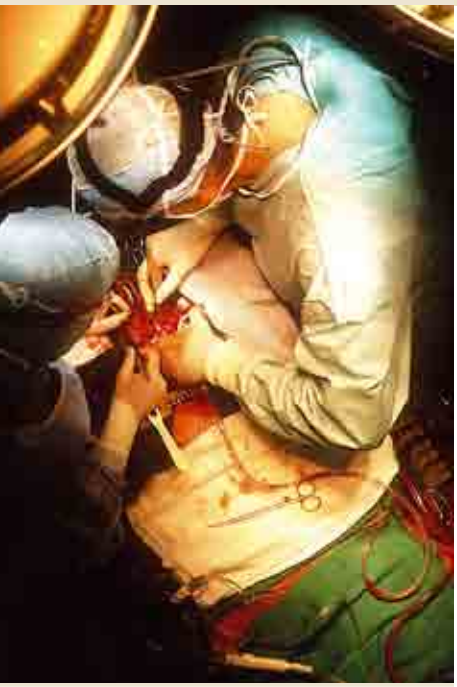
All That Glitters Is Not Gold and All Myxoid Tumors Are Not Myxomas



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Abstract

Primary tumors of the heart are rare, as opposed to metastatic lesions in this organ. Among the primary tumors, cardiac myxoma is the most prevalent neoplasia.

Here, we discuss the varied nature of myxoid lesions, which may be mistaken for myxomas, in two categories: benign and malignant.

Not only should the pathologist, but also the cardiac surgeon, be familiar with the diagnostic pitfalls and the differential diagnoses of myxoid lesions of the heart.

Echocardiographic findings, the rapidity of symptom development, and the location of the tumor, all serve as clues to a better approach in such patients.

Key words: Cardiac tumors, Myxoma, Myxofibrosarcoma

Cardiac myxomas are primary neoplastic lesions, arising in the endocardium. Primary tumors are rare in the heart; nevertheless, myxomas constitute about 80% of these primary tumors.

A wide range of tumors or pseudotumors, albeit rare, may be found in the heart. Therefore, both the pathologist and the surgeon should be familiar with the diagnostic pitfalls and the differential diagnoses. The myxoid lesions, which may be mistaken for myxomas, may be either benign or malignant. In the former group, only to name a few, we can mention papillary fibroelastomas, which occur on leaflets and are sometimes large in size. They have a characteristic gross appearance that is likened to "sea anemone" and in contrast to myxomas, which are vascular lesions, they are avascular structures and do not possess the stellate cells seen in myxomas. Another benign mass mimicking myxoma is cardiac hemangioma, in light of the fact that it is a vascular tumor like myxoma and may also adopt a myxoid stroma. What is used as the diagnostic aid here is the lack of myxoma cell on one hand, and the abundant presence of hemosiderin in myxoma on the other hand. Last but not least, in the first group, we can add organized thrombus as a lesion very likely to be frequently confused with myxoma. Thrombi lack myxoma cells, but possess granulation tissue and areas of scar and hyalinization.

As was mentioned before, the second category of lesions is malignant, and in this group we can name myxosarcomas. They are fibroblastic or fibrohistiocytic tumors that demonstrate extensive myxoid stromal component; however, they should not be considered as benign myxomas evolving into sarcomas. In fact, there is little evidence, if any, that myxomas undergo such progressive malignant transformation. Some 80% of these sarcomas occur in the left atrium, hence the significance of an accurate differential diagnosis from myxomas. A high proportion of myxoid

sarcomas of the heart are initially misdiagnosed as myxomas, but the clues that help differentiate them from myxomas include myxoma or stellate cells, hemosiderin-laden macrophages, and factor-XIII-positive dendritic cells. All these features are lacking in the malignant sarcomas.

In the malignant category of diagnostically challenging tumors, we should add leiomyosarcomas, which are of smooth muscle origins. A myxoid stroma is noted in about 25 % of these tumors, but the dense fascicles of smooth muscle easily distinguish this tumor from cardiac myxoma (1). During the operation, the myxoid appearance of the tumor may further support this assumption that we are dealing with a myxoma, but histopathological examination unexpectedly shows that the mass is not an atrial myxoma but rather a myxoid variant of a primary leiomyosarcoma. For such cases, immunohistochemistry and electron microscopy studies may be utilized to confirm the diagnosis (2).

A great number of malignant tumors in the heart are sarcomatous in origin, and it is not rare to see a patient who presents with symptoms of mitral valve stenosis because of a malignant tumor of the left atrium. We came across a case in the literature where echocardiography first revealed a left atrial mass supposedly interpreted as an atrial myxoma. Upon operation, the tumor appeared lobulated with a smooth surface. It arose from the posterior wall of the left atrium and extended into the mitral annulus. Wide surgical excision was possible with atrial reconstruction and mitral valve replacement. Histopathological and immunohistochemical studies were indicative of a malignant undifferentiated sarcoma (3).

The clinical and echocardiographic findings are extremely crucial to discriminate malignant tumors from benign myxomas, although the diagnosis may not be reached until the histological sections are available. To name a few, a rapidly progressive clinical course, or multiple tumoral growths, and non-septal attachment of the mass all suggest a malignant process. Therefore, careful and precise preoperative evaluation is said to be essential to identify such malignancies (4). Other authors too stress the importance of a comprehensive preoperative workup (5).

Here, we focus again on the intra-operative observation by the surgeon of the left atrial masses attached to the lateral atrial wall. On inspection during surgery, the malignant tumors are prone to infiltrate the mitral annulus and leaflet (6).

Conclusion

In view of the frequency of reports indicating the importance of preoperative workup and intra-operative observation as well as the varied nature of the lesions that may be mistakenly designated as myxoma, one should always remember that all myxoid tumors are not actually myxomas, as all that glitters is not gold.

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