Hypertrophic olivary degeneration after resection of a pontine cavernoma

Case illustration

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Hypertrophic olivary degeneration (HOD), an unusual type of neuronal degeneration, is caused by lesions involving the dentatorubroolivary pathway.1 In contradistinction to other types of neuronal degeneration, HOD is characterized by hypertrophy rather than atrophy of the affected structures.2 We describe a case of HOD in a patient after successful surgical excision of a brainstem cavernoma.

A 32-year-old man presented with new-onset diplopia and headache. Physical examination revealed a right sixth nerve palsy and decreased sensation in the right side of his face. Magnetic resonance (MR) imaging demonstrated an acute hemorrhage and hemosiderin staining within the left dorsal pons and midbrain, which were consistent with a diagnosis of cavernoma. A period of observation was recommended. Two months later he presented again with worsening diplopia and headache. Physical examination revealed a left one and one-half syndrome (paralytic pontine exotropia), right facial hypesthesia, lower motor neuron left facial paresis, lower-extremity hyperreflexia, severe dysmetria of the right upper extremity, and gait ataxia. Magnetic resonance imaging demonstrated enlargement of the lesion, which now extended from the pontomesencephalic junction to the pontomedullary junction and into the left cerebral peduncle (Fig. 1 left).

Total resection of the lesion (Fig. 1 right) was accomplished using a suboccipital transvermian approach with the patient in the sitting position. The lesion extended through the ependyma into the region of the right facial colliculus. The patient recovered well and displayed a significant improvement in his extracocular movements; only a mild right sixth nerve palsy persisted. The facial paresis, dysmetria, and long-tract signs also improved. Serial postoperative MR images demonstrated an uneventful resolution at the resection site. Three months postoperatively, however, changes were evident in the region of the left inferior olivary nucleus, which was located well caudal to the surgical corridor. Serial MR images demonstrated an area of restricted diffusion and hypertrophy of the inferior olivary nucleus (Fig. 2). At no time during the follow-up period was palatal myoclonus, a clinical correlate of HOD, noted. The neuroimaging findings and the clinical course supported the diagnosis of HOD.

Hypertrophic olivary degeneration is usually caused by a lesion in the triangle of Guillian and Mollaret, formed by the red nucleus, contralateral dentate nucleus, inferior olivary nucleus, and their connections.1,2 An examination with electron microscopy has allowed the demonstration of marked proliferation of neurofilaments and mitochondria, suggesting hypertrophy of single surviving cells rather than cellular proliferation.3 The presumed mechanism of HOD is transsynaptic degeneration; neuronal input to the inferior olivary nucleus is lost. Hypertrophic olivary degeneration has been reported following brainstem hemorrhage due to cavernous malformations in two cases.4,5 In the present case the likely mechanism was disruption of the central tegmental tract. Olivary degeneration after injury to any component of the triangle of Guillian and Mollaret should not be confused with neoplastic, vascular, or other primary pathological conditions of the medulla.

References


Fig. 1. Sagittal T1-weighted MR images. **Left:** Preoperative image revealing a large heterogeneous mass expanding the pons, which is consistent with the finding of a cavernous malformation. **Right:** Postoperative image confirming complete resection of the cavernoma.

Fig. 2. **Upper:** Serial axial MR fluid-attenuated inversion-recovery images obtained immediately and 3, 6, and 12 months postoperatively, demonstrating hyperintensity in the left inferior olivary nucleus. The hypertrophy appears maximal at 6 months; regression has begun by 12 months. **Lower:** Sagittal T1-weighted MR image demonstrating hyperintensity in the inferior olive and hemosiderin staining in the surgical cavity.