diseases. Arterial Hb-CO concentrations in other neurodegenerative diseases need to be investigated to clarify the disease specificity of Hb-CO elevation. Although further large cohort studies are required, arterial Hb-CO concentration may be useful for objective monitoring of disease progression in ALS.

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The red ear syndrome

The red ear syndrome (RES) was described by Lance,[1] who suggested associations with upper cervical disorders and atypical trigeminal and glossopharyngeal neuralgias. Recently, Raieli *et al*[2] underlined the close temporal relationship between RES and migraine.

Patient 1

A 22 year old man, with a 12 year history of migraine without and with aura, experienced acute onset of burning and painful ear without other autonomic symptoms. These symptoms were always homolateral to the hemicrania and persisted for about two hours. The RES could be preceded by a headache. He also described sudden attacks of isolated burning ear without headache or autonomic symptoms. This isolated RES was limited to one side and could occur on either side with no preference for one side or the other. The attacks were not related to any particular stimulus. They occurred three or five times a month; approximately half of the episodes were followed by a migraine attack without aura.

Patient 2

A 92 year old woman experienced, 18 years ago, attacks of burning and red left ear associated with autonomic signs, such as left lacrimation. The attacks lasted for 20 minutes to two hours and could occur every day for 15–45 days every 12–18 months. No precipitating factor was found, and the attacks were resistant to non-steroidal anti-inflammatory drugs (indometacin). Subcutaneous sumatriptan was not given because of the age of the patient.

Neurological examination and brain magnetic resonance imaging (MRI) of both patients were normal.

Discussion

Patient 1 appeared to fit the criteria for RES as described by Raieli *et al*.[2] This type of RES occurs more frequently in children than in adults and is associated with a history of migraine with or without aura and of painful and red ear, unilateral or alternating, in isolation or associated with migraine attacks. This hypothesis was previously suggested by Hirsch[3] who reported unilateral and bilateral RES episodes in patients with ''vascular headaches''. Patient 2 was thought to have trigeminal autonomic cephalalgia (TAC).

Despite common elements, the two patients with RES described here differed in age, associated disorders, as well as the response to therapy.

Two different types of RES can be described: the first type occurs in children or young people and is clearly correlated with migraine.[2] These cases can be considered to be idiopathic. The second type occurs in adults and is associated with upper cervical disorders[1] or with TAC. RES has been described in association with diverse etiologies: migraine,[2] upper cervical disorders and temporomandibular joint dysfunction,[1] and TAC, in particular short acting, unilateral headache attacks with conjunctival injection and tearing (SUNCT), and hemicrania continua.[4–6] These associations suggest a common pathophysiological mechanism with activation of the trigeminovascular system. This variability occurs despite the belief that the final common pathway (the trigeminal–autonomic reflex) is presumably the same as in cluster headache.[7]

The trigeminal–autonomic reflex pathway consists of a brainstem connection between the trigeminal nerve and facial parasympathetic outflow.[7] RES ear episodes can be mediated by a cervico–autonomic reflex due to either an upper cervical disorder, or directly by trigemino-autonomic stimulation via the auriculotemporal nerve.[2] Trigeminovascular activation may produce pain that extends beyond the trigeminal territory. Thus the innervation of the earlobe, which is predominantly from the second and third cervical roots, can explain the association with upper cervical disorders.

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Arteriovenous fistula of the superficial temporal artery: an exceptional complication of the pterional approach

Despite the widespread use of the pterional approach in neurosurgical procedures, complications due to iatrogenic injuries of the superficial temporal artery (STA) are extremely rare. Iatrogenic pseudoaneurysms of the STA have been reported as a complication of craniotomy,[1] secondary to placement of external ventricular drainage catheters[2] or of a pin type headholder device.[3] Reported cases of iatrogenic arteriovenous fistula of the STA have occurred after hair transplantation[4] and after temporomandibular arthroscopy.[5] We report a case of iatrogenic arteriovenous fistula of the STA after pterional craniotomy. To the best of our knowledge, such a complication of craniotomy has not been reported before.

A 53 year old man was initially referred to our department with a grade 3 WFNS (World Federation of Neurological Surgeons) subarachnoid haemorrhage. Cerebral angiography revealed an anterior communicating artery aneurysm. A right pterional craniotomy was performed to clip the aneurysm. The superficial temporal artery was incised through a skin incision 7 cm above the tragus, and was coagulated carefully. The surgical procedure and postoperative course were uneventful, and the patient was discharged after two weeks with mild cognitive disturbances. Two months later, he complained of pulsatile tinnitus in the right ear. The tinnitus was exacerbated by lying on the right side. On physical examination, a thrill was palpable and a continuous murmur with systolic accentuation was audible on the pterional scalp incision above the tragus. The murmur and the thrill were abolished by compression of the proximal superficial temporal artery. Selective right external carotid artery angiography revealed an arteriovenous fistula between the main branch of the right STA and the homologous vein (fig 1). An internal carotid artery angiography was also performed, mainly to control the aneurysm, which showed no evidence of any contribution from the intracranial circulation. At operation, the arteriovenous fistula was proximally and distally ligated and excised completely. Postoperatively, the tinnitus disappeared, and the patient was discharged three days later. Six months after surgery there was no sign of recurrence.

Arteriovenous fistulas of the STA are rare lesions that occur most often after trauma or apparently spontaneously. The latent period between STA injury and the presentation of symptoms ranges from some days to 15 years. The presenting symptom usually includes a