Myopericytoma of the Mitral Valve

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***Abstract*—We report the case of a malignant mypericytoma in the left atrium at the level of mitral valve. A 74-year-old man suffering from cardial deficiency, mitral regurgitation and oval neoformation. Following the outcome of specific examinations, he underwent surgical removal of a suspected myxoma-fibroelastoma. After that, the surgical collection was subjected to immunohistochemical staining; the microscopic observation points to mitral valve myopericytoma.**

Keywords--left atrium; malignant myopericytoma, perivascular neoplasms

I. INTRODUCTION

In 1996, Requena and his colleagues suggested that some adult cutaneous myofibromas were benign vascular neoplasms formed by immature (myo)pericytes; based on these observations, they proposed the term myopericytoma [1]. Subsequently, in 1998, Granter et al. described groups of vascular tumors with phenotypic characteristics that overlap with hemangiopericytoma and childhood myofibromatosis [2].; in fact, there is a close histological relationship between myopericytoma, myofibromatosis, solitary myofibrous and infantile hemangiopericytoma [3,4].

The rare cases of myopericytoma have “onion skin”-like proliferation with large, rounded and fused cells with cytoplasm rich in eosinophils; the cells arrange themselves around the vessels which are similar to those present in hemangiopericytoma.

By immunohistochemistry techniques, it has been observed that perivascular cells are reactive to smooth muscle actin (SMA), meanwhile their positivity to desmin is focal only. These results confirm the pericytic origin.

Generally, this type of neoplasm is typical of middle-aged adults at subcutaneous and superficial soft tissue of the extremities; it is characterized by a slow and painless growth.

Malignant myopericytoma, as its benign variant, presents a concentric proliferation of myoid cells that depart from the vascular walls and the same immunohistochemical profile. The malignancy features include elevated cellularity, increased number of mitosis, pleomorphism and necrosis. In addition, the aggressiveness of the neoplasm is independent from the anatomical development site [5].

In this report we show the case of a 74-year-old man suffering from cardial deficiency due to mitral insufficiency from oval neoformation.

II. CASE REPORT

A 74-year-old man with previous bilateral safenectomy, inguinal hernioplasty, arterial hypertension, chronic cerebral vasculopathy and subsequent episode of atrial fibrillation treated with pharmacological cardioversion, has reported ingravescent dyspnea with bilateral pleural effusion. Then, he was admitted for examination and treatment.

During his stay, he underwent a series of exams show in the table *“Table. 1”*.

TABLE I. EXAMS PERFORMED ON THE PATIENT

Exam	Result
Ecodoppler TSA	Calcific sclerosis of mild degree
ECG	Presence of spheroidal moving mass at the level of the rear flap that interferes with the functioning of the valve resulting in high grade steno-sufficiency
Coronarography	Moderate coronary atheromasia without critical stenosis

The reported results are consistent with a diagnosis of mitral insufficiency with left atrial myxoma; therefore, the patient underwent an operation of removal of left atrial neoformation originating from the valvular endothelium.

The samples were sent to the pathological anatomy laboratory as a suspected case of myxoma-fibroelastoma. The macroscopic description shows:

A) Mitral posterior flap of 1,5x0,6 cm with lobulated neoformation with smooth surface of mixoid aspect of 1 cm.

B) Laminar flap of cm 2,5x2 surmounted by rounded neoformation with a smooth surface of 2 cm of maximum diameter, hard-elastic consistency, grey-reddish marbled at the cut.

Formalin fixed, paraffin embedded and hematoxylin and eosin slides taken from surgical samples were microscopically examined. In continuity with fibrous tissue of valvular relevance, a high cellularity proliferation was observed *"Fig. 1"* and *"Fig. 2"*. The lesion presented apparently circumscribed, constituted of medium-size fused or roundish elements, with vesicular nucleus generally without prominent nucleoli, often with perivascular vorticoid concentric arrangement, around narrow vascular lumens, alternating with branched vessels of hemangiopericytoid appearance.

Frequent atypical cellular elements have been found, with dysmorphic hyperchromic nuclei (someone with regressive aspect), also present in the context of the wall of vessels; presence of dense reticulinic component, collagenized in some areas. Mitotic rate up to 5 mitoses/10 HPF in areas with increased cellularity, with occasional atypical mitosis and proliferative index (Ki67) around 20%. Numerous lymphocytes and mast-zellen, some eosinophils are added to the lesion. The central area of the lesion is hemorrhagic, surrounded by hemosiderin deposits.

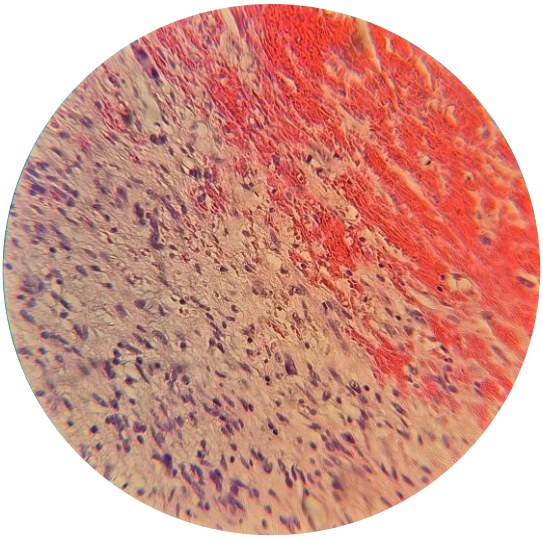

Fig. 1. Lesion observed in hematoxylin and eosin.

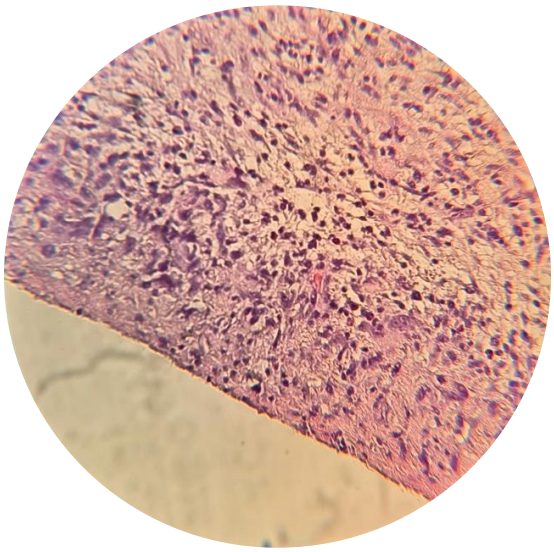

Fig. 2. Lesion observed in hematoxylin and eosin.

Later, the immunohistochemical profile was further investigated. *"Table II"* and *"Fig. 3, 4, 5* and *6"*.

The morphological and immunohistochemical picture suggest the diagnosis of mitral valve myopericytoma (WHO 2013), with intralesional hematoma; the margins were not clearly evaluable on the sample under examination.

TABLE II. IMMUNOHISTOCHEMICAL PROFILE

Staining IHC	Result
Vimentin	+ (spread)
SMA	+ (focal)
Desmin	+ (focal)
EMA	+ (focal)
S-100	-
CD31	-
CD34	-
KERpan	-
BCL-2	-
CD117	-
CD56	-
Calretinin	-

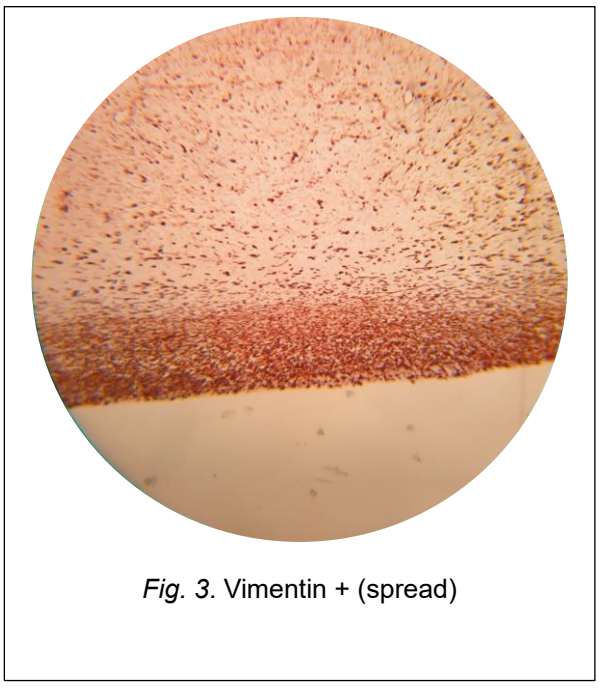

Fig. 3. Vimentin + (spread)

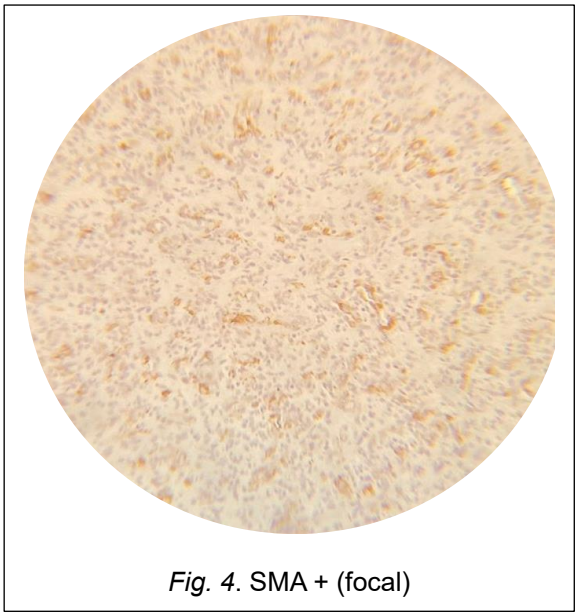

Fig. 4. SMA + (focal)

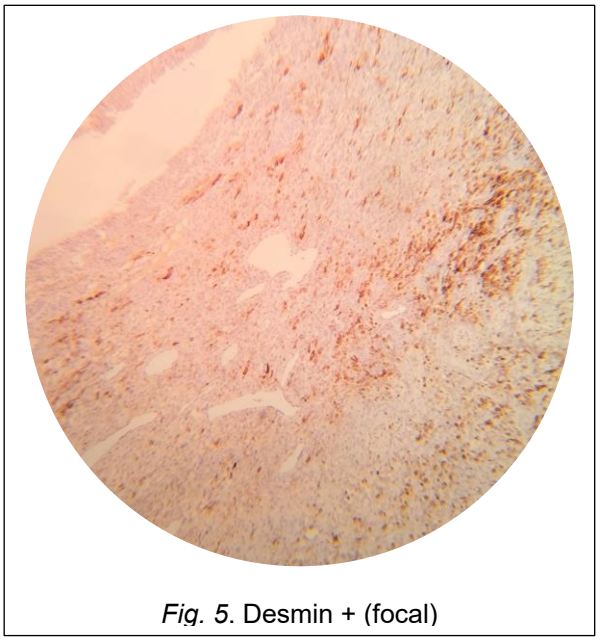

Fig. 5. Desmin + (focal)

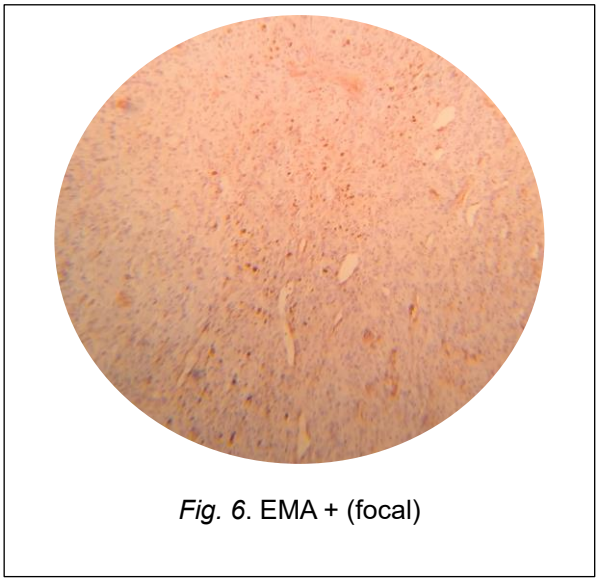

Fig. 6. EMA + (focal)

III. DISCUSSION

Myopericytoma has been described as a vascular tumor with similar characteristics to hemangiopericytoma and childhood myofibromatosis. It is characterized by an "onion skin" pattern with large rounded and fused cells with cytoplasm rich in eosinophils and it stands out from its benign variant by high cellularity, increased number of mitosis, pleomorphism and necrosis. Moreover, it is very aggressive in all anatomical sites.

In this case, the patient was admitted for mitral regurgitation and underwent surgery for the replacement of the mitral valve with biological implant and left atrial neoformation resection (hypothetical myxoma).

The surgical sample was subjected to a immunohistochemical staining in order to determine the nature of the lesion. Microscopic observation guides to mitral valve myopericytoma which is a rare lesion to observe. The presence of cytological atypia, sometimes regressive, and the presence of significant mitotic index with occasional atypical mitosis need attention on the biology of the lesion, not being able to exclude an aggressive behavior of the lesion on morphological basis.

The postoperative course was characterized by heart rhythm instability with atrial fibrillation lasting about 10 hours and subsequent restoration of the sinus rhythm. A pre-discharge echocardiogram was performed indicating a dilated left ventricle with normal parietal thickness, overall contractile and segmentary function of the preserved left ventricle with first grade diastolic dysfunction. Left atrium slightly dilated and no longer evident formations within it. Absence of pericardial effusion and slight bilateral pleural effusion.

At the time of discharge, drug therapy was prescribed to the patient.

After a year, the patient died following a pulmonary embolism.

IV. CONCLUSION

Mitral valve myopericytoma is a very rare malignant neoplasm difficult to diagnose often confused with hemangiopericytoma and myofibromatosis by which it shares similar microscopic characteristics.

It is therefore appropriate to conduct more in-depth studies in order to find a therapy.

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