

Papillary Fibroelastoma of the Heart

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Primary tumors of the heart are rare, with metastases being twenty times more common. Papillary fibroelastoma is the most common primary tumor of heart valves and comprises 7.9% of benign tumors of the heart.

We report the case of a 79 year old man, who presented with the sudden onset of loss of the ability to speak (expressive aphasia), and, a droop of the right side of his face. On admission to hospital, he appeared to be frustrated because of his inability to communicate. On examination, there was a right facial droop with loss of the nasolabial fold. The uvula was deviated to the left. The findings are indicative of a lesion in the left cerebral hemisphere. Computed tomography scan of the brain showed loss of brain tissue (acute infarct) involving the anterior distribution of the left middle cerebral artery, extending into the inferior frontal lobe, anterior to the Sylvian fissure. In right handed people, this lesion involves Broca's motor area of speech. An infarct is caused by the complete occlusion of blood supply, so an examination was undertaken of the carotid arteries by means of Doppler. It showed 60% stenosis (narrowing) of the Right Internal Carotid Artery, severe bilateral stenosis of the External Carotid Arteries, most marked on the right side, where it was 80%. Vascular occlusion in the brain can result from the impaction of material from the left side of the heart. Transesophageal Echocardiography of the heart revealed a mild redundancy of the mitral valve. A slightly irregular mass was attached to the posterior leaflet of the valve. It moved with the valve, and, on occasion, prolapsed into the left ventricle. There was a mild insufficiency of the mitral valve. The differential diagnosis was the possibility of a thrombus (vegetation) versus a papillary fibroelastoma or another benign neoplasm involving the mitral valve.

Magnetic Resonance Imaging of the brain confirmed the presence of an infarct (an area of avascular necrosis) in the left hemisphere, involving the left temporal and insular lobes and the left frontal lobe. There was occlusion of the left middle cerebral artery, involving a branch, distal to the trifurcation.

The patient's speech defect started to improve. He was put onto anticoagulants. There was no fever and blood cultures were negative, which was against the possibility of an infected thrombus on the valve. Surgical resection was undertaken of a 0.5 cm, red, soft polypoid mass which was attached to the middle portion of the mitral valve. The underlying valve tissue was roughly unremarkable.

Histopathologic examination showed the presence of a papillary lesion consisting of arborizing endothelium lined papillary fronds, which varied in thickness and in the degree of hyalinization. Some of the fronds were fibrocellular with a mucopolysaccharide matrix. These features are those of a papillary fibroelastoma (see Figure).

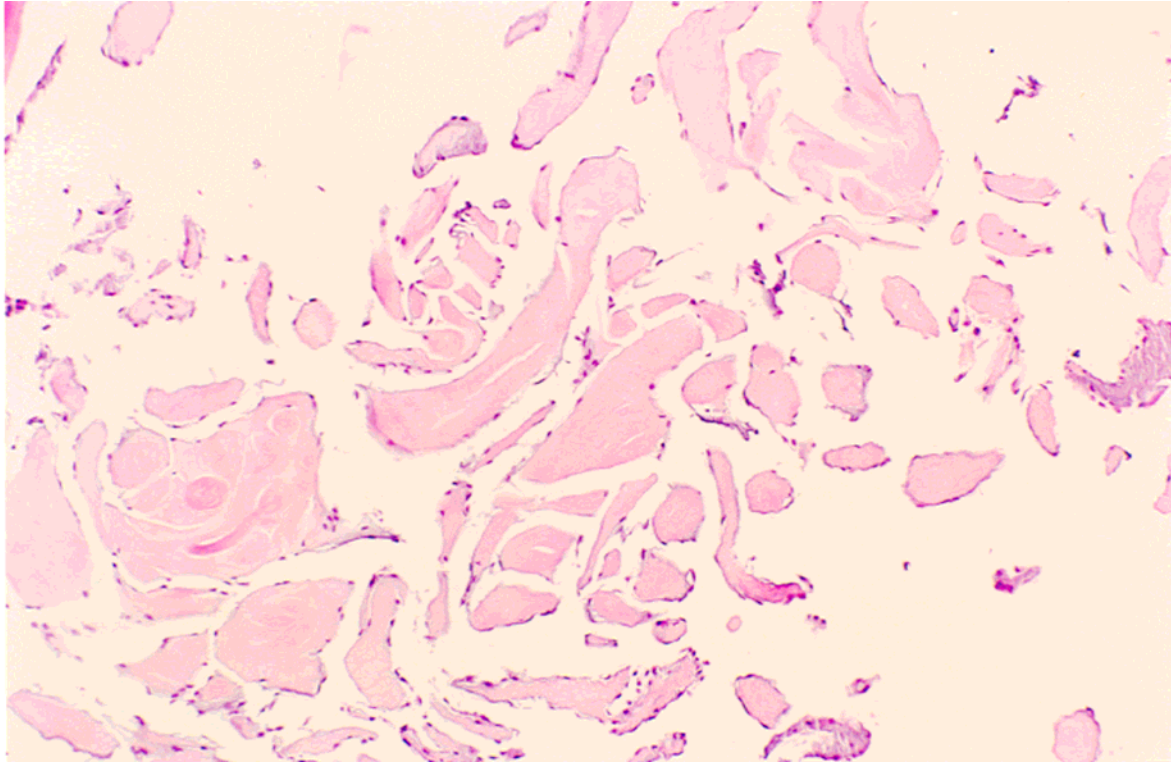


Figure. Histopathology of Papillary Fibroelastoma
Arborizing endothelium lined papillary fronds (Original magnification 40x)

The sequence of events in this patient was the dislodgement of a portion of one of the papillary fronds of the papillary fibroelastoma on the mitral valve and its transfer by the blood stream to the brain with impaction of it into a branch of the middle cerebral artery, distal to the trifurcation, leading to complete occlusion of this vessel. This resulted in complete deprivation of blood supply to the related area of the brain, which included Brocca's motor area of speech and caused the patient's expressive aphasia.