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Neuroimaging Highlight

Ischemic Stroke Due to Carotid Artery Stenosis in a Patient with Hughes Syndrome

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A 23-year-old female presented with sudden right hemiparesis. Brain magnetic resonance imaging (MRI) showed acute left frontal infarcts (Figure 1A). She had no history of dry eyes, dry or ulcerated mouth, joint pain, or rash. High-resolution MRI (HR-MRI) showed an isointense and linear surface enhancement mass within the lumen of the left common carotid artery (Figure 1B and 1C). Contrast-enhanced ultrasound (CEUS) showed that microbubbles could enter between the mass and the carotid vessel wall (asterisk), and the mass was adherent to the intima of the vessel wall at the proximal end (Figure 1D). Digital subtraction angiography showed a regular intraluminal filling defect resulting in severe stenosis (Figure 1E). Transesophageal echocardiography revealed no intracardiac embolic sources or congenital heart disease. Routine coagulation tests showed significantly prolonged activated partial thromboplastin time (APTT) (76.00 s, normal value: 28.00–43.00 s). Mixing studies showed an abnormal APTT Immediate MixAPTT (64.50 s) and an abnormal APTT Incubated MixAPTT (69.20 s). Intraoperatively, the mass was found to be tightly adherent to the intima of the vessel wall at the proximal end (Figure 1F). A pathological examination of the contents of the carotid artery stenosis (Figure 1G-1I) showed a large number of fibrin threads, capillaries, scattered erythrocytes, and leukocytes with nuclear disintegration and early organization with the growth of fibroblasts. Pathological findings confirmed the diagnosis of an organized thrombus. Serological findings based on antibodies against cardiolipin (IgG > 120.00 MPLU/ml (positive: ≥12.0)), beta-2 glycoprotein I (IgG >200 AU/ml (normal value: 0–20)), and lupus anticoagulant confirmed Hughes Syndrome or antiphospholipid syndrome (APS). The patient was then treated with warfarin without relapse at the one-year follow-up.

APS, coined by Hughes in 1983,[1] is a disorder characterized by recurrent arterial or venous thrombosis, pathologic pregnancy, and consistently positive antiphospholipid antibodies.[2] APS may be idiopathic or secondary to an underlying disease, including systemic lupus erythematosus and other connective tissue diseases, neoplasms, infectious diseases, and drug use.[1] However, little is known about the mechanisms leading to thrombosis in APS. Proposed mechanisms include endothelial dysfunction, hypercoagulable state, elevated levels of complement activation products, and platelet activation.[3] Neurological manifestations, especially cerebral ischemia, are common in APS, but an organized intralumina+l thrombus, presenting as a mass in the common carotid artery, has not been reported previously.[4] The presence of lupus anticoagulant leads to a significant prolongation of the APTT value, and its presence is confirmed by mixed studies. The paradoxical prolonged APTT value with arterial thrombosis in young patients possibly suggests such a disease.

It is recommended that oral anticoagulation be maintained after thrombosis, with a target international normalized ratio between 2.0 and 3.0. For patients with a history of fetal loss, acetylsalicylic acid is recommended for prevention. Hydroxychloroquine has been used for the prevention of systemic lupus erythematosus and secondary APS. The clinical scope of this syndrome has expanded, and important advances in understanding its pathogenesis and clinical management have been made in recent years. It is an important cause of stroke in young people, usually causing in situ thrombosis of the intracranial vessels of the brain; a chronic internal carotid artery thrombus is a rare manifestation of the disease.

The differential diagnosis of carotid artery thrombosis includes carotid webs, which also causes blood flow eddies due to vascular web and thus leads to thrombosis. Angiography and arterial wall imaging showed a thrombus in the carotid artery but did not demonstrate web-like structures in our case. Since Ehrenfeld et al. reported the success of surgical treatment for a carotid web in 1967, carotid endarterectomy has been the treatment of choice for carotid webs.[5,6] Presently, there is no unified standard for treating an organized thrombosis because of the lack of high-quality clinical trials. Organized thrombus is similar to atherosclerotic plaques of the internal carotid artery. Surgical removal of either the carotid web or organized thrombus may be safe; however, there is a standard treatment of acute intraluminal thrombus presenting as an intraluminal mass, which is heparinization until the thrombus resolves. Carotid endarterectomy or stenting for acute intraluminal thrombus

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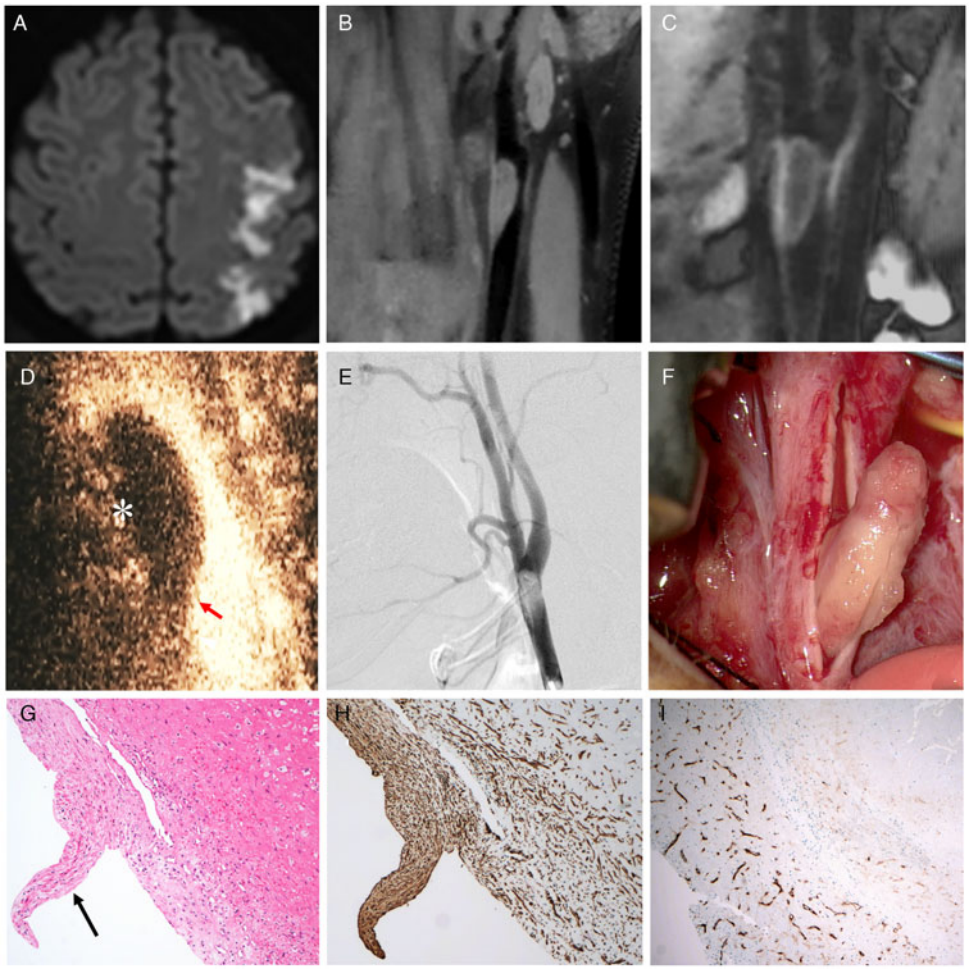

Figure 1: (A). Brain MRI showing acute embolic infarcts in the left frontal lobe. Isointense (B) and linear surface enhancement (C) mass in the left common carotid artery (arrow) on coronal HR-MR. (D) Contrast-enhanced ultrasound (CEUS) image of the carotid mass, microbubbles could enter between the mass and the carotid vessel wall (asterisk), and the mass was tightly attached to the intima of the vessel wall (red arrow). (E) Digital subtraction angiography showed severe stenosis of the common carotid artery with regular filling defects (arrow). (F) Intraoperatively, the mass was found to be attached to the intima only at the proximal end. Pathologic examination of the mass. (G) showed a large number of fibrin threads, capillaries, scattered red blood cells, and leukocytes with nuclear disintegration. Note the early organization with the growth of fibroblasts (SMA (+)) and capillaries (CD34 (+)) (H and I, respectively). Fibrous stripe-like structures are attached to the intima (see arrow).

is dangerous. And, it is still difficult to draw a significant management conclusion from a single case, which is a major limitation of this report. Further accumulation of such cases, analyses, and histological evidence in comparison with atherosclerotic stenosis can help to stratify stroke risk and determine surgical indications.

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Competing Interest. The authors declare that they have no conflict of interest.

Statement of Authorship. The author Y Du from Shenzhen Polytechnic contributed to the data collection and analysis, drafted and revised the manuscript. The author S Zheng from Peking University Shenzhen Hospital, China, contributed to the data collection and analysis. The author H Qi from Peking University Shenzhen Hospital, China, contributed to the data collection and analysis. The corresponding author Z Han from Peking University Shenzhen Hospital, China, critically reviewed and revised the manuscript.

Data Sharing Statement. Anyone can get access to the materials given reasonable reason.

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