharged home after recovering spontaneously from this episode of confusion, but continued to deteriorate, and died of metastatic melanoma 3 months later, and 5 months after initial referral.

**Discussion**

We describe both a rare cause of bilateral sensorineural hearing loss and an unusual presentation of metastatic melanoma. The majority of neoplasms at the CPA are vestibular schwannomas [3]. Metastatic tumours found at the CPA constitute only 0.2% of lesions found at this site and
these most commonly originate from breast, lung, kidney, stomach, and larynx [3]. A systematic literature review (Medline database search terms used were ‘cerebello-pontine angle’, ‘melanoma’, ‘internal auditory meatus’, and ‘bilateral’) identified nine papers which reported 11 cases of bilateral cerebellopontine angle metastatic melanoma [4–12]. Details of these cases are summarised in Table 1.

Recent data suggests incidence of malignant melanoma within the UK continues to increase and is now at around 17 cases per 100,000 population per year [13]. Melanoma is a common cause of intracranial metastasis and 50% of patients with metastatic melanoma have cerebral involvement [14]. Despite this, melanoma rarely presents at the CPA. It has been suggested that metastasis to this site is haematological; at autopsy, metastatic melanoma has been found around blood vessels of the temporal bone [15]. From the temporal bone, metastatic melanoma is thought to track along cranial nerves VII and VIII via their neural sheaths or vasa nervosum [15]. This case also describes the progression of melanoma metastases to the posterior cranial fossa from the IAM, suggestive of leptomeningeal spread from this site. It is still unclear why the majority of such melanomas present bilaterally.

Vestibular schwannomas present as bilateral disease in 5% of cases; this occurs mainly in patients with neurofibromatosis type II [10]. Accordingly, there have been cases of misdiagnosis of bilateral cerebellopontine angle melanoma as both vestibular schwannoma [4] and neurofibromatosis type II [5]. Furthermore, diagnosis of metastasis to the CPA is not immediately obvious due to a latent

Table 1. Summary of previously published case reports of bilateral acoustic melanomas. All cases developed further intracranial metastasis following initial diagnosis

<table>
<thead>
<tr>
<th>First author, date of publication</th>
<th>Extra-cranial metastasis</th>
<th>Development of additional intracranial metastasis</th>
<th>Treatment modality</th>
<th>Survival post-treatment (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Delerue, 1991 [7]</td>
<td>No</td>
<td>n/d</td>
<td>Surgical</td>
<td>4</td>
</tr>
<tr>
<td>Tu, 1994 [8]</td>
<td>Yes</td>
<td>Yes</td>
<td>n/d</td>
<td>n/d</td>
</tr>
<tr>
<td>Arriaga, 1995 [9]</td>
<td>No</td>
<td>n/d</td>
<td>Surgical</td>
<td>5</td>
</tr>
<tr>
<td>Shinogami, 1998 [10]</td>
<td>No</td>
<td>Yes</td>
<td>Surgical, radiotherapy</td>
<td>12</td>
</tr>
<tr>
<td>Jacob, 2007 [11]</td>
<td>No</td>
<td>Yes</td>
<td>Surgical</td>
<td>0</td>
</tr>
<tr>
<td>Brackmann, 2007 [12]</td>
<td>Yes</td>
<td>Yes</td>
<td>Radiotherapy</td>
<td>9</td>
</tr>
<tr>
<td>Brackmann, 2007 [12]</td>
<td>Yes</td>
<td>Yes</td>
<td>GKS, Chemotherapy</td>
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</tr>
<tr>
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<td>n/d</td>
<td>n/d</td>
<td>n/d</td>
</tr>
<tr>
<td>Gerganov, 2008 [4]</td>
<td>Yes</td>
<td>n/d</td>
<td>Surgical</td>
<td>n/d</td>
</tr>
</tbody>
</table>

n/d – not described within paper


