Case reports

British Heart Journal, 1975, 37, 94-97.

Burkitt's lymphoma presenting with heart block

T. O. Cole, Ed. B. Attah, and G. C. Onyemelukwe

From the Departments of Medicine and Pathology, University College Hospital, Ibadan, Nigeria

This case report describes a 12-year-old boy with cardiac involvement by Burkitt's lymphoma producing complete atrioventricular block which at one stage showed phénomène d'accrochage. Electrocardiographic findings are discussed and the importance of routine electrocardiogram in Burkitt's lymphoma stressed.

Burkitt's lymphoma is the commonest childhood malignancy in most of the endemic areas in tropical and subtropical Africa (Burkitt and Wright, 1970a) including Ibadan, Nigeria (Edington and Maclean, 1964). The disease is invariably extensive and multifocal and has a wide tissue and organ distribution. Cardiac involvement is not infrequent (Burkitt and Wright, 1970b). Often this consists of nodular deposits on the epicardium, most commonly the right atrium. The myocardium may be involved directly or by extension of the epicardial deposits. Tumour deposits also occasionally appear in the endocardium with consequent polypoid masses in the cardiac chambers. Despite the relative frequency and wide distribution in the heart, Burkitt's lymphoma, to our knowledge, has not been previously described as a cause of intracardiac conduction defects.

We report a case of Burkitt's lymphoma to record a rare presentation, heart block, and to discuss the probable significance of electrocardiographic changes in the diagnosis and assessment of patients with this rapidly fatal malignancy.

Case report

A 12-year-old schoolboy was admitted to University College Hospital, Ibadan with a 3-week history of exertional dyspnoea, anorexia, and a facial swelling. The swelling, which was painless, had apparently gradually increased in size. He had no associated excessive lacrimation, epistaxis, or any oral bleeding. He had palpitation and mild ankle oedema, but no other cardiovascular manifestations. Systemic inquiries revealed nothing relevant. There was no family history of heart disease.

Physical examination revealed a well-developed but slightly dyspnoeic young boy with moderate conjunctival pallor. A hard globular mass with an approximate dia-

meter of 3 cm was present between the left epicanthus and the bridge of the nose. It was clearly delineated, slightly mobile, and not tender; the skin on the surface was shiny and freely mobile. The jugular venous pressure was slightly raised. The pulse was irregular and collapsing. The blood pressure in the right arm was 115/70 mmHg. Examination of the heart revealed no enlargement. The first heart sound was soft and a grade 1 ejection systolic murmur was audible at the apex. Other significant findings were a slight degree of ankle oedema, few basal crepitations in the lung bases, and a moderately enlarged and tender liver. There was no recognizable evidence of active rheumatic fever or infective endocarditis. The neurological system was normal.

The following laboratory data were obtained: haemoglobin 6.2 g/100 ml, with film appearance compatible with iron deficiency anaemia, haemoglobin genotype AA, blood group O Rh +ve, WBC 13,500/mm³ (neutrophils 51%, eosinophils 8%, lymphocytes 28%, monocytes 12% and myelocytes 1%), ESR 56 mm/hr (Westergren). Liver function tests, blood urea, electrolytes, and bicarbonate were within normal limits. Antistreptolysin-O titre was normal and serial blood cultures revealed no growth. Chest x-ray showed slight enlargement of the heart and x-ray of the skull was reported as normal.

The initial electrocardiogram performed on the second day of admission (Fig. 1) displayed the pattern of Wenkebach conduction with varying 3:2 and 2:1 AV ratio. The pattern remained unchanged after treatment of the anaemia and heart failure with packed red blood cell transfusion augmented by 20 mg frusemide. Digitalis was not given. Dyspnoea, however, improved and the ejection systolic murmur disappeared. The patient remained on bed rest while the various investigations were being carried out.

Three days later, he began to have bouts of slow but regular heart action with occasional cannon wave in the neck associated with general discomfort. Electrocardiographic monitoring was started. Rhythm strips revealed 2:1 AV block with occasional periods of complete

FIG. 1 12-lead electrocardiogram showing atrial rate of 98 a minute and Mobitz Type I partial AV block with varying 3:2 and 2:1 conduction ratio. Shortest PR interval of the conducted beat is 0.2 sec.

FIG. 2 Continuous electrocardiogram showing complete heart block with 3:1 AV accrochage. PR or RP interval changes more rapidly when relatively prolonged.

heart block; atrial rate was 108 a minute and ventricular rate in the latter about 44 a minute. At this stage, with the maxillary mass, monocytosis, and associated varying degrees of heart block in a boy of his age, Burkitt's lymphoma was strongly suspected and arrangements for biopsy of the mass were made.

Sublingual isoprenaline 15 mg 2-hourly was prescribed. Before he could have the first dose, a seemingly persistent heart block developed. Episodic hooking of every third P with R wave before or after, occurred when the PR or RP interval was short (*phénomène d'accrochage*). The atrial rate of 120 a minute was con-

FIG. 3 Interventricular septum showing tumour deposit in AV node region.

sistently treble that of the ventricles (Fig. 2). Cannon waves, though still periodic, became more persistent and disturbing. He eventually started the sublingual isoprenaline with no significant effect. The dose was increased the next day to 30 mg 2-hourly. This further accelerated the atrial rate but had no effect on that of the ventricles. On the fifth day acceleration of the atrial rate continued but the ventricular rate gradually slowed down. At an atrial rate of 130 a minute, accrochage disappeared; ventricular rate was 28 a minute. Transvenous pacing equipment was not available. The patient progressively deteriorated despite the combination of isoprenaline and the available intensive care facilities. He died 8 days after being admitted.

Postmortem findings

The body was that of a fairly well-nourished boy. There was a 2.0 × 3.0 cm tumour mass in the left upper eyelid and a small tumour nodule in the left upper jaw gingiva above the premolar teeth. No superficial lymph node enlargement was present.

The heart weighed 229 g. The epicardial surface was glistening and there was a 3.5 × 2.0 × 2.0 cm firm grey nodular tumour deposit on the posterior surface of the right atrium and the posterior interatrial groove. The right coronary artery was not infiltrated and was patent. There was nodular infiltration of the lower right atrial wall, and along the interatrial septum and the right atrioventricular groove anterolaterally. One tumour nodule presented as a large, irregular polyp in the right atrium. There was a 1.5 × 0.8 cm tumour nodule in the region of the atrioventricular node. The myocardium in the region of tumour deposits including the AV node showed total replacement by the tumour (Fig. 3). The foramen ovale was closed. Both ventricles were of normal appearance and the valves were unremarkable. The left ventricular wall measured 1.1 cm thickness and the

right 0.3 cm. Apart from the region of the AV node, the ventricular myocardium was slightly pale and was otherwise normal. There was no evidence of myocardial infarction, fibrosis, or tumour.

The lungs were slightly oedematous and were free of tumour deposits. The liver, left kidney, gastric antrum, and second part of duodenum contained firm grey discrete tumour nodules measuring from 0.5 to 4.5 cm in diameter. The abdominal lymph nodes were enlarged and replaced by tumour. Other organs were of normal appearance. The diagnosis of Burkitt's lymphoma was made. Schistosomiasis of the urinary bladder was an incidental finding.

Microscopical examination, including histochemistry, confirmed the diagnosis of Burkitt's lymphoma involving the left upper eyelid, gingiva, heart, abdominal lymph nodes, liver, stomach, duodenum, and left kidney. The infiltrate was destructive, destroying the atrial myocardium, atrioventricular node, and other organs involved. Multiple sections from the interventricular septum and the conducting system below the AV node revealed no tumour infiltration. There was also no evidence of tumour deposit or infarction in multiple sections taken from other portions of the left ventricle. The coronary arteries showed no evidence of arteritis, atheroma, or other abnormalities.

Discussion

Primary or metastatic tumours involving the conducting system are well recognized, though relatively rare, causes of heart block (Katz and Pick, 1956; Buckberg and Fowler, 1961; Goggio, Harkness, and Palmer, 1961). The case described here indicates that Burkitt's lymphoma, which not uncommonly involves the heart, is another possible

cause of heart block. To our knowledge, this has not been previously reported.

At necropsy, the lymphoma sites included an area around the AV node. This is compatible with the electrocardiographic changes observed during his management. The initial intracardiac conduction disturbance was Mobitz Type I heart block with varying 3:2 and 2:1 AV ratio and a prolonged PR interval of the conducted beat. Later 2:1 AV ratio became persistent with PR interval of the conducted beat remaining prolonged. At this stage, he had episodes of heart block which became permanent as the atrial rate increased. It is usually impossible to differentiate between Mobitz Types I and II AV block and further, mixed forms may occur. The poor ventricular response to the increasing atrial rate and the rapid progression to complete heart block (Langendorf and Pick, 1968; Stock, 1970) strongly favour the 2:1 AV block in this patient as Mobitz Type II.

There is increasing clinical and electrocardiographic evidence that the anatomical site of block is different in Mobitz Types I and II (Langendorf and Pick, 1968; Stock, 1970; Krikler, 1971). Type I is usually the result of a lesion in the AV node; in type II the lesion is more peripherally situated in the AV conduction system and is frequently associated with fascicular block. Recent electrophysiological observations (Watanabe and Dreifus, 1967) and His bundle electrocardiographic findings by Narula and Samet (1970) support this. It seems possible, therefore, that progression in the degree of heart block in this patient could have been the result of a rapid and degenerative growth of the lymphoma from the AV node into the bundle of His. Acceleration of the atrial rate, of unknown cause, could also have been contributory.

Another interesting feature in this case was the development of 3:1 atrioventricular synchronization which came in spells (*phénomène d'accrochage*) as described by Segers, Lequime, and Denolin (1947). PP interval was 0.48 sec and RR 1.44 sec. The PR or RP interval was never longer than 0.2 sec. Fixed synchronization was not observed in this patient probably because of the persistently rapid atrial rate. As shown in Fig. 2, synchronization occurred only when the interval between the P and R waves was short. The mechanism responsible for atrioventricular synchronization is still not clear. However, there is at the moment, experimental evidence to suspect both mechanical and electrical mechanism.

The case presented confirms the possibility of cardiac involvement of Burkitt's lymphoma as a

cause of intracardiac conduction disturbances. The electrocardiographic changes observed are compatible with the sites involved. It is, therefore, reasonable to include this lymphoma in the differential diagnosis of lesions producing intracardiac conduction defects and other possible electrocardiographic changes. Early recognition and treatment of the tumour involved may revert conduction disturbances to normal (Shelburne and Aronson, 1940). Routine electrocardiograms, therefore, may be of great value in suspected or confirmed cases of Burkitt's lymphoma in respect of the recognition of cardiac involvement. If present, it may also be useful in assessing the severity and distribution in the heart of this rapidly fatal malignancy.

We are grateful to the Medical Illustration Unit, Medical School, University of Ibadan, for production of the Figures and to Mr. R. A. O. Aminu for preparation of the manuscript.

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- Requests for reprints to Dr. T. O. Cole, Department of Medicine, University College Hospital, Ibadan, Nigeria.