‘Twenty-four-and-a-half’ syndrome and contralateral hemifacial spasm due to pontine carvernoma

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DESCRIPTION
A 50-year-old man who enjoyed good health in the past was admitted for diplopia, acute hearing loss of both ears and left-sided limb clumsiness on waking up. Physical examination showed right one-and-a-half syndrome (figure 1A–I and video 1), lower motor neuron weakness of the right face muscles (video 2), hemifacial spasm on the left side of the face (video 3), bilateral hearing loss and ataxia of left limbs (video 4). MRI of the brain revealed a right pontine carvernoma with perilesional oedema (figure 2A,B). Urgent operation for removal of pontine carvernoma was performed and his condition improved afterwards.

Figure 1 (A) Looking straight, (B) looking towards the right, (C) looking towards the left, (D) looking straight up, (E) looking straight down, (F) looking up and right, (G) looking down and right, (H) looking up and left and (I) looking down and left.
‘Twenty-four-and-a-half’ syndrome is a novel pontine neuro-ophthalmological condition characterised by ‘one-and-a-half’ syndrome with additional ipsilateral seventh and bilateral eighth cranial nerve palsies ($1^{1/2}+7+8+8=24^{1/2}$). This is caused by a right pontine carvernoma with perilesional oedema involving the ipsilateral abducens nucleus and the adjacent medial longitudinal fasciculus (MLF), or alternatively the involvement of the MLF plus the paramedian pontine reticular formation, along with ipsilateral facial nerve in this patient. Deafness is likely due to involvement of the cochlear nucleus complex bilaterally. The contralateral hemifacial spasm may be related to oedema of the contralateral dorsal pontine tegmentum causing hyperexcitability of the contralateral facial nerve motoneurons.
Learning points

▸ ‘Twenty-four-and-a-half’ syndrome is a novel pontine neuro-ophthalmological condition characterised by ‘one-and-a-half’ syndrome with additional ipsilateral seventh and bilateral eighth cranial nerve palsies.

▸ It is caused by a right pontine cavernoma with perilesional oedema involving the ipsilateral abducens nucleus and the adjacent medial longitudinal fasciculus (MLF), or alternatively the involvement of the MLF plus the paramedian pontine reticular formation, along with ipsilateral facial nerve in this patient. Deafness is likely due to involvement of the cochlear nucleus complex bilaterally.

▸ The contralateral hemifacial spasm may be related to oedema of the contralateral dorsal pontine tegmentum causing hyperexcitability of the contralateral facial nerve motoneurons.

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REFERENCES


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