CASE REPORTS

EPILEPTIFORM ATTACKS DUE TO MYXOMA OF THE RIGHT AURICLE

BY

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Attacks of dizziness and syncope have been relatively common features in the recorded cases of primary intracardiac tumour and of intracardiac ball thrombus (Wood, 1814; von Recklinghausen, 1885; Schwarz and Biloon, 1931; Gorlitzer, 1934; Elson, 1934; Fawcett and Ward, 1939; Garvin, 1941; Wright, Flynn, and Druet, 1944; Burnett and Davidson, 1945; Evans and Benson, 1948). Actual epileptiform convulsions have been mentioned only by McLeod (1883) and Young and Hunter (1947).

The patient described here was first seen by one of us because she had had a number of generalized fits occurring during the course of a prolonged and unexplained febrile illness.

Case Report

A married woman, aged 52 years, was first seen on March 4, 1948, having had four or five epileptic attacks. At the age of six years she had an attack of rheumatic fever, and for a few years she suffered from intermittent joint pains. There was nothing to suggest that there was involvement of the heart at the time. She subsequently married and had two uneventful pregnancies. In 1937 glycosuria was found and she was treated as a diabetic by diet only. In the same year bilateral lens extractions were performed for cataract.

The present illness started in November, 1946, with attacks of fever and rigors, each attack lasting a few days followed by complete recovery. During these attacks there was some shortness of breath and palpitation, and in some there had been a complaint of left mammary pain radiating down the left arm, unrelated to exertion. She was admitted to St. Luke's Hospital, Guildford, in January, 1947. She had intermittent fever to about 101° F. and a persistent tachycardia (110-130). There was no clinical evidence of a heart lesion. Blood culture was negative. There was a mild microcytic anæmia with a polymorphonuclear leucocytosis varying between 17,000 and 40,000 per c.mm. The radiological appearance of the heart was said to suggest mitral stenosis. The lungs were normal. The Wassermann reaction was negative. There was then no evidence of diabetes. She was discharged at the end of five weeks at the request of her relatives.

She was admitted to the Royal Masonic Hospital in July, 1947, under the care of Sir Horace Evans, having had an attack of pleurisy with blood-stained sputum the previous month. On examination then, she appeared thin and sallow. There was no cardiac enlargement but a short presystolic bruit was heard. The spleen was palpable. There was a secondary anæmia with a white cell count that varied between 3000 and 12,000 per c.mm. Blood cultures were negative. The blood Wassermann and Kahn tests were positive. X-rays of the chest showed evidence of infarction of both lungs. A diagnosis of mitral stenosis with bacterial endocarditis was made and she was given penicillin two million units daily for a long period without material change in her condition. After returning home her condition remained much the same and she continued to have intermittent fever.

In February, 1948, she had a generalized epileptic attack without warning, which was not followed by any focal symptoms or signs. Further similar attacks occurred in the same month. She was seen by one of us for the first time in March, 1948, and appeared moderately wasted with a generalized *café-au-lait* tint to the skin. In addition there was some brownish pigmentation of

the forearms, abdomen, and hard palate. The pulse rate was 90 a minute, temperature normal and blood pressure 86/60. No cardiac abnormality was observed and the spleen was not palpable. Blood count was normal apart from a mild microcytic anæmia and blood culture was negative. The urine contained a moderate amount of pus with a growth of *B. coli* on culture. In view of the hypotension, pigmentation, and past history suggestive of endocarditis, it was considered possible that the suprarenal glands had been damaged by emboli with consequent Addison's disease. The administration of D.O.C.A. ten milligrams daily raised the blood pressure to a maximum of 116/85, but resulted in widespread œdema and was discontinued. In April, 1948, a systolic murmur was heard over the whole præcordium with a mid-diastolic murmur to the left of the sternum. Fluoroscopy showed slight backward displacement of the barium in the right oblique view; the right auricle was prominent in the anterior view. Three days later the diastolic murmur was no longer audible and a rough systolic thrill was felt. The presence of dyspnœa with an enlarged tender liver was taken to indicate cardiac failure but there was no engorgement of the neck veins or œdema. At this time the urinary infection became severe but responded to sulphonamide therapy in about three weeks, and the patient was discharged in June feeling fairly well but having occasional fever up to 101° F.

FIG. 1.

FIG. 2.

FIG. 1. and 2.—Gangrene of nose and symmetrical gangrene of feet.

She was re-admitted in July, 1948, complaining of increasing weakness and occasional shivering attacks. The physical signs were unchanged and there was an intermittent pyrexia to 102° F. The urine was normal. In spite of further negative blood cultures, it was considered that active endocarditis was present and a course of penicillin therapy was commenced, 125,000 units three-hourly. This was continued for thirty-eight days, the patient being afebrile for the last eighteen days. She was discharged on September 4 and re-admitted on November 11. She had been fairly well until four weeks before admission when she complained of frontal and occipital headache, with intermittent severe abdominal pain of sudden onset, associated with precipitate defæcation. These symptoms occurred about four times daily. Examination showed an extreme degree of emaciation with generalized brownish pigmentation of the skin and a malar flush. There was no cyanosis. The apical first sound had a snapping quality; there was a loud diffuse systolic murmur and a soft diastolic murmur to the left of the sternum, the latter audible in the erect posture only.

Two days later she was found to have a deep purplish discoloration of the tip of the nose, fingers, and feet with some purpuric patches at the elbows. Her temperature was 95° F. and her pulse rate 80 a minute, the pulse being almost imperceptible at the wrist. The diastolic murmur was audible in any posture. A diagnosis of ball thrombus in the left auricle was suggested at this stage by the variable physical signs, the clinical diagnosis of mitral disease, and the symmetrical peripheral gangrene.

During the next few days there were frequent attacks of auricular flutter with two to one block, each attack lasting a few minutes and not giving rise to any new subjective disturbance. The patient's condition became slowly worse without the development of any signs of congestive heart failure, and she died seven days after the onset of gangrene.

FIG. 3.—Cardiac myxoma exposed *in situ* showing origin from the inter-auricular septum, and the free end of the tumour lying in the tricuspid orifice.

Necropsy. There was evidence of infarction of the bases of both lungs. The pericardium was normal. The right auricle was considerably enlarged and the left ventricle unusually small. A large solid mass could be felt through the right auricular wall, and on opening this chamber a firm greyish-yellow mass was found arising from the inter-auricular septum in the region of the fossa ovalis and extending down into the tricuspid orifice. The latter was dilated, the surfaces of the valves were roughened and the points of contact of the mass with the valves could be clearly seen. The mitral, pulmonary, and aortic valves were normal and there was no evidence of recent or old endo-

carditis. The liver was enlarged and smooth and showed marked "nutmeg" change. The spleen was enlarged but appeared of normal texture.

Macroscopically, the intracardiac mass appeared on section to consist of a firm grey mucinous and semi-gelatinous substance with a slightly laminated organized thrombus covering the surface with some small discrete hæmorrhages.

Microscopically the tumour consisted of areas of spindle cells interspersed with a few stellate cells in a mucinous matrix, with externally, a firmly attached organized and partly necrotic thrombus. The myxomatous part of the tumour and the thrombus merged into one another with no dividing line. The vessels were very thin with single endothelial lining and there was considerable round cell and polymorphonuclear perivascular infiltration with phagocytic ingestion of hæmosiderin.

Several blocks were made, sections from one of which stained with the specific muci-carmines stained well, the others having much less affinity for this dye on account of the intense necrosis present. For the same reason Weigert's elastic stain demonstrated only a few fibres, usually abundant in the endocardial myxomata.

The tumour had, therefore, the histology and staining reactions of a myxoma associated with an intimately fused necrotic thrombus.

DISCUSSION

This patient had an illness of five years duration, characterized by progressive wasting, and during the last two years by an intermittent pyrexia, hæmoptysis, palpitation, convulsions, and finally peripheral gangrene. In addition, there was a past history of rheumatic fever and glycosuria. For a time the diagnosis of mitral stenosis with an added endocarditis seemed quite clear, but the course of the illness and the persistently negative blood cultures ultimately made this diagnosis doubtful. Finally, the occurrence of symmetrical peripheral gangrene led to the suggestion that the supposed mitral stenosis was complicated by the presence of a ball thrombus in the left auricle. This seemed at the time to account for many of the unusual features of the illness, in particular the fits, the variable cardiac murmurs, and the intermittent enlargement of the liver and spleen.

The finding of a tumour in the right auricle without any evidence of mitral disease or of old endocarditis increased the difficulty of explaining certain of the symptoms and signs. It must be concluded that the fits were due to cerebral anoxia following temporary impaction of the tumour in the tricuspid orifice. (In theory they might have been associated with paroxysmal auricular flutter but there was no change in the pulse rate to suggest this during the two fits that were observed in hospital). The attacks of dyspnœa and the final gangrene could be explained in a similar way. The hæmoptyses could be explained by the occurrence of pulmonary infarction by detached fragments of tumour or its covering thrombus. The fever, leucocytosis, and the response to penicillin remained to be explained. The presence of a necrotic thrombus covering the tumour suggests that there had been in fact an endocarditis confined to the surface of the tumour, and that this infection had finally been controlled by chemotherapy.

There has been considerable doubt in the past as to the existence of cardiac tumours. Some have regarded them as organized thrombi. It is, however, now generally accepted that these masses have a neoplastic origin. (Welch, 1899; Ribbert, 1924; Yater, 1931; Fawcett and Ward, 1939).

The diagnosis of this condition during life is clearly very difficult, both on account of its rarity and of the great variability in the clinical manifestations. It is to be expected that symptoms directly attributable to the presence of an intra-cardiac mass would be very similar whether the mass is a myxoma or a ball (mass) thrombus. Evans and Benson (1948) reviewed the symptomatology of these two conditions, and pointed out the difficulty of dissociating the symptoms of the thrombus itself from those of the mitral stenosis that accompanies the condition. Variability of the clinical picture would seem to be the feature of real value, and the occurrence of intermittent congestive heart failure with perhaps syncopal attacks and peripheral cyanosis or gangrene should suggest

intermittent obstruction of the mitral or tricuspid orifices. Yater (1931) stated that every patient diagnosed as having tricuspid stenosis should be suspected of having a cardiac tumour, giving as his reason that "inflammatory stenosis of that orifice is exceedingly rare." This should perhaps be qualified by adding that evidence of mitral disease must be absent.

It was remarkable that the presence of a tumour or thrombus which almost completely obstructed the passage of blood through the heart was compatible with the continuation of life over quite a long period. Strouse (1938) described a patient who died in 1935 from myxoma of the right auricle the symptoms of which had been present for 43 years. The patient described by Macoun (1949) lived for four years and the patient here described lived for five years. In the latter it must be concluded that the obstruction of the tricuspid orifice was intermittent in the cardiac cycle. Since the main filling of the ventricle occurs during diastole it is probable that the tumour only obstructed the orifice during auricular systole.

The diagnosis of this condition during life is of great interest but the value of such a diagnosis is likely to remain essentially academic, since it is unlikely that any steps could be taken to remove an intra-auricular tumour; the comment of Burns (1809) is still mainly true: "if blended with other diseases its presence can rarely before death be ascertained; and if discovered it would not alter the plan of treatment."

SUMMARY

The clinical course is described of a patient who ultimately died of a myxoma of the right auricle. Among the features were: epileptiform convulsions, hæmoptysis, hypotension, and peripheral gangrene. The diagnosis was complicated by the presence of fever due to an endocarditis on the surface of the tumour.

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