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RESEARCH ARTICLE

PAPILLARY FIBROELASTOMA : A CASE REPORT AND REVIEW OF THE LITERATURE

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Abstract

Papillary fibroelastoma (PFEs) is a rare primary benign cardiac tumour which can cause severe embolic complications. We report the case of a 44-year-old with a sessile masse adhering to the aortic valve objectified by a transthoracic echocardiography. This mass was excised surgically and analysed histologically. The diagnosis of papillary fibroelastoma was then confirmed.

Introduction:-

Papillary fibroelastomaaccounts for about 7% of primitives heart tumours [1]. Before the appearance of ultrasound, the diagnosis was occasionally during heart surgery or autopsy [1,2].

Fibroelastoma originates from the endocardium [1-4]; he is usually asymptomatic but may also be the origin of serious complications such as accidents cerebral ischemic attacks, acute coronary syndromes, embolism lung disease and sudden death. We report a case of papillary fibroelastoma of the aortic valve discovered in a 44-year-old man presenting with dyspnea.

Patient and Method:-

It was a 44-year-old patient with no notable medical history, who presented with progressive worsening dyspnea associated with a feeling of blurred vision and lightheadedness. On clinical examination, the patient was in good general condition, heart sounds were regular with a systolic murmur at the aortic focus radiating to the neck vessels. Pleuro-pulmonary examination and the rest of the somatic examination were unremarkable. Chest X-ray showed a normal-sized cardiac silhouette and good parenchymal transparency. The electrocardiogram was unremarkable. Transthoracic echocardiography revealed severe calcified aortic stenosis and normal-sized left ventricle with preserved systolic function. A 15mm mass was noted on the aortic valve, without stenosis or mitral regurgitation. The patient underwent preoperative coronary angiography, which was normal, and the laboratory tests were unremarkable. The patient underwent surgery with a vertical median sternotomy, revealing a 15mm mass on the atrial side of the aortic valve, friable and easily detachable. The operative specimen was sent to the laboratory for histopathological examination. Microscopic examination revealed a papillary tumor lesion lined by regular endothelial covering resting on hyalinized and focally edematous connective tissue (Figure 1). The final diagnosis was papillary fibroelastoma of the aortic valve.

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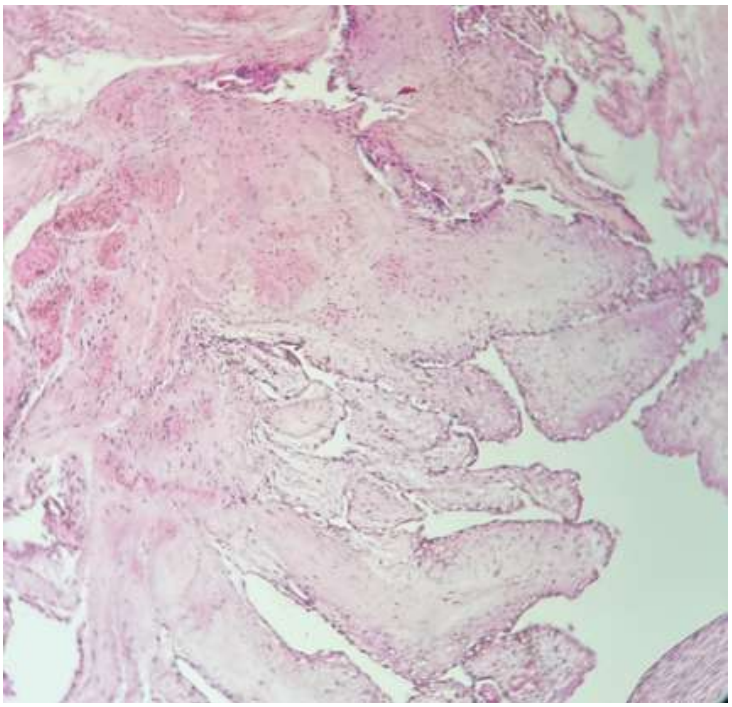

Figure 1:- Papillary formations lined by regular endothelial lining resting on hyalinized connective tissue in the center (hematoxylin-eosin staining).

Discussion:-

The emerging discipline of cardio-oncology covers the intersection of neoplasia and heart disease. This includes neoplasms directly involving the heart as well as indirect effects neoplasms may exert on cardiac structure and/or function.

Cardiac neoplasms can be categorized according to their histological characteristics (benign or malignant) and their site of origin (metastatic or primary). Metastatic lesions are exponentially (20- to 30-fold) more common than primary lesions and uniformly malignant. In contrast, primary lesions can be either benign or malignant.

Papillary fibroelastoma is the second-most common primary cardiac neoplasm (behind cardiac myxoma). Whether they represent neoplasms or not has also been debated, with some contending that they may be reactive because of the increased incidence in patients with a history of endocardial injury, such as prior instrumentation (2). However, rKRAS mutations have been documented recently in up to 80% of analysed lesions, suggesting that at least a subset of PFEs may have neoplastic origins (3).

Although PFEs may arise on any endocardium lined surface, 88% occur on valvular surfaces, with the left-sided valves more commonly affected than those on the right (4). PFEs can affect individuals of any age, although the mean age of detection is 60years. Some investigators have reported a female predominance, whereas others have suggested that PFEs are more common in men (5, 4).

The majority of patients are asymptomatic. When symptoms do develop, the most common are embolic, followed by heart failure, and last, sudden cardiac death (6–7). If the PFE is left-sided, embolic events may lead to territorial infarction (e.g., renal, mesenteric, cerebral, retinal, or limb, among others) (5,6,8). Right-sided PFEs may cause pulmonary embolism with subsequent pulmonary hypertension (9,10,11). Unusual presentations that have been reported in published studies include fever and thrombocytopenia. Although PFEs are typically< 1 cm they can occasionally be larger and obstructive, potentially resulting in sudden death (9,12,13).

PFEsfrequently have aclassicappearance on imaging, arisingfrom a stalk. PFEswilltypicallyexhibit a “frond-like” appearancethat has been likened to aseaanemone. This maybe harder to appreciate on smalllesionswith certain modalities, especially if the spatial resolutionis not sufficient. Echocardiographically, they have

acharacteristicspeckledappearance, withstipplingnear the edges. CMR (Cardiacmagneticresonance) may show some T2 hyperintensity in theselesionsrelated to connective tissue and mucopolysaccharide matrix. Significantenhancementisatypical, given the avascular nature of theselesions.

Macroscopically, the size varyingfromtwomillimeters to sevencentimeters [14]. Its implantation base ispedunculated. His body formsmanyfolds and itsconsists of arborizing, papillary projections arisingfrom a central fibrousstalk.

Microscopically, such projections are composed of avascularfibroelasticcores and lined by a single endocardialcelllayer. The underlying connective tissue isrich in collagenfibers, elasticfibers, glycosaminoglicans and cellssmooth muscle [14].

The clinicalcontext, the evolutionundertreatment anticoagulant and/or antibiotic and imaging data are necessary to suggest the diagnosis [15,16]. Surgical excision is the referencetreatment, mostoftenmaintaining the native valve [15-16]. In a minority of cases (typicallythoseinvolving large or multiple lesions), reconstruction withpericardium or valve replacement maybenecessary.

For our patient, the postoperativeimmediatewere simple with a stable hemodynamic state and a stay in intensive for 48 hours.

Conclusion:-

Although fibroelastoma is a tumor histologicallybenign, its evolution can be marked by complications fatal such as systemic and pulmonary embolisms, and death sudden. Its treatment is surgical by excision of the tumour.

The postoperative prognosis is excellent and the risk of recurrence tumour is low.

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