· 临床病理讨论 ·

Clinicopathological Conference

A 61-year-old man with thickened heart

(the 35th case)

Institute of Geriatric Cardiology, Chinese PLA General Hospital

Presentation of case

A 61-year-old man was admitted to the hospital because of chest distress and shortness of breath. His electrocardiogram (ECG) revealed complete left bundle branch block, and he was diagnosed as myocarditis, and didn't get regular treatment. Since 1986, he had occurred precordial pain on exertion generally, and relieved after a rest for 10min. He was diagnosed as coronary artery disease. After that, he had a history of 16-year chest pain, which usually responded to sublingual Suxiao Jiuxin pills. In 2002, he noted a decrease of exercise tolerance and increased fatigue. His echocardiogram revealed a "thick heart". The patient was ever admitted to a local hospital several times and treated as coronary artery disease, but the result was less satisfactory. In January 2008, he developed palpitation without motivation, accompanied with chest distress, pain, and dyspnea. These symptoms can not be relieved after buccaling Suxiao Jiuxin pills. His ECG showed atrial fibrillation and complete left bundle branch block. Digoxin administration didn't relieve the symptoms at all. On September 21, 2008, he suffered nocturnal dyspnea, cyanosis, abdominal distention and edema of lower extremities, and was brought to an emergency room at a local hospital immediately. There he was diagnosed as acute heart failure and was treated with furosemide, Lanatoside C and nitroprusside sodium, and then his conditions were improved. However, on September 28, 2008 when he was lying in a hospital bed, he had a sudden loss of consciousness for 3 — 5min and recovered without any emergency medical treatment. His ECG showed rapid atrial fibrillation with heart rate of 160/min when he was recovered. After that he suffered sudden loss of consciousness for twice. On October 14, 2008, a coronary angiography revealed that left anterior descending artery and left circumflex artery were both 20% stenosis. Then he was hospitalized with the diagnosis of cardiac dysfunction.

Being a physical worker, the patient denied any history of hypertension, hyperlipemia and diabetes mellitus. A history of alcohol abuse (> 200ml/d) in the past 40 years was reported, but he already stopped drinking for 7 years.

Physical examination: the temperature was 36.5℃, the blood pressure 128/70mmHg, the pulse 78/min, respiration rate 18/min. He was conscious and cooperative, and could answer all questions. He had light cyanosis of lips, light engorgement of jugular veins. No moist rales over both lung fields was auscultated. The heart on percussion was found to be enlarged to the left. Cardiac rhythm was irregular at 108/min. At cardiac apex, there was a grade-Ⅱ/Ⅵ soft systolic murmur. The abdomen was flat. The liver was palpated 3cm below the right costal margin and was firm without tenderness. Hepatojugular reflux was negative. There was no shifting dullness and edema of legs.

Accessory examination: the complete blood

收稿日期:2009-07-03

作者单位:100853 北京市,解放军总医院老年心血管病研究所

通讯作者:蓝云锋,Tel:15011546007

count and levels of electrolytes and glucose, as well as the results of tests of coagulation, renal function, liver function and myocardial enzymes were all within normal ranges. A chest X-ray showed an enlarged heart without pulmonary congestion. An electrocardiogram obtained after the patient's arrival showed atrial fibrillation, left bundle-branch block (Fig 1). While on June 4th, 2009, his ECG changed to atrial fibrillation and complete right bundle branch block (Fig 2). The dynamic electrocardiogram revealed atrial fibrillation and several long pauses, and the longest was 3.01s (Fig 3).

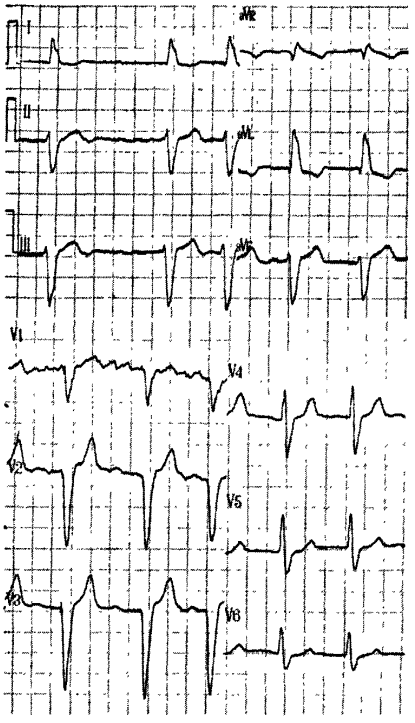

Fig 1 Atrial fibrillation, left bundle-branch block

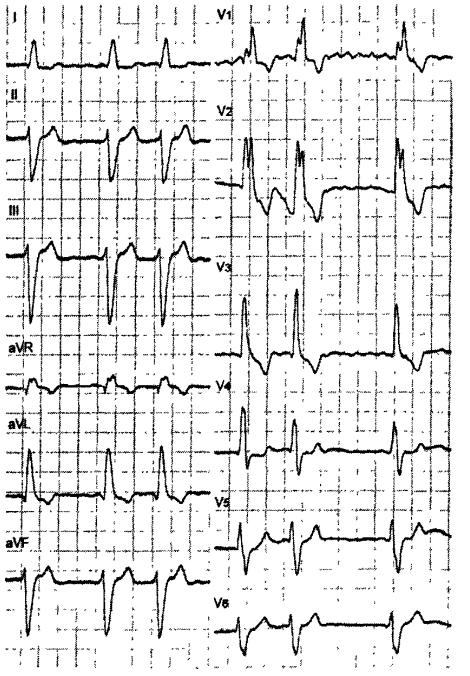

Fig 2 Atrial fibrillation, right bundle branch block

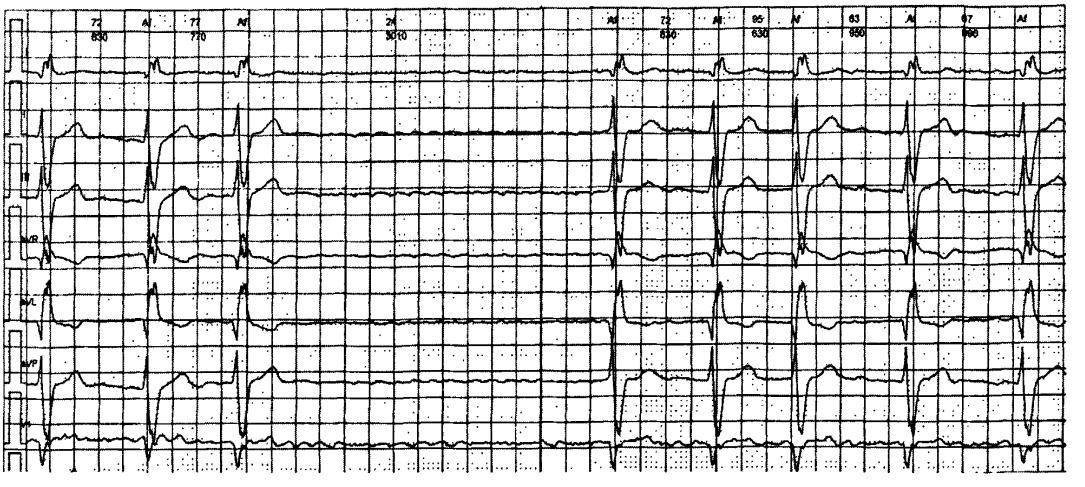

Fig 3 Long pause

A transesophageal echocardiography showed that the left ventricle appeared to be thickened, and the septum was more thick than the free wall (18mm *vs* 11mm). The ejection fraction was 67%. The aortic valve was anatomically and functionally normal while there was light mitral incompetence and mild tricuspid regnigatation. A heart MRI plain scan showed cardiac dilatation, including left ventricle, left and right atria. The left ventricle appeared to be thickened, and the part below septal membranous part was 21mm in systolic phase and 19mm in diastolic phase. There was mild left ventricular outflow obstruction.

Discussion

Dr. LAN Yunfeng: This patient is a 61 years old man, who has a long course of chronic disease, which aggravated recently. His main clinical features included: progressively worsened physical capacity, chest distress and chest pain on exertion recently, interrupted edema of lower limbs, and episode of sudden loss of consciousness. Objective detections showed: atrial fibrillation, intermittent complete left or right bundle branch block, long pause. The patient also had an important feature, the paroxysmal loss of consciousness without uncertain reason. It had high possibility of cardiogenic cause, and can not rule out the cause of transient ischemic attacks.

Dr. ZHANG Wenli: According to the patient's features, we should differenciate the diagnosis between hypertrophic cardiomyopathy (HCM) and dilated cardiomyopathy(DCM).

HCM is characterized by: idiopathic hypertrophy of the left (and sometimes right) ventricle, which is not consistent with the features of hemodynamics; heart failure due to diastolic dysfunction; ischemia even in the absence of coronary artery disease; and various arrhythmias. HCM can be generally divided into two categories: obstructive and non-obstructive. The most of this patient's clinical features support to diagnose as HCM. MRI plain scan showed that the interventricular septum was 21mm thick (normal value $<$ 11mm), accompanied with mild left ventricular outflow obstruction. These findings met the diagnosis criteria of HCM.

However, his chest X-ray and heart MRI plain scan both showed an enlarged heart, including left ventricle, left and right atria. The radiological and clinical features may confirm the diagnosis of DCM instead of HCM. The enlarged heart may be attributed to mitral incompetence and tricuspid incompetence. It may be explained by the change of hemodynamics. It was possible that this case was a HCM with DCM-like features. Besides, sustained tachycardia is also an important cause of reversible cardiac dilatation.

From the result of the echocardiography and heart MRI plain scan test, this case can be dingnosed as asymmetric septal hypertrophy accompanied with light obstruction of left ventricular outflow.

Dr. LU Caiyi: The following differential diagnosis should be considered: HCM, DCM and coronary artery disease.

HCM: The positive family history in the disease accounts for 50%, which belongs to autosomal dominant inheritance. It is characterized by dissymmetric hypertrophy of the left ventricle. Sometimes, the hypertrophy is concentric which is not consistent with the hemodynamics. It can be divided into obstructive and non-obstructive types. The latter is usually presented as apical hypertrophy. The key features of the disease include diastolic dysfunction of left ventricle, arrhythmias, angina, apopsychia, and even sudden death. The patient's condition consisted with these features, so the diagnosis of hypertrophic obstructive cardiomyopathy was obtained.

DCM: the chest X-ray and the heart MRI plain scan both reported an enlarged heart, including left ventricle, left and right atria. The echocardiogram revealed mitral incompetence and tricuspid incompetence, accompanied with atrial fibrilation. However, all auxiliary examinations didn't show left ventricular enlargement and contractile dysfunction. They didn't support the diagnosis of DCM which is characterized by dilatation of heart and decline in contractile force.

Coronary artery disease: the patient had recurrent chest distress on exertion which responded well to pimobendan. But the coronary angiography didn't reveal serious coronary artery disorder. In ECG, ischemic changes were persistent. It didn't support to diagnose the patient as coronary artery disease. In the case of HCM, parts of myocardial overgrow so thickly that it causes relative insufficiency of blood-supply. The symptoms resemble the features of coronary artery disease, so it be-

comes confusing.

Some features, such as dilatation of heart, didn't support to diagnose the patient as HCM. I think, these enlarged heart chambers may be the result of the change in hemodynamics. Persistently pachyntic left ventricle resulted in diastolic dysfunction, which induced the increase of end-diastolic pressure, followed by the increase of the pressure of atria sinistrum, pulmonary vascular bed and pulmonary arterial pressure increasing, and then the pressure load of the atria sinistrum and cor dextrum system was increased. On the one hand, an enlarged atria sinistrum increased arrhythmogenesis such as atrial fibrilation. On the other hand, a high pressure load in cor dextrum system would cause the changes such as right heart chambers enlargment and relative tricuspid incompetence. In the course of disease, the patient suffered apopsychia for several times. The following causes could be taken into consideration. Firstly, it may be caused by Adams-Stokes syndrome due to ventricular long pause, because the patient had complained intermittent complete left or right bundle branch block and long R-R interval. Secondly, atrial fibrilation would augment the excitability in sympathetic nervous system, aggravate left ventricular outflow tract obstruction, and afterwards induce the heart stroke volume sharply down and apopsychia. Thirdly, thrombus evoked by atrial fibrilation may fall off, and then would lead to transient ischemic attacks. The disease would display symptoms in nervous system immediately after the attack, but this patient is an exception.

The major risks of HCM include sudden death (50%), heart failure(25%) and endocarditis. The treatment must aim directly at these risk factors. In order to prevent sudden death, an implantable cardioverter-defibrillator or oral administration of β receptor blocker should be chosen. Because of atrial ventricular block, the patient can not take oral administration of β receptor antagonist unless a pacemaker is implanted. Furthermore, the pacemaker could improve diastolic function. In order to resolve the problem of outflow obstruction, surgical intervention or apical pacing maybe a good choice. Moreover, positive treatment to prevent endocarditis must be considered. In the daily life, the patient should be suggested to avoid exertion, especially heavy activity. As for atrial fibrilation, anticoagulant therapy is necessary.

(Translator: LAN Yunfeng)

一例61岁男性心肌增厚的病例讨论

1 病历摘要

患者王XX，61岁男性，因“反复胸闷26年，加重伴发作性晕厥9月余”于2009-5-18入院。1983年，患者在劳累时感胸闷、气短，心电图检查发现“完全性左束支传导阻滞”，当地诊断为“心肌炎”，未正规治疗。1986年以后逐渐出现劳累时心前区疼痛，每次历时十余分钟，症状轻，可自行缓解。当地医院诊断“冠心病”，因平素一般体力活动尚可，仅剧烈活动时诱发上述症状，含服速效救心丸可使症状好转，仍未正规检查治疗。2002年起，运动耐力显著下降。超声心动图发现“心肌肥厚”，并因此多次住院，按“冠心病”治疗，效果差。2008年1月，无诱因心悸，伴胸闷、胸痛和呼吸困难，含服速效救心丸无效。心电图示“心房颤动、完全性左束支传导阻滞”。口服地高辛等治疗，效果不明显。2008-9-21，病情加重，伴有夜间呼吸困难、唇绀、腹胀、下肢水肿等症状，当地医院诊断为心功能不全。经呋塞米、毛花苷C、硝普钠等治疗，症状好转。2008-9-28，卧位休息时突然意识丧失，在3～5min后自行恢复，其后心电图提示“房颤(快室率型)”。此后类似症状发作2次。2008-10-14，冠脉造影见前降支及回旋支各有一处斑块影，伴局部＜20%的局限性狭窄。为进一步诊治入院。患者平素从事体力劳动，既往无高血压、高血脂及糖尿病病史。饮酒40年(＞200ml/d)，戒酒7年。家族中无类似病患者。

查体：体温36.5℃，脉搏78次/min，呼吸18次/min，血压128/70mmHg。神志清楚，自动体位。口唇轻度发绀，颈静脉轻度充盈，双肺呼吸音清，未闻及干湿啰音。心浊音界向左扩大，心率108次/min，律不齐，心音强弱不等，心尖部闻及2/6级柔和收缩期杂音。腹部平坦，肝肋下3cm，质硬无压痛，肝颈反流征阴性；无移动性浊音，双下肢无水肿。

辅助检查：全血细胞检查、电解质、血糖、凝血功能、肾功能、肝功能、心肌酶均正常。胸片报告心影增大，无肺淤血。入院后心电图为心房颤动、完全性

左束支传导阻滞(图1;见377页)。2009-6-4心电图为心房颤动、完全性右束支传导阻滞(图2;见377页)。动态心电图报告为心房颤动,心室长间歇(最长3.01s,图3;见377页)。经食管超声心动图提示:左室肥厚,室间隔比游离壁明显增厚(18mm/11mm)。左室射血分数67%。主动脉瓣正常,二、三尖瓣轻-中度关闭不全。心脏MRI平扫结果发现心脏扩大(左室、左房和右房)。室间隔膜部下方可见显著的收缩期增厚(21mm∶19mm),左室流出道轻度受阻。

2 讨 论

蓝云锋医师:该患者为中年男性,慢性病程,近期加重。主要表现为进行性的体力下降、近期劳累性胸闷胸痛、下肢浮肿,伴有发作性意识丧失。客观检查发现有心房纤颤(快室率型)、间歇性完全左或完全右束支传导阻滞,心室长间歇;心脏超声和核磁检查发现非对称性心肌肥厚和左室流出道轻度梗阻、部分瓣膜病变;冠脉造影见斑块和部分管腔轻度狭窄。诊断考虑肥厚型梗阻性心肌病,心律失常:心房纤颤(快室率型),间歇性完全性左或右束支传导阻滞、心室长间歇。发作性意识不清原因待查:心源性可能性大,不除外短暂脑缺血发作等。

张文莉副主任医师:针对该患者的临床表现特点,应在肥厚型心肌病和扩张型心肌病之间进行鉴别诊断。肥厚型心肌病具有如下特点:与血流动力学特征不相适应的左心室肥厚,常为不对称肥厚并累及室间隔;因舒张功能障碍导致的心衰;即使没有冠脉病变也可能发生心绞痛以及各种心律失常的发生。肥厚性心肌病大体可分为梗阻性和非梗阻性。该患者的临床特点大多数都支持肥厚型心肌病的诊断,心脏MRI检查室间隔厚度达到21mm(正常者<11mm),并伴有流出道的轻度狭窄,达到肥厚型梗阻性心肌病的诊断标准。但也有疑点:在胸片和心脏MRI检查中都发现心脏多腔室增大,从影像和临床表现,需要考虑扩张型心肌病的诊断。我认为,这是三尖瓣关闭不全和二尖瓣关闭不全所致的心脏增大,是血流动力学改变所致。也许,该患者本就是一个肥厚型心肌病扩张型心肌病样症状。另外,持续的心动过速也可导致可逆性的心肌扩大。根据超声心动图和心脏MRI的检查结果,可诊断该患者为非对称性室间隔肥厚型心肌病伴轻度流出道梗阻。

卢才义主任医师:该患者的鉴别诊断应考虑这么几个方面:肥厚型心肌病、扩张型心肌病、冠心病。

肥厚型心肌病:该病约50%病例有家族史,为常染色体显性遗传。主要特征为非对称性的左心室壁增厚(偶尔为与血流动力学效应不相称的同心性肥厚,如没有高血压病史等),可分为梗阻型和非梗阻型,后者还有一个特殊类型为心尖肥厚型心肌病。肥厚型心肌病主要特征为左心舒张功能障碍和多种心律失常,部分患者可有心绞痛、晕厥等临床表现,可发生猝死。该患者符合上述特征。考虑诊断为肥厚型梗阻性心肌病。需要考虑的鉴别诊断如上所述。

扩张型心肌病:该患者胸片和MRI平扫见到心影增大,左、右心房和右心室增大。超声心动图报告二、三尖瓣关闭不全,伴有心房纤颤,但左心室无明显扩大,收缩功能障碍。不符合该型心肌病心腔扩大、收缩力降低的特征。

冠心病:患者病程中有劳累性胸闷,经血管扩张药治疗有效。但冠脉造影未发现严重冠脉病变,心电图"缺血性"改变为持续性,不支持冠心病诊断。肥厚型心肌病因局部心肌肥厚,可造成心肌供血相对不足,产生类似心绞痛的临床特点。容易混淆。

在患者的客观检查中,有一些特征与典型的肥厚型心肌病不符。如多心腔扩大等。我认为这些腔室的扩大可能是肥厚的左心室长期舒张功能障碍,导致左室舒张末期压力增高,进而引起左房-肺血管床-肺动脉压力增高,造成左房和右心系统压力负荷增加。一方面左房增大,促进心房纤颤等心律失常发生,另一方面右心系统高压力负荷,造成右侧心腔扩大和三尖瓣相对关闭不全等病理改变。患者病程中多次发作性晕厥,可考虑以下几方面原因:其一,心室长间歇致阿-斯综合征发作,该患者有间歇性左右束支传导阻滞和长RR间期,支持此诊断;其二,心房纤颤时交感神经系统兴奋性增加,左心室流出道梗阻加重致心排量急剧降低所致晕厥;其三,心房纤颤诱发的心房血栓脱落引起的短暂脑缺血发作,此类病变一般在发作后即刻会遗留一些神经系统症状,该患者没有。

肥厚型梗阻性心肌病的危害主要有猝死、心衰和心内膜炎。其中50%的病例发生猝死,心衰者为25%左右。所以治疗也针对上述危险因素展开。预防猝死的手段包括埋藏式自动复律器和β受体拮抗剂,该患者有房室传导阻滞的潜在风险,不适合后者,除非在起搏器的保护下应用,同时还可改善舒张功能;针对流出道梗阻,可用外科手术或心尖部起搏等方法治疗;同时要积极预防感染性心内膜炎。在日常生活中,建议患者避免劳累,尤其避免急速的运动。针对持续心房颤动,抗凝治疗必不可少。

(参加讨论医师:蓝云锋、张文莉、卢才义)

(蓝云锋 整理)