MEDICAL GRAND ROUNDS

NOVEMBER 20, 1958

MYXOMA OF THE LEFT ATRIUM

CASE 1.

A 50-YEAR-OLD WHITE WOMAN WAS ADMITTED FOR THE FIRST TIME ON [redacted] 1958 FOR EVALUATION OF CARDIAC STATUS.

SHE WAS TOLD THAT SHE HAD HAD AN OBSCURE FEBRILE DISEASE IN CHILDHOOD BUT WAS PERFECTLY WELL UNTIL [redacted] 1957, WHEN SHE DEVELOPED COUGH AND FEVER TO 103 DEGREES. SHE RECOVERED IN ABOUT 2 WEEKS BUT SORE THROAT AND EASY FATIGABILITY PERSISTED. IN [redacted] 1957 A NONPRODUCTIVE COUGH, MADE WORSE BY LYING DOWN, WAS NOTED. DYSPNEA ALSO APPEARED WHEN SHE LAY FLAT IN BED. EXAMINATION AT THE TIME SHOWED TACHYCARDIA AND "CLOUDINESS AT THE BASES OF THE LUNGS." SHE IMPROVED SOMEWHAT WITH THE PASSAGE OF TIME BUT IN [redacted], 1958 CARDIAC ENLARGEMENT AND BASAL RALES WERE DETECTED. TREATMENT WITH DIGITALIS AND LOW SODIUM DIET RESULTED IN NO DISCERNIBLE BENEFIT. HEMOPTYSIS AND PERIPHERAL EDEMA WERE DENIED AT THIS TIME. CAREFUL EXAMINATION AT THE TIME SHOWED THE FOLLOWING:

PULSE RATE 120
BP 94/80
NO VENOUS DISTENSION
MODERATE CARDIOMEGALY
MARKED ACCENTUATION OF P_2
LOUD M-1
THIRD HEART SOUND AT APEX
FAINT AORTIC SYSTOLIC MURMUR
MOIST RALES AT BOTH BASES
NO HEPATOMEGALY OR PERIPHERAL EDEMA
ECG: NORMAL AXIS, DIGITALIS EFFECT, AND PROLONGED PR INTERVAL.

THE PHYSICIAN'S DIAGNOSES AT THIS TIME INCLUDED LEFT ATRIAL MYXOMA AND MITRAL STENOSIS.

ON ADMISSION TO [redacted], PHYSICAL FINDINGS WERE UNCHANGED EXCEPT THAT AN APICAL PRESYSTOLIC RUMBLE WAS EASILY AUDIBLE. CHEST FILMS SHOWED CARDIOMEGALY AND PULMONARY CONGESTION.

CARDIAC CATHETERIZATION ON [redacted], 1958, SHOWED MARKED PULMONARY HYPERTENSION WITH A LA-LV DIASTOLIC GRADIENT OF 24 MM. HG. THERE WAS NO EVIDENCE OF LV FAILURE OR SHUNTS OF ANY KIND. RA PRESSURE WAS 0. CARDIAC OUTPUT WAS 3.56 L./MIN. NEAR THE CONCLUSION OF THE PROCEDURE, SHE SUDDENLY BECAME COMATOSE AND SHORTLY THEREAFTER SHOWED A

RIGHT HEMIPARESIS AND APHASIA. THE HEMIPARESIS IMPROVED OVER THE ENSUING WEEK BUT THE APHASIA WAS STILL RATHER MARKED AT DISCHARGE (8 DAYS AFTER ADMISSION).

SHE WAS READMITTED ON [redacted] 1958, AT WHICH TIME THE APHASIA AND RIGHT HEMIPARESIS HAD ALMOST DISAPPEARED. THE CARDIAC FINDINGS WERE UNCHANGED. SHE COMPLAINED OF PAIN AND STIFFNESS IN THE RIGHT HAND AND WAS MILDLY FEBRILE. SEDIMENTATION RATE WAS 40-50 MM. REPEATED BLOOD CULTURES WERE NEGATIVE. CARDIAC FILMS SHOWED BIVENTRICULAR ENLARGEMENT AND PROMINENT PULMONARY ARTERY SEGMENT. ECG SHOWED TENDENCY TO RAD AND DIGITALIS EFFECT.

OPERATION WAS POSTPONED UNTIL [redacted] BECAUSE OF THE POSSIBILITY OF ACTIVE RHEUMATIC FEVER. AT SURGERY, "A LARGE MASS, NEARLY FILLING THE LEFT ATRIUM, WAS ENCOUNTERED. THIS WAS SOFT AND GELATINOUS AND /AROSE/ FROM A 2 CM. ... PEDICLE IN THE VICINITY OF THE FORAMINAL VALLEY...." IT PARTLY OCCLUDED THE RIGHT PULMONARY VEINS AND ALMOST COMPLETELY OBSTRUCTED THE MITRAL ORIFICE.

THE TUMOR WAS REMOVED BUT SEVERE BLOOD LOSS WAS ENCOUNTERED IN THE PROCESS. ALTHOUGH THE BLOOD WAS RAPIDLY REPLACED AND OTHER SUPPORTIVE MEASURES INSTITUTED, RESTORATION OF ARTERIAL BLOOD PRESSURE COULD NOT BE SATISFACTORILY ACCOMPLISHED. SHE EXPIRED 12 HOURS POST-OPERATIVELY.

CASE II.

A 37-YEAR-OLD WHITE WOMAN WAS ADMITTED ON [redacted], 1958, COMPLAINING OF SHORTNESS OF BREATH, CHEST PAIN, AND ANKLE SWELLING.

IN [redacted] 1958, PRECORDIAL PAIN RADIATING TO LEFT ARM AND UNASSOCIATED WITH STRESS APPEARED. SHE WAS FOUND TO HAVE SEVERE ANEMIA (7.0 GMS. HEMOGLOBIN) AND WHAT WAS THOUGHT TO BE A HEMIC MURMUR. IN [redacted] 1958, RAPID HEART BEAT, SEVERE DYSPNEA AT REST, AND HEMOPTYSIS APPEARED (GRADUALLY). ANKLE EDEMA BECAME WORSE. DIGITALIS AND DIURETICS PRODUCED SOME IMPROVEMENT. SHE ALSO DESCRIBED "CHOKING UP" WHENEVER SHE LAY ON HER RIGHT SIDE. SHE WAS FINALLY REFERRED TO DR. HUGH WILSON AS A POSSIBLE CANDIDATE FOR CARDIAC SURGERY.

ON ADMISSION, PULSE RATE WAS REGULAR AT 90 AND BP WAS 100/70. TEMPERATURE WAS NORMAL. THE PATIENT WAS COMFORTABLE BUT SEEMED SLIGHTLY DUSKY. THE NECK VEINS WERE MODERATELY DISTENDED. THE LUNGS WERE CLEAR. THE HEART WAS MODERATELY ENLARGED AND P2 WAS MARKEDLY ACCENTUATED. AN ATYPICAL MID-DIASTOLIC APICAL RUMBLE WAS AUDIBLE. THE LIVER WAS MODERATELY ENLARGED AND TENDER. THERE WAS MARKED PITTING EDEMA FROM FEET TO KNEES. WHEN THE PATIENT WAS PUT ON HER LEFT SIDE (FLAT), SHE RAPIDLY BECAME CYANOTIC AND DYSPNEIC. PULSE RATE INCREASED DRAMATICALLY AND THE MANEUVER WAS ABANDONED AFTER 20-30 SECONDS.

ECG SHOWED RAD, DIGITALIS EFFECT, AND BROAD, FLAT P WAVES. CARDIAC FILMS SHOWED LA ENLARGEMENT.

RIGHT HEART CATHETERIZATION ON [redacted] SHOWED PULMONARY HYPERTENSION (82/43) AND A PA WEDGE PRESSURE OF 30 MM. HG. THERE WAS NO ARTERIAL DESATURATION BUT CARDIAC OUTPUT WAS VERY LOW (1.6 L./MIN.). RA MEAN PRESSURE WAS 11.4 MM. HG. CINEANGIOCARDIOGRAPHY SUGGESTED BLOCKAGE TO LA OUTFLOW BUT DID NOT CLEARLY DEMONSTRATE A TUMOR MASS IN THE CHAMBER.

THE PATIENT WAS THOUGHT, NEVERTHELESS, TO HAVE A LEFT ATRIAL TUMOR AND ATTEMPTS TO REDUCE EDEMA WERE INSTITUTED PRIOR TO OPEN-HEART SURGERY. ON [redacted] SHE COMPLAINED OF MALAISE AND WORSENING DYSPNEA. ON [redacted], SHE SUDDENLY BECAME EXTREMELY DYSPNEIC AND CYANOTIC. IN SPITE OF VIGOROUS TREATMENT WITH OXYGEN AND OTHER ADJUNCTS, THE HEART STOPPED AND COULD NOT BE RESTARTED.

AUTOPSY CONFIRMED THE PRESENCE OF A LARGE LEFT ATRIAL MYXOMA.

DISCUSSION

THE TRADITIONAL VIEW THAT PRIMARY TUMORS OF THE HEART ARE UNDIAGNOSABLE DURING LIFE HAS GONE HAND-IN-HAND WITH THE FACT THAT DEFINITIVE TREATMENT WAS, UNTIL RECENTLY, IMPOSSIBLE. WITH THE ADVENT OF CARDIAC SURGERY AND THE INTRODUCTION OF OPEN-HEART TECHNIQUES, INTEREST IN THE DIAGNOSTIC ASPECTS OF INTRACARDIAC (PRIMARY) TUMORS HAS BEEN REKINDLED.

THE INTRACARDIAC TUMOR WHICH IS OF GREATEST CLINICAL INTEREST IS THAT WHICH WAS FOUND IN THE 2 CASES JUST PRESENTED. AT LEAST HALF OF THE REPORTED PRIMARY TUMORS OF THE HEART WERE MYXOMAS (PRICHARD) AND OF THESE, 75 PER CENT WERE IN THE LEFT ATRIUM. MOST ARE POLYPOID AND ARE ATTACHED TO OR OVERLIE THE FOSSA OVALIS (MAHAIM). THEY MAY, BY VIRTUE OF THEIR LOCATION, OBSTRUCT LEFT ATRIAL OUTFLOW OR PULMONARY VENOUS INFLOW OR BOTH. FURTHER, SINCE THEY ARE POLYPOID, THEY MAY PRODUCE SUCH MECHANICAL INTERFERENCE INTERMITTENTLY. ANOTHER FEATURE OF IMPORTANCE IS THEIR FREQUENT ASSOCIATION WITH EMBOLI. IT HAS BEEN HELD, IN FACT, THAT THE INTRACARDIAC MYXOMAS ARE NOT TRUE PRIMARY TUMORS BUT ARE PROBABLY THROMBOTIC IN ORIGIN. THE PREVAILING VIEW SEEMS TO BE THAT THEY ARE TRUE CONNECTIVE TISSUE TUMORS, RARELY MALIGNANT, AND BECOME MYXOID BECAUSE OF THEIR CONSTANT MOVEMENT IN THE LEFT ATRIUM. IN ANY EVENT, THEY MAY BECOME VERY FRIABLE, OR MAY BE OVERLAIN BY FRIABLE CLOT, SO THAT RELEASE OF EMBOLI IS NOT UNCOMMON.

FROM THE ABOVE, CERTAIN REASONABLY CHARACTERISTIC CLINICAL FEATURES CAN BE PREDICTED AND ARE SUBSTANTIATED IN THE LITERATURE (DOANE AND PRESSMAN, KROOPF AND PETERSON, PANSCH, FIELD ET AL, WEINSTEIN AND ARATA, LIKOFF ET AL):

1. BLOCKAGE TO LEFT ATRIAL OUTFLOW, PRODUCING CHARACTERISTIC SIGNS AND SYMPTOMS OF MITRAL STENOSIS WITH OR WITHOUT INSUFFICIENCY.

2. EMBOLIC PHENOMENA, OF ANY TYPE, INCLUDING THAT CHARACTERIZED BY MYRIADS OF TINY EMBOLI PRODUCING GENERAL SYSTEMIC SYMPTOMS BUT NOT SPECIFIC SIGNS AS DO LARGER SINGLE EMBOLI TO THE BRAIN, KIDNEYS, ETC.

FOR THESE REASONS, MANY CASES HAVE BEEN THOUGHT TO HAVE RHEUMATIC MITRAL DISEASE WITH SUPERIMPOSED ACTIVE RHEUMATIC FEVER OR SUBACUTE BACTERIAL ENDOCARDITIS. OF GREATEST IMPORTANCE, CLINICAL EXPERIENCE ALSO SHOWS THAT MOST SUCH PATIENTS HAVE DEVELOPED SIGNS OF BLOCKAGE TO LEFT ATRIAL OUTFLOW VERY RAPIDLY, WITHOUT ANTECEDENT HEART DISEASE OR SUBSTANTIABLE RHEUMATIC FEVER. TREATMENT WITH DIGITALIS AND DIURETICS HAS BEEN NOTORIOUSLY INEFFECTIVE. EXACERBATION OF ACUTE SIGNS AS A RESULT OF CHANGE IN BODY POSITION HAS BEEN AN UNRELIABLE GUIDE TO DIAGNOSIS. IN SOME CASES (KROOPF AND PETERSON) ANTERIOR CHEST PAIN WAS ONE OF THE PRESENTING COMPLAINTS. OTHER FEATURES ARE THE RAPID DEVELOPMENT OF RIGHT-SIDED ECG PATTERNS AND A-V CONDUCTION DISTURBANCES AND X-RAY SIGNS OF LEFT ATRIAL AND RIGHT VENTRICULAR ENLARGEMENT. ANGIOCARDIOGRAPHY HAS ALSO YIELDED DIAGNOSTIC INFORMATION.

IT IS AXIOMATIC THAT THE ONLY LOGICAL TREATMENT OF MECHANICAL BLOCKAGE TO FLOW THROUGH THE LEFT ATRIUM IS BY MECHANICAL MEANS. THIS DICTUM APPLIES AT THE PRESENT TIME TO BLOCKAGE AS THE RESULT OF LEFT ATRIAL MYXOMAS AS WELL AS TO THAT DUE TO MITRAL STENOSIS. THE RAPID DEVELOPMENT, IN PREVIOUSLY WELL PATIENTS, OF SIGNS AND SYMPTOMS SUGGESTIVE OF BLOCKAGE TO LEFT ATRIAL OUTFLOW, ESPECIALLY IF ASSOCIATED WITH EMBOLIC PHENOMENA WITH OR WITHOUT FEVER, SHOULD SUGGEST LEFT ATRIAL MYXOMA.

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