Ramsay Hunt Syndrome Associated with Brain Stem Enhancement

Sabine Sartoretti-Schefer, Spyros Kollias and Anton Valavanis

AJNR Am J Neuroradiol 1999, 20 (2) 278-280
http://www.ajnr.org/content/20/2/278

This information is current as of April 20, 2024.
Case Report

Ramsay Hunt Syndrome Associated with Brain Stem Enhancement

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Summary: Postcontrast T1-weighted MR images in a patient with Ramsay Hunt syndrome showed an enhancing lesion in the region of the nucleus of the pontine facial nerve and abnormal enhancement of the intrameatal, labyrinthine, and tympanic facial nerve segments and of the geniculate ganglion, as well as enhancement of the vestibulocochlear nerve and parts of the membranous labyrinth. This enhancement most probably resulted from a primary neuritis of the intrameatal nerve trunks of the seventh and eighth cranial nerves.

MR images in patients with Ramsay Hunt syndrome (herpes zoster oticus) usually show intensely enhancing intrameatal nerve segments of the seventh and eighth cranial nerves and enhancement of parts of the membranous labyrinth and of different intratemporal facial nerve segments (1–5). However, only one report has described an abnormal T2-weighted hyperintense signal in the region of the pontine facial nerve nucleus in a patient with herpes zoster oticus (6). We present a case of clinically and laboratory proved Ramsay Hunt syndrome in which MR studies showed not only the typical MR characteristics mentioned above but also an enhancing, ischemic lesion in the pons on postcontrast T1-weighted images, most probably related to inflammation.

Case Report

A 58-year-old previously healthy man had acute and complete peripheral facial nerve palsy, hearing loss, and vertigo, which had ensued over the preceding 28 hours. Physical examination revealed painful cutaneous auricular vesicles within the external auditory canal and on the auricle; laboratory examination of the blood serum showed elevated IgM antibodies against varicella zoster virus. A severe sensorineural hearing loss was determined, and electroneurography revealed a neural degeneration of 85% maximum over the following 7 days. A diagnosis of Ramsay Hunt syndrome (herpes zoster oticus) was established. On the third day after onset of the palsy, an MR examination was performed on a 1.5-T MR unit using a phased-array dual coil with a diameter of 5 inches applied to both ears. Transverse precontrast T1-weighted images were acquired with a section thickness of 3 mm, a gap of 0.5 mm, a field of view (FOV) of 180 mm, a matrix of 256 × 192, and parameters of 500/163 (TR/TE/exci-

Received February 9, 1998; accepted after revision May 27.

From the Institute of Neuroradiology, University Hospital of Zürich, Frauenklinikstrasse 10, CH-8091 Zürich, Switzerland.

Address reprint requests to Sabine Sartoretti-Schefer, MD.

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cytes and plasma cells in the geniculate ganglion have rarely been reported (9, 10), whereas neuritis of the trunks of the seventh and eighth nerves within the internal auditory canal and of the intratemporal facial nerve segments has been described several times (10–12). Moreover, because cases of histologically proved herpetic inflammation of the facial nerve have been reported in which no inflammatory infiltrates were found within the geniculate ganglion (7, 11, 12), it has been concluded that geniculate ganglionitis does not exist as a specific pathologic entity (9) but occurs only in conjunction with simultaneous infection of the trunks of the seventh and eighth nerves within the internal auditory canal and secondarily spreads to the geniculate ganglion in some patients (10–12). Infection of the vestibulocochlear nerve as well as of the vestibular ganglion and the spiral ganglion has been demonstrated histologically as well (10). The infection presumably spreads to the membranous labyrinth via the vestibulocochlear nerve (5, 6) and via interneural connections that form anastomoses between the vestibular and cochlear nerves and the labyrinth (8).

Scattered inflammatory infiltrates along the intrapontine facial nerve from its nuclear origin within the caudal and lateral pons to its nerve root exit zone at the lateral pons have been described histologically (12). However, to date, involvement of the pontine nucleus and the intrapontine nerve course has not been demonstrated on postcontrast T1-weighted images. A single report has described an abnormal T2 hyperintensity in the region of the intrapontine seventh nerve nucleus in a patient with Ramsay Hunt syndrome (6), consistent with inflammatory involvement.

Cerebral angiitis has been reported in patients with herpes zoster ophthalmicus (13, 14). In these cases, retrograde spread of the virus along the intracranial branches of the trigeminal nerve to the arterial wall with secondary vascular wall inflammation (necrotizing angiitis) and vessel thrombosis was suspected, since the trigeminal nerve (especially the branches of the ophthalmic division) provides the sensory innervation of the intracranial portion of the internal carotid artery and of segments of the middle and anterior cerebral artery (13). This hypothesis is supported by electron microscopic findings showing viral particles in vascular smooth muscle cells and giant cells within the vessel wall, suggesting a direct viral infection of the vessel wall (13, 15). Hematogenous or contiguous dissemination via CSF pathways may cause cerebral angiitis as well (14).

In our patient with varicella zoster viral infection, a contrast-enhancing lesion in the region of the intrapontine facial nerve nucleus without elevated signal on T2-weighted images was seen on postcontrast T1-weighted images together with abnormal enhancement of the peripheral facial nerve along the distal intrameatal, labyrinthine, and tympanic nerve segments and of the geniculate ganglion, consistent with inflammation of the facial nerve nucleus within the pons; of the peripheral facial nerve itself; and of the geniculate ganglion. The primary site of infection was located within the nerve trunks within the internal auditory canal, from where the virus spread in a retrograde fashion along the subarachnoid nerve segment through the parapontine cistern to the pons and in an anterograde fashion along the peripheral nerves to the geniculate ganglion (16).

But why didn’t the T2-weighted images show any abnormality at the site of focal enhancement in the pons, similar to that seen in patients with rhombencephalitis (16)? In accordance with pathologic observations, intrapontine inflammation due to viral spread of varicella zoster viral infection to the brain stem may lead to a local angiitic vessel involvement and to vessel occlusion, with secondary
local ischemic infarction and breakdown of the blood-brain barrier. Therefore, the slight enhancement seen within the pons without associated hyperintensity on the T2-weighted images could have been caused by the presence of a small focus of subacute local ischemia.

Conclusion

Postcontrast T1-weighted MR images in a patient with Ramsay Hunt syndrome showed an enhancing lesion in the region of the pontine facial nerve nucleus and abnormal enhancement of the intrameatal and intratemporal nerve trunks, including the labyrinthine and tympanic nerve segments, the geniculate ganglion, the vestibulocochlear nerve, and parts of the membranous labyrinth. This resulted most probably from a primary neuritis of the intrameatal nerve trunks of the seventh and eighth cranial nerves with secondary anterograde and retrograde spread of the inflammation to the intratemporal nerve segments and to the brain stem.

References