Basilar Artery Occlusion in a Young Patient With Wegener’s Granulomatosis

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Background and Purpose Stroke is an uncommon sequela of Wegener’s granulomatosis, resulting primarily from vasculitic infiltration of small to medium-sized vessels. We describe a young patient with Wegener’s granulomatosis and basilar artery occlusion.

Case Description A 26-year-old man with histopathologically confirmed Wegener’s granulomatosis suffered a brain stem stroke 24 hours after open lung biopsy. Angiography revealed midbasilar artery occlusion.

Wegener’s granulomatosis is a multisystem, necrotizing, granulomatous vasculitis that can afflict the central nervous system. Although peripheral nerves are frequently involved in patients with this disorder, cerebrovascular events are rare. Most of these have been attributed to small-vessel vasculitis. We describe a patient with histopathologically proven Wegener’s granulomatosis who developed a brain stem infarction secondary to basilar artery occlusion.

Case Report A 26-year-old white man was in good health until 3 months before admission when he experienced the gradual onset of rhinorrhea, epistaxis, and headache. Despite treatment for presumed nonallergic nasal rhinitis, his nasal complaints persisted, and he developed hemoptysis, lethargy, weight loss, and low-grade fevers. Nasal biopsy revealed epithelioid histiocytes and giant cells with necrosis but without vasculitis. A chest x-ray showed a 5-cm cavitary lesion in the left lung field, without evidence of lymphadenopathy or pleural or parenchymal disease. A diagnosis of Wegener’s granulomatosis was suspected, and given the absence of histopathological evidence of necrotizing granulomatous vasculitis, the patient was admitted to the hospital for an open lung biopsy.

The patient’s medical history was unremarkable. There was no history of cocaine or tobacco use, intravenous drug use, or human immunodeficiency virus risk factors. He was taking 500 mg/d of ciprofloxacin (for sinusitis), but he denied use of over-the-counter decongestants containing phenylpropanolamine. Physical examination revealed no abnormalities in the lungs, heart, or skin. There was a large clot of blood in the left nasal cavity. Neurological examination revealed no abnormalities. Laboratory data demonstrated mild leukocytosis (16 000/mm³), urine analysis was normal.

The patient underwent a thoracotomy with wedge resection of the lung mass without intraoperative complication. Twenty-four hours after the procedure, the patient developed clouding of consciousness, dysarthria, and right hemiplegia. Additionally, the patient was found to have an ophthalmoparesis involving the third and sixth nerves on the left. A more complete oculomotor examination was limited by the patient’s level of consciousness. Brain computed tomography, at that time, was normal. A lumbar puncture revealed a glucose level of 73 mg/dL, total protein of 21 mg/dL, 0 white blood cells, 10 red blood cells, and cerebrospinal fluid immunoglobulin index of 0.317 (normal values, 0.091 to 0.133). Cytoxan (100 mg/d) and dexamethasone (16 mg/d) were initiated. T2-weighted magnetic resonance imaging of the brain, performed 1 day after the event, showed increased signal in the left ventral and paramedian pons consistent with an infarction (Fig 1). Angiography, limited to the posterior circulation, revealed abrupt termination of flow in the basilar artery distal to the anterior inferior cerebellar artery (Fig 2). The proximal vertebral arteries were normal, and there was no evidence of segmental narrowing or dilation of small vessels. Laboratory data (obtained after the onset of neurological symptoms) demonstrated a significant leukocytosis (36 400/mm³), elevated erythrocyte sedimentation rate (90 mm/h), positive rheumatoid factor (1:320), elevated C-reactive protein (26.5 mg/dL), positive antineutrophil cytoplasmic antibody, a positive lupus anticoagulant (dilute thromboplastin inhibition ratio, 1.3 [normal values, 0.8 to 1.1]), and an elevated creatinine (blood urea nitrogen, 22; creatinine, 1.6). The transthoracic echocardiogram was normal, and 24-hour Holter monitor was only notable for mild ventricular ectopy.

Conclusions Patients with Wegener’s granulomatosis may experience premature large cerebral vessel occlusion. Putative etiologies in our patient include vasculitis, postoperative hypercoagulability, and/or intraoperative neck positioning leading to embolization. 

Key Words  • angiography  • basilar artery  • occlusion  • Wegener’s granulomatosis
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FIG 1. Magnetic resonance imaging (MRI) demonstrates pontine infarction. A, T2-weighted MRI (repetition time, 3000 milliseconds; echo time, 80 milliseconds) of the midpons demonstrates increased signal intensity in the ventral area (arrowhead) consistent with infarction. Hyperintensities and air-fluid levels in sinuses represent preexisting sinusitis. B, T2-weighted MRI caudal to that in panel A demonstrates increased signal intensity in the central and paramedial areas (arrowhead) consistent with infarction. Note absence of basilar artery flow void seen in panel A, consistent with proximal midbasilar artery occlusion.

Histological evaluation of the patient’s lung biopsy revealed necrotizing granuloma and extensive vasculitis. Skin lesions developed after the stroke, and biopsy of the skin showed leukocytoclastic vasculitis. A definitive diagnosis of Wegener’s granulomatosis was made, and cytotoxan and steroid treatment were continued.

On discharge from a comprehensive rehabilitation unit, the patient was more independent, needing only minimal assistance for ambulation and activities of daily living. His ophthalmoparesis completely resolved.

Discussion

The respiratory, renal, and central nervous system involvement, positive antineutrophil cytoplasmic antibody, and histopathologic evidence of necrotizing granulomatous vasculitis constitute a definitive diagnosis of Wegener’s granulomatosis in this patient. 3,4

A recent study examined the neurological manifestations of this disorder and found that cerebrovascular events are rare, occurring in 13 of 324 patients with Wegener’s granulomatosis. 5 Only 2 of the 13 patients underwent angiography, and in both cases the studies were negative. Indeed, with rare exceptions, previous angiograms in patients with Wegener’s granulomatosis and stroke have also failed to confirm vasculitis of the central nervous system. 5 The lack of sensitivity of angiography in this disorder has been attributed to predominant affection of small vessels (50 to 300 μm). 4 Intracerebral large-vessel involvement in Wegener’s granulomatosis has thus far been limited to postmortem histopathologic studies. 5,6,7 One of these reports described bilateral anterior cerebral artery thrombosis in an elderly white man. 7 Postmortem neuropathological examination suggested that these large-vessel occlusions resulted from necrotizing vasculitis and direct invasion of granulomatous lesion from the nasal cavity. To our knowledge, our patient represents the first radiographic demonstration of large-vessel occlusion in Wegener’s granulomatosis.

The mechanism of basilar occlusion in our patient remains less clear. Angiography consistent with vasculitis demonstrates focal narrowing or dilation in more than one vascular distribution. 8
artery, no other posterior circulation vessels were affected; the anterior circulation was not examined. While these observations call into question the role of vasculitis in this case, the finding of an elevated erythrocyte sedimentation rate, a positive antineutrophil cytoplasmic antibody, an increased white count, and the onset of skin lesions around the time of stroke suggest that the stroke occurred during a generalized "vasculitic flare." Because no specific clinical or serological markers of vasculitic exacerbation have been established, in the absence of cerebral histopathology, vasculitic involvement of the basilar artery cannot be proven.

In addition to vasculitis, cerebrovascular events in Wegener’s granulomatosis have been attributed to direct involvement of vessels by necrotizing granuloma. The absence of such a lesion on magnetic resonance imaging makes this an unlikely mechanism for stroke in this patient.

The temporal relation of the patient’s stroke to the open lung biopsy raises the possibility that postoperative factors contributed to the patient’s basilar occlusion. Recently, the clinical features of posterior circulation stroke after general surgery were surveyed. The majority of the patients examined (8 of 12) experienced infarction 24 to 48 hours after surgery, as did our patient. A multiplicity of putative mechanisms were postulated: (1) hypoperfusion in the setting of stenotic or vasculitic intracerebral vessels, (2) postoperative hypercoagulability, (3) vessel wall injury secondary to surgical instrumentation, (4) air or fat emboli introduced during surgery, and (5) neck rotation leading to compression or dissection of the vertebral artery with subsequent stasis of flow, thrombosis, and embolization once movement is resumed.

In our patient, hypoperfusion is unlikely given the absence of documented hypotension in the perioperative or postoperative period. Similarly, the patient did not have a central venous line placed before surgery, and therefore vessel injury is doubtful. Finally, the 24-hour lag in stroke onset after surgery makes the likelihood of intraoperative air or fat emboli small.

Hypercoagulability around the time of surgery represents a potential etiology in our patient’s stroke, particularly given the coexisting positive serum lupus anticoagulant. The contribution of lupus anticoagulant to the patient’s hypercoagulable state may have been particularly important given the patient’s vasculitic flare. As previously postulated, these procoagulant factors in the setting of reduced neck mobility during surgery could have led to intraoperative vertebral thrombosis with subsequent embolization to the basilar artery postoperatively.

In summary, we present the first angiographic evidence of basilar occlusion in a patient with histopathologically proven Wegener’s granulomatosis. The patient’s stroke may have resulted from vasculitis, hypercoagulability, intraoperative neck position, a paradoxical embolus, or a combination of these etiologic factors. Future studies may clarify the prevalence of large-vessel involvement in Wegener’s granulomatosis as well as the temporal relation of stroke to surgical procedures in patients with autoimmune vasculitides.

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References

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