Neurol Sci (2000) 21:329-331 © Springer-Verlag 2000

CASE REPORT

R.M. Ruggieri • L. Manfrè • F. Calbucci • F. Piccoli

Therapeutic considerations in cerebellopontine angle lipomas inducing hemifacial spasm

Received: 20 January 2000 / Accepted in revised form: 24 November 2000

Abstract Lipoma is a very rare tumour at the cerebellopontine angle. We report a case of incomplete hemifacial spasm, associated with a lipoma involving and compressing both facial and acoustic nerves at their origin in the brainstem. The patient was treated with medical therapy (botulinum toxin A) and surgery. We present a review of the last ten years of the literature, with particular regard to management.

Key words Cerebellopontine syndrome • Lipoma • Hemifacial spasm • Botulinum toxin • Facial nerve lesion • Acoustic nerve lesion

R.M. Ruggieri (✉) • F. Piccoli
Institute of Neuropsychiatry
University of Palermo
Via G. La Loggia 1, I-90129 Palermo, Italy

L. Manfrè
Institute of Radiology P. Cignolini
University of Palermo, Palermo, Italy

F. Calbucci
Neurosurgery Department
Bellaria Hospital, Bologna, Italy

Introduction

Hemifacial spasm (HFS) is caused by a neurovascular conflict in more than two-thirds of cases [1-3]. Only in 1% of adult patients [4] it is the consequence of a tumour located in the cerebellopontine angle (CPA), among which lipoma is very rare.

We describe a patient suffering from an incomplete HFS, due to a lipoma involving and compressing both facial and acoustic nerves at their origin from the brainstem. We compare medical therapy with botulinum toxin A and surgical treatment for this condition.

Case report

A 45-year-old woman came under our observation complaining of right blepharospasm associated with a vascular pulse in the ear of the same side. Her history started 8 years previously with vertigo and tinnitus in the right ear, gradually disappearing in few months without any medical care. Two years later, she noticed quivering of the right eyelid, especially in winter or during stressful periods. After three years, the symptoms became persistent and she came under our observation.

Neurological examination showed paroxysmal clonic twitches of the right orbicularis oculi muscle, resulting in a partial hemifacial spasm. We did not find other neurological signs. Audiometry did not disclose hearing impairment. Blood analysis showed a slight increase in cholesterol level (270 mg/ml).

Brain magnetic resonance angiography, associated with conventional T1-weighted (Fig. 1) and T2-weighted spin echo sequences (0.75 mm thickness), and computed tomography (CT) showed the presence of a small lipoma (10 mm diameter) in the right cerebellopontine angle. Supplementary fat- (Fig. 2a) and water-suppressed scans (Fig. 2b) (Dixon technique) demonstrated the fatty tissue

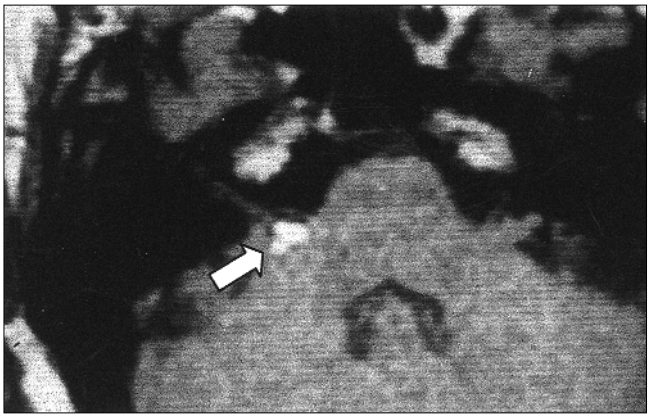

Fig. 1 *Cerebellopontine angle lipoma at the level of the root-entry zone of the cranial nerves VII and VIII*. T1-weighted spin echo MR image. MRI confirms the fatty nature of the lesion, as high signal intensity is detected on short TR sequences (*arrow*)

involving the cranial nerves VII and VIII at their origin in the brainstem.

The patient was started on treatment with botulinum toxin type A. Injection of the toxin in the right orbicularis oculi muscle led to a complete resolution of her symptoms, without any side effects.

One year later she preferred to undergo surgical care. At operation, cranial nerves VII and VIII were incorporated into the lipoma tissue. Lipoma also displaced facial nerves nearby a small vessel. The mass could not be totally removed. In order to decompress the facial nerve, a partial excision was performed and the complex (nerves VII-VIII and lipoma) was then isolated from a small adjacent artery. Histological analysis confirmed the diagnosis of lipoma.

Postoperatively, the HFS disappeared, but the patient noted right tinnitus and hearing loss. At a 10-month follow-up, she still complained of these symptoms. Audiometry and brainstem evoked potentials showed the presence of an ipsilateral acoustic nerve lesion.

Discussion

Lipoma is a rare intracranial tumour [5], and an unusual cause of hemifacial spasm. It is most often located in the pericallosal cisterns, and sometimes it is associated with corpus callosum hypoplasia [6]. Usually, this tumour is an occasional finding, unless it occurs in the CPA. In this location, it induces focal signs and symptoms in 80% of cases by compressing the cranial nerves [7].

Many hypotheses have been postulated to explain the presence of a lipoma in the central nervous system. According to one reliable report [8], this lesion cannot be considered a

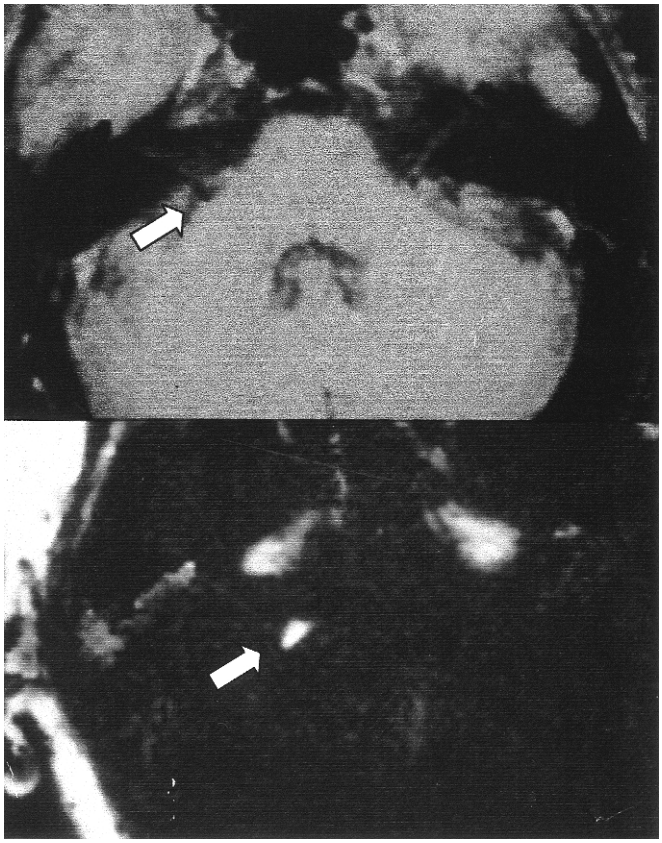

Fig. 2a, b *Cerebellopontine angle lipoma*. Axial MR fat (**a**) and water (**b**) suppression Dixon technique. Saturation of the signal from lipids can be appreciated on fat-saturated image (*arrow* in **a**), and loss of signal is detected. Water suppression image only depicts fatty tissues, as the small lipoma and extracranial fat (*arrow* in **b**)

real neoplasm or hamartoma, but rather a congenital malformation, in which case it could be the result of an abnormal persistence and maldifferentiation of the meninx primitiva.

In our patient, though both cranial nerves VII and VIII were involved in the lesion, the only persisting symptom was an incomplete hemifacial spasm. Although this symptom usually spreads to the rest of the face [4], the lower facial muscles were not affected during the 5-year preoperative period. Despite her past history of vertigo and tinnitus, her right acoustic nerve appeared clinically intact before surgical treatment.

Many authors reported a higher frequency of acoustic rather than facial symptoms associated with a CPA lipoma [6]. This can be explained by the fact that HFS is often treated without performing MRI. In fact, the request for imaging studies depends on the strategy of the selected treatment [4].

Our patient decided to undergo a surgical approach, although she was informed of all the possible risks.

Reviewing the last 10 years of the literature**,** we found many reports of cases affected by surgical complications.

The most common side effects included various degree of auditory impairment [6, 9-11] and facial palsy [9, 11, 12]. There has also been reported transient dysphagia [12], uvula deviation and cerebellar signs [11].

Surgery is helpful in relieving symptoms, but it should be also considered that the mass cannot be totally removed. Any attempt is technically hazardous [11], because of its strong involvement with cranial nerves and arteries [6, 12]. Though a partial excision is generally performed, the risk of local lesions remains still high.

In our patient, partial excision was associated with microvascular decompression. To improve benefit, this technique has to be preferred. In fact, the lesion can be directly or only indirectly responsible for the HFS, by displacing cranial nerve VII nearby a vessel and inducing a neurovascular conflict, as occurred in our patient. Clinical remission is obtained by separating the two structures.

Considering that lipoma is a benign mass, and that HFS is the only persisting symptom, we suggest that local injections of botulinum toxin A are preferable. Since action lasts for a short period, therapy should be repeated every 3-4 months. Remission is obtained in more than 90% of the patients [13, 14].

Acknowledgement We are indebted to Dr. Robert Esser, M.D., for his assistance and careful review of the manuscript.

Sommario *Il lipoma è un raro tumore dell'angolo pontocerebellare. Descriviamo il caso di un emispasmo facciale associato ad un lipoma situato all'interno del pacchetto acustico-facciale, all'origine del tronco encefalico. La paziente è stata sottoposta a terapia medica (tossina botulinica A) e chirurgica. Presentiamo anche una revisione degli ultimi 10 anni di letteratura, ponendo particolare attenzione al tipo di trattamento scelto.*

References

1. Wilkins RH (1991) Hemifacial spasm: a review. Surg Neurol 36:251-277
2. Evidente VG, Adler CH (1988) Hemifacial spasm and other craniofacial movement disorders. Mayo Clin Proc 73:67-71
3. Jannetta PJ, Abbasy M, Maroon JC, Morales-Ramos F, Albin MS (1977) Etiology and definite microsurgical treatment of hemifacial spasm. Operative techniques and results in forty-seven patients. J Neurosurg 43(3):321-328
4. Sprik C, Wirtschafter JD (1988) Hemifacial spasm due to intracranial tumor. An international survey of Botulinum toxin investigators. Ophthalmology 95:1042-1045
5. Kazner E, Stochdorph O, Wende S, Grumme T (1980) Intracranial lipoma. Diagnostic and therapeutic considerations. J Neurosurg 52:234-245
6. Rosenbloom SB, Carson BS, Wang H, Rosenbaum A, Udvarhelyi GB (1985) Cerebellopontine angle lipoma. Surg Neurol 23:134-138
7. Maiuri F, Cirillo S, Simonetti L, De Simone MR, Gangemi M (1988) Intracranial lipomas. Diagnostic and therapeutic considerations. J Neurosurg Sci 32:161-167
8. Truwit CL, Barkovich AJ (1990) Pathogenesis of intracranial lipoma: an MR study in 42 patients. Am J Roentgenol 155:855-864
9. Levin JM, Lee JE (1987) Hemifacial spasm due to cerebellopontine angle lipoma: case report. Neurology 37:337-339
10. Jallo JI, Palumbo SJ, Buchheit WA (1994) Cerebellopontine angle lipoma: case report. Neurosurgery 34(5):912-914
11. Inoue T, Maeyama R, Ogawa H (1995) Hemifacial spasm resulting from cerebellopontine angle lipoma: case report. Neurosurgery 36:846-850
12. Salpietro FM, Alafaci C, Napoli P, Cipri S, Tomasello F (1994) Cerebellopontine angle lipoma. Case report. J Neurosurg Sci 38:55-58
13. Moore P (1995) Botulinum toxin treatment. Blackwell Science, Oxford
14. Mauriello JA, Coniaris H, Haupt EJ (1987) Use of botulinum toxin injections in the treatment of one hundred patients with facial dyskinesias. Ophthalmology 94:976-979