Demyelinating Disease Presenting As Wallenberg's Syndrome

Report of a Patient

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SUMMARY In clinical usage, Wallenberg's lateral medullary syndrome has become synonymous with lateral medullary infarction due to occlusion of one of the vertebral or posterior inferior cerebellar arteries. We report a patient in whom the pathological process was demyelination.

A 51-YEAR-OLD, right-handed man was in good health until he noted the abrupt onset of difficulty in walking. Later the same day, he experienced slurred speech, vertigo, dysphagia and hiccups. He visited a physician the following day because of coughing, and "bronchitis" was diagnosed.

On the third day of illness, he developed drooling from the right side of his mouth and "numbness" over the left side of his body. He was admitted to a local hospital where "evidence of cerebellar and brainstem dysfunction" was noted. A contrast-enhanced cranial CT-scan and lumbar puncture were normal. Ten days later the patient was transferred to the Denver Veterans Administration Medical Center for further evaluation.

His general physical examination was within normal limits, including examination of the heart and extracranial blood vessels. Mental status was normal. Multiple abnormalities of head and neck function were noted. Pain and temperature perceptions were diminished on the left side of the face. There was weakness of the entire right side of the face. Speech was dysarthric and on phonation the palate deviated to the left. His tongue deviated slightly to the right on protrusion. There was a right Horner's syndrome. Strength was normal in all extremities. Pain and temperature perceptions were diminished over the left side of the body. Joint position and vibratory sensations were intact in the legs. Movement of the right upper extremity was ataxic. Gait was wide-based and marked by a tendency to fall to the right. Muscle stretch reflexes were normal, and plantar responses were flexor.

The illness was complicated by recurrent aspiration pneumonitis and adult respiratory distress syndrome. Hypoxemia, hypotension and gastrointestinal hemorrhage developed, culminating in the patient's death 4 weeks after the onset of his symptoms.

Neuropathological Findings

At autopsy scalp, skull and meninges were unremarkable. The brain weighed 1,450 g and showed normal configuration and vasculature. The vessels showed minimal atherosclerotic changes. Perfusion of the vertebral arteries in the neck showed good flow; no thrombosis could be detected in the right posterior inferior cerebellar artery. The spinal cord was not removed.

On gross examination of multiple sections of brain, a single lesion was found. This consisted of an enlargement of the dorsal two-thirds of the right medulla.

Microscopic sections of this region demonstrated a well-demarcated, rounded area of total demyelination which extended inferiorty to the right olivary nucleus and across the midline dorsally (fig. 1). The medial lemnisci and pyramids were spared. At the margin, moderate spongiosis was present. Centrally, the lesion was hypercellular with reactive astrocytes predominating. Neuronal cell bodies, axons and spheroids were abundant (fig. 2), and individual dorsal medullary nuclei were discernible. Oligodendrocytes were virtually absent and atypical forms were not observed. Pleomorphic microglia were numerous in the surrounding parenchyma. Macrophages were somewhat less abundant. The Virchow-Robin spaces contained many macrophages and occasional lymphocytes; the vessels were otherwise normal.

FIGURE 1. Section of the medulla showing topography of demyelination. LFB-PAS. X6.5
Discussion

Classically, Wallenberg's syndrome involves ipsilateral cerebellar signs, palatal weakness, facial numbness and Horner's syndrome along with contralateral loss of pain and temperature perception over the body. Variations on this theme are common, as there is variability in both the direct and the collateral vascular supply to the brainstem. The non-classical features observed in our patient, such as diminished pain/temperature perception in the contralateral face and dysfunction of the ipsilateral seventh and twelfth cranial nerves, are previously reported and anatomically reasonable manifestations of the lateral medullary syndrome.

Diagnoses other than ischemic infarction are seldom considered when this clinical picture develops suddenly or within a few days. Yet, without question, the manifestations of the syndrome are dependent on the anatomy of the brainstem and not on any particular pathophysiologic process. The syndrome has been noted in association with mechanical trauma to the vertebral artery in the neck, vertebral arteritis, metastatic neoplasm, hematoma, aneurysm of the vertebral artery, herpetic brainstem encephalitis and arteriovenous malformations. To our knowledge, this is the first report of Wallenberg's syndrome as the presenting manifestation of a demyelinating process.

Our patient demonstrated classic pathological findings of demyelination. The lesion was indistinguishable from those seen in multiple sclerosis, but lacking additional lesions, such a diagnosis would be semantically inappropriate, at best. Unfortunately, evoked potentials and CSF immunoglobulin studies were not done. Thus, there is no laboratory support for such a diagnosis. Other illnesses, such as Binswanger's disease or central pontine myelinolysis, are associated with histologic changes of demyelination, but these diagnoses seem unlikely in our patient, given the absence of associated factors such as hypertension, alcoholism or severe electrolyte abnormalities. Moreover, an isolated lesion in this location would be unusual for either of these processes.

The benign prognosis of this syndrome militates against pathological confirmation of an ischemic etiology in most cases. While we do not propose that processes other than ischemic infarction are common causes of isolated lateral medullary syndrome, this patient serves as a reminder that the syndrome is not pathologically homogeneous.

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References

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