THE DIAGNOSTIC APPROACH TO AURICULAR MYXOMAS*

EDMOND PAQUET, M.D., *Quebec, Que.*

THE PURPOSE OF THIS PAPER is to consider the most recent data on auricular myxomas and report briefly four new cases.

Many investigators have written comprehensive studies on cardiac tumours (Leriche and Bauer,[11] Yater,[14] and others). More recently, Mahaim[12] published a complete monograph in which he reviewed and analyzed a great number of cases, thus reviving interest in these tumours, usually oddities discovered by chance.

Apparently, primary tumours of the heart are relatively infrequent as compared with cardiac or pericardial metastases. Primary tumours are benign and of the myxoma type. Myxomata are mostly found in the left auricle but not infrequently in the right auricle; thus, two cases in our series were found in the latter.

The association of a congenital cardiac anomaly with auricular myxoma as in the first patient in our series, who also had an interauricular septal defect, seems to be entirely fortuitous. As to the age and sex incidence, results of statistical surveys vary greatly and are not conclusive.

Anatomically, the heart appears dilated and the auricle strikingly enlarged. The valves are not damaged, but it is common to find the atrioventricular orifice obstructed by the tumour, which partially fills the auricular cavity and tends to slip into the ventricle.

More often solitary and of variable size, myxomata are attached to the interauricular septum by a short pedicle derived from the endocardium and inserted around the foramen ovale. In addition, the tumour appears as a smooth, glittering, globular or polypoid mass, occasionally villous, reddish or yellowish-grey, semigelatinous or transparent, soft or elastic. Microscopically, the tumour mass is enclosed in a continuous endothelial layer. Its cellular content, though differentiated, is poorly specialized. Most of the few cells it contains appear ramified, spindle-shaped or round. Connective as well as elastic fibres are disseminated through a mucoid-reactive substratum, i.e. pink staining with mucicarmine or violet with thionine. In addition to these, it is common to find many new-formed vessels along with hæmorrhagic plaques and hæmosiderin deposits. The predominance of either connective fibres or vascular tissue gives rise to distinctive types called fibromyxoma or angiomyxoma. It is of interest to note that the origin and nature of these tumours have for many years given rise to two schools of thought. Some investigators have supported the view that a myxoma is nothing less than a degenerated organized thrombus. Nevertheless, on the basis of embryology, it is now generally acknowledged that a myxoma is a true neoplasm. Accumulating evidence has proved that it is derived from the embryonic rests of the fossa ovalis. It is known that the fossa ovalis is the part where the last phase of the developmental cycle of the heart takes place. Thus, it is likely that the predilection of these tumours for the foramen ovale is not purely coincidental. Proponents of this modern concept find abundant support for their views.

It must be kept in mind, as Anderson[2] stated, that such tumours projecting into a cardiac cavity may be associated with thrombosis on their surface and embolizing potentialities. Furthermore, it is noteworthy, as Brown[1] remarks, that an organized thrombus never undergoes myxomatous degeneration in peripheral vessels where such thrombosis is commonest. He also calls attention to the fact that myxomata are much less frequent in the ventricles, where thrombosis is observed more often than in the auricles.

In any event, whatever the true nature of this neoplasm, it concerns the clinician primarily because of the mechanical accident it produces in the heart. Acting like a foreign body in the auricular cavity, this pedunculated, mobile tumour manifests itself clinically by cardiac embarrassment. This appears first by occlusion of the atrio-ventricular orifice, evidenced by a syndrome resembling mitral stenosis. It cannot be emphasized too strongly, however, that a previous history of rheumatic fever is usually lacking. If rheumatic fever has occurred previously as may happen occasionally, the diagnosis is of course impossible to make. Auricular filling is also partially responsible for congestive heart failure. Intermittent at first, the cardiac disturbances become progressive, then persistent and refractory to the usual therapeutic measures.

*From the Department of Cardiology, Director—Prof. Lenègre, Boucicaut Hospital, Paris.

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The impossibility of satisfactorily establishing the cause of a severe organic cardiopathy, and the symptomatic variability observed with postural changes of the patient, appear as the most prominent features of the disease. The chief manifestations are dyspnœa, auscultatory changes, neurological disturbances and embolic phenomena.

Dyspnœa is an important source of trouble. Intermittent at first, it becomes progressive and severe. Not infrequently, attacks of paroxysmal dyspnœa associated with cyanosis occur in the supine position. The attacks are likely to depend on transitory occlusion of the pulmonary veins or mitral valve and consequently pulmonary stasis. If chronic and progressive, this mechanical embarrassment will produce pulmonary hypertension and right ventricular hypertrophy.

Auscultatory findings are of interest, for they suggest the diagnosis of mitral stenosis and may vary with postural changes. If the patient is in a standing or sitting position, the tumour would tend to narrow the mitral orifice and produce the auditory signs so characteristic of mitral stenosis, whereas with the patient in the supine position one would hear a systolic murmur. Nevertheless, it must be noted that the majority of cardiologists disagree on the constancy and exactness of such findings.

Finally, it is commonly observed that blood pressure and particularly pulse pressure are low in these patients, probably as a result of the same obstructive mechanism.

Sudden occlusion of the mitral valve may produce cerebral anoxia and consequently dizziness, unconsciousness, syncopal faints or epileptiform attacks and even motor disturbances.

Of greater severity are embolic complications. The sources of the emboli may be thrombi detached from the surface of the tumour or a tumoral fragment. In order of frequency, cerebral, pulmonary or peripheral embolism may be observed. It should be pointed out that cerebral embolism has no diagnostic value, for it is a common complication of severe affections of the left heart. It must be emphasized, moreover, that pulmonary embolism is mostly encountered with right auricular myxomata.

It may be possible occasionally to make the diagnosis of myxoma of the left auricle from peripheral embolism. Obviously, such an embolus would be myxomatous, and would have to be removed and examined. The conditions of such a diagnosis are of course hazardous and unreliable. The occurrence of anginal pains is not unusual and has led to the suggestion that they may depend on coronary embolism, but anatomical studies do not support such an hypothesis. Occasionally, the clinical features may be suggestive of bacterial endocarditis. However, splenomegaly is absent and blood cultures are persistently negative in cases of myxoma.

The course of the disease is influenced primarily by the size of the tumour and its growth rate and to a lesser extent by the length of the pedicle. Sudden death may be caused by acute occlusion of the mitral orifice or pulmonary veins.

The electrocardiogram shows mainly P waves of increased height and duration and right ventricular strain as evidenced by RS-T segment depression and inversion of T waves. It should be noted, however, that the latter features may be due to digitalis. Occasionally, arrhythmias may occur, either auricular paroxysmal tachycardia, auricular flutter or fibrillation, premature contractions or bundle branch blocks. Thus, the electrocardiographic changes are of no value in making the diagnosis.

Roentgenographic examination of the heart is likewise of no significant value, showing an increase of cardiac shadow or a left auricular hypertrophy, right ventricular hypertrophy and not infrequently increase in pulmonary arteries. Of considerable interest, however, is angiocardiography, often confirming the presence of the tumour. In antero-posterior and lateral projections, filling defects are seen in an enlarged auricle. Recent studies[7] suggest that it would be possible to differentiate tumour from intra-auricular thrombosis by the greater size, relatively more rounded shape, well-marked contour and homogeneous aspect of the former. Such features at least are not seen in rheumatic mitral stenosis. Cardiac catheterization is of diagnostic aid in the case of right auricular myxoma, revealing high pressure in the auricle. In our third case, normal pressures were found in the right ventricle as well as pulmonary capillaries, but the pressure was significantly increased in the right auricle. From this finding, the diagnosis was suspected and subsequently confirmed by angiocardiography.

Finally, it is important to note that in a patient with isolated tricuspid stenosis, the possibility of a right auricular myxoma must first be examined, for this valve is rarely involved by the rheumatic process.

TABLE I.

Case and sex	*Age*	*Clinical aspect*	*B.P.*	*Venous pressure*	*Angiocardiography*	*Cardiac catheterization*	*Operative findings*	*Other findings*
1. F.	43	Mitral stenosis? Interauricular septal defect. Chronic cor pulmonale.	110/60 to 120/70	Increased	Right-to-left shunt	Capillary pressure? Right ventricular, Right auricular, Pulm. arterial hypertension.	No mitral stenosis.	No rheumatic fever. Intra-parietal myxomas of the right auricle and pulmonary circulation.
2. M.	52	Mitral regurgitation. Left heart failure. Subacute bacterial endocarditis?	100/60	Normal				Angina pectoris—Myocardial infarct. Myxoma left auricle. Visceral myxomatous embolism. Sudden death. No rheumatic fever.
3. M.	39	Right heart failure. Tumour of the right auricle.	100/60 to 120/80	Increased	Filling defect of the right auricle.	Right auricular hypertension.	Removal of a right auricular myxoma.	No rheumatic fever.
4. M.	41	Mitral stenosis.	110/60	Normal		Compatible with mitral stenosis.	For commissurotomy. No mitral stenosis.	Myxoma of the left auricle. Rheumatic fever: very doubtful.

CASE REPORTS

Case 1: C.O., a 43-year-old woman, had a post-partum cardiac decompensation with a syndrome resembling mitral stenosis. No history of rheumatic fever. Positive Wassermann reaction. The course of the disease was marked by intermittent periods of improvement and relapse of right heart failure. Signs on auscultation were compatible with mitral stenosis.

Radiologically, increase of cardiac shadow; left auricle normal in size; right ventricle ++; right auricle +; pulmonary arteries dilated and pulsatile. Interauricular septal defect? E.C.G.: right ventricular hypertrophy ++; right auricular hypertrophy. Digitalis waves. B.P.: 120/70-110/60 mm. Hg. Venous pressure: 22.5 cm.-24.5-25. Cardiac catheterization: pulmonary arterial pressure 72; right ventricle pressure 50; right auricle 15; capillary pressures cannot be measured. Angiocardiography: right-to-left-shunt.

Surgical exploration: no mitral stenosis. Postoperative death by ventricular fibrillation.

Autopsy findings: left auricle and ventricle normal; right auricle and ventricle strikingly dilated and hypertrophied. Myxomatous tumours were found, totally included in the wall of the right auricle and pulmonary arteries. Myxomatous nodules were disseminated in the pulmonary vessels, without infarction. It should be noted that these findings are exceedingly rare. Interauricular septal defect was present. Real difficulty in diagnosis was encountered because of the exceptional site of the tumour, producing no true obstruction. This unusual process, because of its peculiar character, namely site and metastatic dissemination, was responsible clinically for chronic cor pulmonale.

Case 2: N.Y., a 52-year-old man, had angina pectoris followed by myocardial infarction; no previous history of rheumatic fever. There were clinical signs of mitral regurgitation complicated by left heart failure. Radiologically, the heart shadow was increased, the left auricle slightly enlarged, and both ventricles were hypertrophied. E.C.G.: right ventricular hypertrophy. B.P.: 100/60. Venous pressure: 5–8–4.5 cm. In the course of the disease, there was prolonged hyperthermia with cerebral embolism manifested by left hemiplegia and without splenomegaly. Subacute bacterial endocarditis? Sudden death.

Autopsy: dilated heart. The enlarged left auricle contained a pedunculated myxoma. There was considerable infarction of the posterior wall of the left ventricle. Coronary arteries: arteriosclerosis +++. Multiple visceral myxomatous embolism with infarcts to the kidneys, spleen and right temporal lobe (sylvian artery).

Case 3: B.V., a 39-year-old man. No rheumatic fever. Clinical signs of right heart failure. Radiologically, right ventricular hypertrophy +++; right auricular hypertrophy ++. E.C.G.: Right ventricular and auricular hypertrophy, complete right bundle branch block. Digitalis waves. BP: 120/80-100/60. Cardiac catheterization: right auricular as well as venous hypertension. Capillary pressure 1; pulm. arteries 11; right ventricle 8; right auricle 13; superior vena cava 18; venous pressure 26–33–23 cm. Angiocardiography: filling defect of the right auricle.

Subsequently, there were attacks of paroxysmal dyspnœa with cyanosis when the patient bent forward to take up something. Venous pressure before an attack 13.5–18.5–14.5; during an attack, it increased to 32–35.5–34.

Operative and post-mortem findings: right auricular myxoma. Postoperative death. This patient presented the characteristic features of a right auricular tumour as revealed by cardiac catheterization and angiocardiography.

Case 4: H.R., a 41-year-old man, hospitalized for mitral stenosis complicated by congestive heart failure. However, the rheumatic fever history was very doubtful. Signs on auscultation were compatible with mitral stenosis. Radiologically, both ventricles were hypertrophied; left auricle ++.

E.C.G.: Right ventricular hypertrophy, bilateral auricular hypertrophy, incomplete right bundle branch block. B.P.: 110/60. Normal venous pressure. Cardiac catheterization showed pressures compatible with mitral stenosis. Capillary pressure 35; pulm. arteries 54; right ventricle 27.5; right auricle 3.5. Thoracotomy for commissurotomy; no mitral stenosis, but left auricular myxoma found. Clinically, the most evident diagnosis was mitral stenosis.

SUMMARY

The diagnosis of auricular myxoma is facilitated by the following findings:

1. Mitral stenosis without rheumatic etiology.

2. Symptomatic variability with postural changes. Because of this well-recognized fact, it would be of interest to examine the patient in different positions, especially by auscultation and electrocardiography, and possibly so to observe suggestive differences.

3. Progressively severe heart failure not improved by the usual treatment.

4. Confirmatory findings at angiocardiography and cardiac catheterization. As these tumours are pedunculated and benign, their surgical removal is indicated and rational. Although patients operated on so far have not survived because of technical or organic difficulties, nevertheless it is hoped that with increasing progress in cardiac surgery, particularly with use of artificial heart and lungs, myxomata will soon be radically cured by early operation.

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RÉSUMÉ

Les myxomes auriculaires sont des tumeurs bénignes, pédiculées, siègeant plus souvent dans l'oreillette gauche. Ils entraînent une obstruction auriculo-ventriculaire, se manifestant d'abord par le syndrôme du rétrécissement mitral sans antécédents rhumatismaux puis par une insuffisance cardiaque progressivement sévère et pouvant s'accompagner d'accidents emboliques. L'évolution est irrémédiable et fatale, se terminant souvent par la mort subite.

Quoique peu fréquents, ils sont de plus en plus observés, grâce aux nouvelles méthodes qui en facilitent le diagnostic: angiocardiographie et cathétérisme cardiaque. Nous en rapportons quatre cas: deux de l'oreillette gauche, deux de l'oreillette droite.

Le traitement chirurgical s'impose malgré les échecs actuels car les progrès techniques de la chirurgie cardiaque offrent les plus grandes possibilités pour leur cure radicale. E.P.

MEDITERRANEAN ANÆMIA IN CHINESE CANADIANS*

J. K. SIDDOO, M.D.,† C. J. COADY, M.D.,‡ L. MORGAN-DEAN, R.T.,¶ and W. H. PERRY, M.D., M.R.C.P.(E.), F.R.C.P.[C.],§ *Vancouver*

IT IS THE PURPOSE of this communication to present a study of four Chinese families including three cases of Mediterranean anæmia and eleven persons with the trait.

*From the Hæmatology Laboratory, Department of Pathology, Vancouver General Hospital, and the Department of Medicine, University of British Columbia.
†Associate Resident in Pathology, Vancouver General Hospital.
‡Resident in Pathology, Vancouver General Hospital.
¶Chief Technician, Hæmatology Laboratory, Vancouver General Hospital.
§Hæmatologist, Vancouver General Hospital; Asst. Prof. of Medicine, University of British Columbia.

Criteria for the diagnosis of Mediterranean anæmia were divided into essential and supplementary. The essential criteria were marked anæmia, morphological abnormalities of red cells with target cells, microcytes, poikilocytes and hypochromia, demonstration of the trait in other members of the family, increased resistance of red cells to hæmolysis in hypotonic saline, and refractoriness of the anæmia to iron therapy. The supplementary criteria included values of fetal hæmoglobin above 2%, the absence of other abnormal hæmoglobins demonstrable by paper electrophoresis, reticulocytosis in excess of 1.5%, the presence of nucleated red cells in the circulating blood and, finally, clinical evidence of pallor, periodic attacks of fever, splenomegaly and hepatomegaly.