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Giant left atrial myxoma induces mitral valve obstruction and pulmonary hypertension

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Primary intracardiac tumor in the site of the left ventricle was described in 1559 and myxoma seems to constitute 40-50% of the primary intracardiac tumors. Atrial myxomas are sporadic in origin with unknown etiology and nearly 90% appear to be solitary and pedunculated; 75-85% seen in the left atrial cavity. It is often attached to the atrial septum. It occurs in middle age with the mean age of 56 years and more common in women. It could be familial in up to 10% of cases and when this occurs, they are more likely to be multiple and located in the ventricle also. The symptoms of atrial myxoma are related to location, embolization, and propensity to obstruct blood flow through the heart and are clinically difficult to diagnose. The diagnosis was possible only at post-mortem till 1951, after which diagnosis of the left atrial tumor has been confirmed by angiography. Till today echocardiography is a tool to diagnose intracardiac masses and it poses challenges in discrimination between primary cardiac tumors, such as myxoma, and other cardiac masses; which can be specified by histopathological examination. As literature reveals there are low surgical mortality and long-term good results in patients with cardiac myxoma. Histologically presence of myxoma cells in the background of the myxoid stroma is the diagnostic point. But the most challenging differential diagnosis is with mural thrombi showing myxoid changes. Calretinin is a novel marker which is positive in the myxoma cells and hence helps in differentiation from mural myxoid thrombi. Positive expression of Calretinin marker indicates that the myxoma cells may originate from endocardial sensory nerve tissue. In conclusion, Giant Left Atrial Myxoma is a rare benign lesion of the heart which can be treated by surgical removal. In general, it appears small and asymptomatic. However, it may present as large mass rarely because of the asymptomatic course despite the large size of a tumor and clinically mimic malignancy. Embolization of its fragments may cause pulmonary embolism when located in the right and systemic organ infarction when located on the left. Interference with mitral flow mimics mitral stenosis with massively dilated left atrium, pulmonary hypertension and chronic heart failure which progress over an appreciable duration of time.

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