

## MYXOMA OF THE HEART VALVES \*

### REPORT OF A CASE

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Although all tumors of the heart are rare, primary tumors of the heart do occur in the form of either sarcoma, fibroma, lipoma, myoma or myxoma, or combinations of two or more of these. With the exception of sarcoma these tumors tend to be benign. There has been considerable confusion and controversy about the classification of myxoma and fibroma of the heart ever since Czapek<sup>1</sup> asserted that many of the alleged myxomas of the heart were really organized thrombi. Thorel,<sup>2</sup> Stahr,<sup>3</sup> and Husten<sup>4</sup> concurred with Czapek in this opinion, while Ribbert,<sup>5</sup> Link,<sup>6</sup> Mandelstamm,<sup>7</sup> and Hagedorn<sup>8</sup> presented strong arguments in favor of the neoplastic nature of these tumors. The differentiation is made difficult by the microscopic similarity between myxoma and organized thrombi. Both show a homogeneous, poorly cellular matrix, and both are covered with endothelium. They differ, however, in that the myxomas are avascular, and usually have mucin and elastic fibers present, while organized thrombi are vascular, generally contain degenerated blood pigment, and have neither mucin nor elastic fibers. When these differences are considered, along with the gross appearance, there is little room for argument, for the organized thrombus is typically a smooth flat tumor with a broad base and more or less evidence of contracture and scarring, while the myxoma is a pedunculated, lobulated tumor with a narrow base and no evidence of contracture.

Although myxomas have been found in all chambers of the heart the sites of predilection are the interauricular septum of the left auricle and the heart valves. The myxomas that are found on the heart wall and septum are characteristically large tumors that may grow to a size sufficient to fill the entire chamber in which they are located and cause valvular insufficiency by occlusion of the valves.

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Choisy<sup>9</sup> first described a heart polyp of the left auricle situated in the valvula foraminis ovalis. Within the next few years similar cases were reported by de Puisaye,<sup>10</sup> LeGendre,<sup>11</sup> and Proust.<sup>12</sup> These tumors had the lobulated appearance and microscopic peculiarities of the myxoma. The valvular myxomas are more uncommon than those in the heart wall and are much smaller, varying from 6 to 15 mm. in diameter. They resemble the larger growths in that they are pedunculated, but show a characteristic papillary structure which the larger ones do not have. They are found on all the heart valves but are slightly more common on the tricuspid valve.

A total of 22 myxomas located on the heart valves have been reported. These are tabulated in the accompanying table. A consideration of the table will show that these tumors may occur at any age from birth to old age, and in either sex. As a rule, they are found accidentally at autopsy, having failed to give rise to clinical signs or symptoms during life.

#### REPORT OF CASE

*Clinical History:* A. H., a negress, 62 years of age, was admitted to St. Luke's Hospital on Dec. 26, 1932 because of failing vision in both eyes. Except for a pelvic operation 8 years before she had always been well.

The patient was an obese colored female with normal vital signs and, except for the eyes, normal physical findings. The pupils were equal, but both lenses showed large opacities. After a preliminary iridectomy the patient was discharged and readmitted 3 weeks later for a right cataract extraction, which was uneventful. On the 6th postoperative day she suddenly collapsed and showed the typical picture of circulatory failure, the blood pressure being 60/40 and the pulse feeble. Roentgenograms and electrocardiograms offered no assistance in making a diagnosis. She became steadily worse and died the next day.

#### POSTMORTEM EXAMINATION

The body was that of an obese colored female. The positive findings were limited to the heart and pericardial sac. On opening the pericardial sac about 100 cc. of cloudy fluid were seen. The pericardium was hemorrhagic, and covered with small tags of fibrin. The heart weighed 325 gm. and the left ventricle measured 1.5 cm. in thickness. None of the valves was thickened, and there was no evidence of endocarditis. On the anterior cusp of the tricuspid valve, situated in the middle of the valve and about 5 mm. from its

TABLE I  
Summary of Previously Reported Valvular Myxomas

Author	Year	Type of Tumor	Situation	Age yrs.	Sex	Remarks
Luschka <sup>18</sup>	1857	Myxoma?	Pulmonic	40	F	Old endocarditis present. Author considered it inflammatory
Curtis <sup>14</sup>	1871	Myxoma	Mitral	83	F	
Debove <sup>15</sup>	1873	Myxoma	Tricuspid	38	M	All were highly papillary
Ribbert <sup>1</sup>	1897	Myxoma	Pulmonic	"	"	
			Tricuspid	"	"	
Guth <sup>16</sup>	1898	Papillary myxoma	Tricuspid	54	F	Other valves normal Endocardium thickened. Multiple fibromas in walls
Simmonds <sup>17</sup>	1901	Papillary fibroma	Tricuspid	25	..	
Reitmann <sup>18</sup>	1905	Hyalofibroma	Pulmonic	74	M	Papillary; matrix very poor in cells Valve thickened
Leonhardt <sup>19</sup>	1905	Myxoma	Mitral	26	F	
Djewisitzky <sup>20</sup>	1906	Myxoma	Aortic	38	M	Patient had a hypernephroma Kochlin considered these tumors to be Lambd excrescences
Blumgart <sup>21</sup>	1907	Fibromyxoma	Mitral	86	F	
Hagedorn <sup>2</sup>	1908	Myxoma	Mitral	Middle aged	M	Patient had a hypernephroma Kochlin considered these tumors to be Lambd excrescences
Kochlin <sup>22</sup>	1908	Myxoma	Pulmonic	19	F	
			"	53	"	Some thickening of cusp Similar growth on tricuspid
			Aortic	60	"	
Forel <sup>23</sup>	1910	Fibroma	Aortic			Some thickening of cusp Similar growth on tricuspid
Dean and Falconer <sup>24</sup>	1913	Myxoma	Pulmonic	53	M	
Kornfeld <sup>25</sup>	1928	Myxoma	Pulmonic	68	M	Similar growth on tricuspid
Abrahamet <sup>26</sup>	1931	Myxoma	Pulmonic	Newborn infant		
			Aortic	"		

edge, was a small spherical tumor measuring 6 mm. in diameter and projecting 4 mm. above the surface of the valve, to which it was attached by a short wide pedicle. Its surface was finely nodular and it had a translucent gelatinous appearance (Fig. 1). The coronaries were thin and showed no evidence of sclerosis.

Permission for examination of the brain was not obtained, but it was assumed that the patient died of a cerebral accident.

#### MICROSCOPIC EXAMINATION

The cusp containing the tumor was excised, and sections were prepared with hematoxylin and eosin, Van Gieson's, thionin, mucicarmine, and Weigert's elastic tissue stains.

There is no microscopic thickening of the cusp, and there is well marked differentiation of the layers of the valve. A narrow pedicle attaches the tumor to the cusp, and the endothelium and elastic laminae are reflected onto the pedicle. The pedicle very shortly spreads out, and divides into two main limbs, from each of which numerous papilliform processes arise (Fig. 2).

Stained with hematoxylin and eosin the tumor matrix is pink. There is a complete endothelial covering over the entire tumor. The papillae contain only a few cells and there are no blood vessels present; the pedicle, on the other hand, contains many stellate and spindle-shaped cells which have fine processes extending from them into the fine fibrillar groundwork present throughout the entire tumor (Fig. 3).

The test for mucin with thionin is only faintly positive; the stain with mucicarmine, however, is strongly positive, the entire matrix taking a deep red stain. Van Gieson's connective tissue stain shows a large number of coarse, pink-staining fibrils in the pedicle which extend into the papillae and become lost. The presence of elastic fibers is proved by Weigert's elastic tissue stain, which shows a large number of vertically placed, blue-staining fibers throughout the entire tumor.

The tumor presented then is one arising by a well circumscribed pedicle from the tricuspid valve, and which is free of any inflammatory change. The tumor contains mucin and elastic fibers. Part of this tumor, however, consists of hyaline connective tissue.

## DISCUSSION AND SUMMARY

When a tumor is found on a heart valve it may be either a Lambl excrescence, a product of an inflammatory reaction or a neoplasm. Organized thrombi do not have to be considered, since they do not occur on valves.

Lambl<sup>27</sup> first described the excrescences which bear his name as thread-like structures 2 to 3 mm. in diameter, occurring on the aortic valve. These are papillary in form and resemble myxomas microscopically. Koechlin<sup>22</sup> felt that all the valvular myxomas should be called Lambl excrescences. In examining 150 bodies he found thread-like growths on the valves in 20 per cent of the cases. However, all the Lambl excrescences reported have occurred on the aortic cusps with the exception of 2 cases reported by Koechlin,<sup>22</sup> in which they were found on the mitral valves, and in these there was evidence of an old endocarditis present. Whereas Lambl excrescences are generally found in a heart exhibiting endocardial changes myxomas occur in hearts with a normal endocardium. Moreover, myxomas are larger, measuring 6 to 10 mm. in diameter, and are more compact. Inflammatory growths on the valves are generally accompanied by gross thickening of the valves, destruction of valvular contours, and by microscopic scarring.

If it be assumed, as did Ribbert<sup>5</sup> and Leonhardt,<sup>19</sup> that true mucin occurs in a true tumor, and could come about in no other way, then these valvular growths can be readily differentiated by the various chemical and color reactions characteristic of mucin. Ribbert believed that the myxomas were true tumors which originated from embryonic connective tissue, and that ultimately the mucoïd tissue gave rise to fibrous tissue. The fibroma described by Hagedorn<sup>8</sup> represents the endpoint in this development.

Reitmann,<sup>18</sup> on the contrary, believing that the myxomas represented a stage of degeneration of connective tissue tumors, designated his case as a hyalofibroma.

Whether the myxomas are really neoplasms, or whether they represent a degenerative process in connective tissue growths is still open to question. The fact is, however, that they do represent a group of tumors that occur in the heart, and have rather uniform morphological characteristics.

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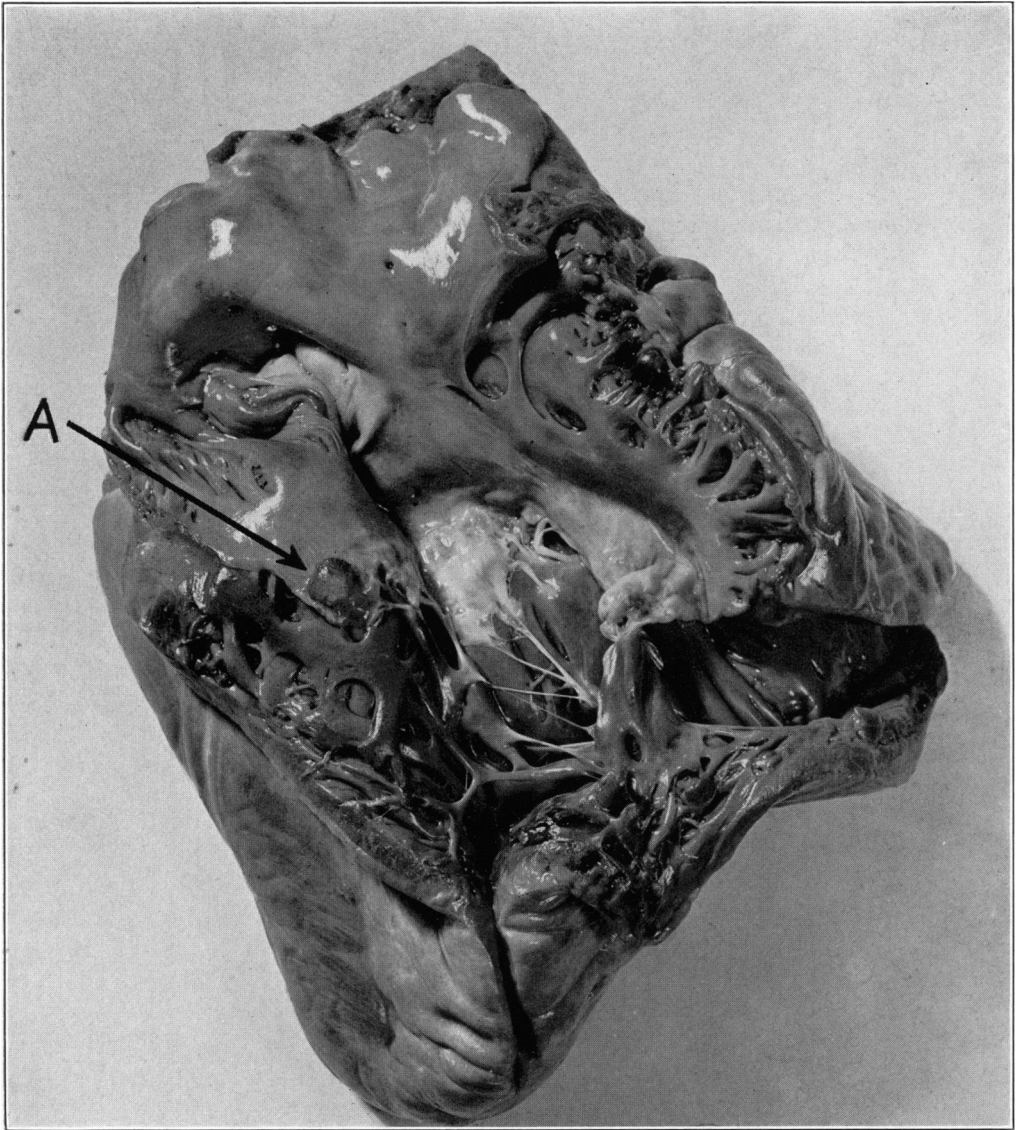
DESCRIPTION OF PLATES

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PLATE 104

FIG. 1. Heart showing a myxoma (A) attached to the tricuspid valve on the auricular surface. The right ventricle has been cut open. Approximately natural size.

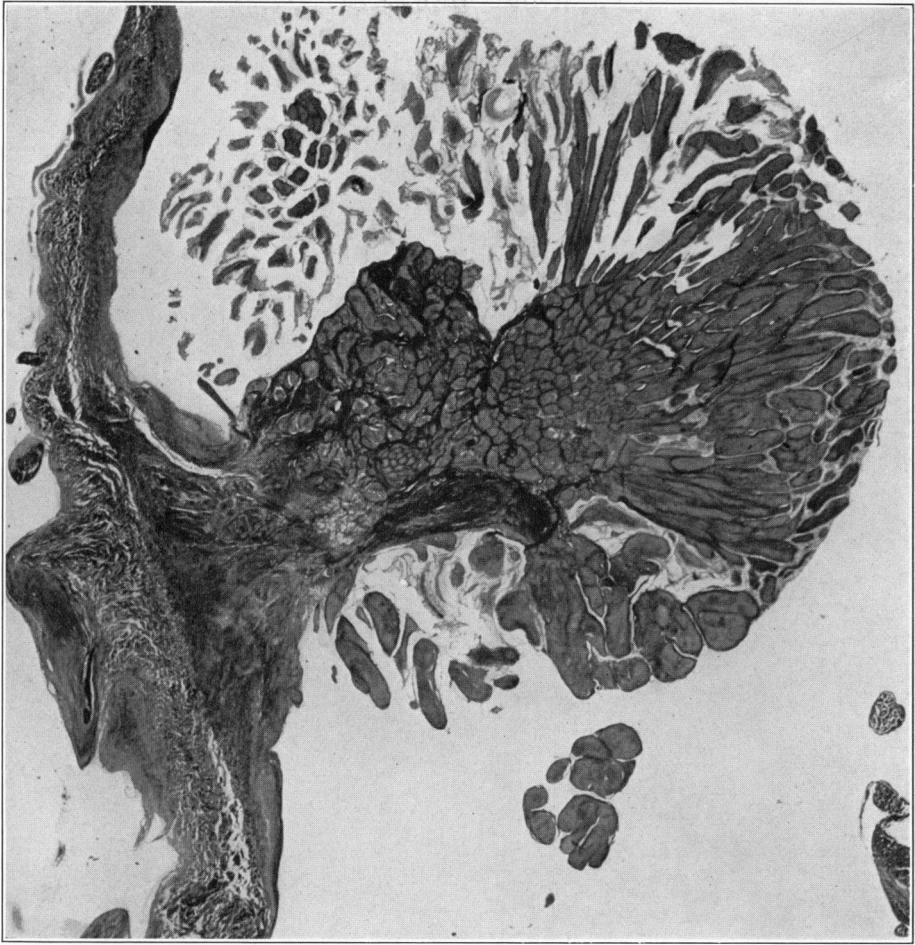




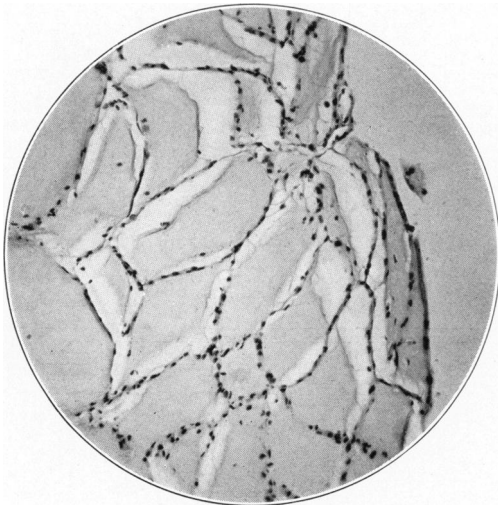
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PLATE 105

- FIG. 2. Microscopic section showing a myxoma attached to the tricuspid valve.  
× 18.
- FIG. 3. Cross-section of several papillae of the myxoma showing the endothelial  
cells covering them, and the poorly cellular matrix. × 100.



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