·临床病理讨论·

Clinicopathological Conference

A 81 years old woman with dissecting aortic aneurysm

(The second case)

Case Presentation

Institute of Geriatric Cardiology, General Hospital of PLA

A 81 years old woman was admitted to the hospital on Feb. 11, 2001 because of drowsiness for half a day.

On Feb. 10, the patient did some housework and went to bed earlier than usual because of fatigue. At 9 o'clock next morning, she was found still sleeping soundly, with rapid respiration and fecal and urinary incontinence. She could respond to stimulation and answer questions. Blood pressure taken at that time was 120/80mmHg(1mmHg = 0.1333kPa) and serum glucose was 16mmol/L. Then she was transferred to our hospital urgently. At the time of her arrival at the emergency department, she became lethargic and cyanotic. The white cell count was 13 000 per cubic millimeter with 66 percent of neutrophils. Arterial blood gas values were as follows: pH 7.1, $PaCO_2$ 74.7 mmHg, HCO_3^- 16.7mmol/L, PaO_2 26.1mmHg. Immediate trachea intubation and artificial ventilation were performed and sodium bicarbonate was supplemented to correct acidosis. An hour later, the patient came around. Her cyanosis was relieved and the blood gases were improved. Then she was transferred to in-patient department for further treatment.

On arrival at the in-patient department, the temperature was 36℃, pulse rate 70, and respiratory rate 21. The blood pressure was 135/67mmHg. On physical examination, the patient was alert with trachea cannula and pulmotor. No murmur was heard on the neck. The lungs were clear and small amount of crackles were present at both lung bases. Heart rhythm was regular and no murmur was heard at any auscultation area. The abdomen was normal, and no peripheral edema was found.

Laboratory tests were performed. The urine was positive (+) for protein but was otherwise normal. Blood chemical values were as follows: SGPT 49 U/L, SGOT 46 U/L, blood urea nitrogen 21.2mmol/L, serum glucose 16mmol/L, creatine kinase (CK) 359 U/L. The serum levels of CK-MB, sodium, potassium and chloride were normal. An electrocardiogram revealed elevated T waves in leads Ⅱ, avF and biphasic T waves in lead V_2. A chest radiograph showed increased lung markings with striated shadow in right lower lobe, normal hilar shadow, distortion of the aorta, prominent and calcified aortic knob, elevated left diaphragm and normal cardiac size.

The patient had hypertension and diabetes mellitus type 2 complicated by diabetic retinopathy. She also had history of chronic bronchitis, chronic obstructive pulmonary emphysema and chronic respiratory failure.

Mechanical ventilation was continued after hospitalization while other effective therapeutic measures such as anti-infection, anti-hypertension, blood-lowering and expectorant were adopted. On the second and third hospital day, CK was measured to be 964 U/L, 643 U/L, respectively, while CK-MB, GPT and GOT remained normal. Electrocardiogram showed inverted T waves in leads V_1-V_4. These changes were thought to be related to hypoxemia and therefore no specific treatment was given. During this period of time, blood pressure was found occasionally high with peak blood pressure being 180/90mmHg, which fell to normal through antihypertensive agents. Purulent sputum was changed to mucoid sputum. No fever was observed and arterial blood gases were normal. On Feb. 16, myocardial enzymes resturned to

normal limits and high upright T waves in lead Ⅰ, Ⅱ, V_4-V_6 were found. But on Feb. 18, peripheral edema appeared and a chest radiograph showed blurring of hilar shadow, hypo-translucent right lung field and suspicious widening of upper mediastinum. A bedside thoracic ultrasonogram showed right-sided pleural effusion. Peripheral edema disappeared after taking diuretics. Later on, gradual withdrawal of mechanical ventilation started. In the morning of Feb. 21, endotracheal tube was successfully removed and mechanical ventilation was withdrawn. After that, the patient's condition remained stable from Feb. 21 to Feb. 25 with normal arterial blood gase values and she could expectorate herself. Arterial blood gases tested on Feb. 25 showed: pH 7.4, $PaCO_2$ 59.3 mmHg, HCO_3^- 31.7mmol/L, and PaO_2 80.1mmHg. Then on Feb. 26, the patient felt quite well all day long and had a long conversation with her family members that afternoon. At 20:20 that night when she was resting on bed, she suddenly felt a severe chest pain accompanied by dyspnea, cyanosis, profuse sweating and subsequent unconsciousness. At that time, the respiratory rate was 8, and the heart rate 65. The blood pressure was 0. Emergent treatment was instituted. At 20:23, the patient's breathing and heartbeat stopped. Resuscitation measures including mechanical ventilation all failed and the patient died.

Clinical Discussion

Dr. ZHENG Qiufu: This was a case of sudden death. Acute pulmonary thromboembolism (PTE) was first considered for the reason that the patient presented chest pain and dyspnea——two symptoms of the triad. Statistical data showed that less than one third of the patients suffering from pulmonary thromboembolism has typical triad and most patients present only one or two symptoms. Once pulmonary thromboembolism occurs, patient's breathing becomes rapid and heart rate increases. In this case, the blood pressure was zero, respiratory rate was 8 and heart rate 65 at the onset. It might be possible that more than 50 percent of pulmonary artery was blocked by emboli. Admission chest X-ray showed such findings as broadening of upper mediastinum, cloudy hilar shadow, left-sided pleural effusion and elevated diaphragm, which were all compatible with the manifestations of pulmonary thromboembolism. If a single etiologic factor is considered, the possible course of the disease might be as such: pulmonary thromboembolism with less than 50 percent of pulmonary arteries block already existed on admission. With the breaking up or autolysis of emboli, the patient's condition got improved for a short time. Thereafter, a massive pulmonary thromboembolism was superimposed and led to sudden death of the patient. Had bed-side hemodynamic monitoring been performed at that time, the presence of pulmonary hypertension and normal or low pulmonary capillary wedge pressure would have gone far to clinch the diagnosis.

Dr. XIE Hengge: The patient had history of lethargy and fecal and urinary incontinence, which tended to suggest neurological diseases. Generally speaking, patients with cerebrovascular disease, whether massive brain infarction or brain hemorrhage, should display increasingly elevated blood pressure with the occurrence of disturbance of consciousness. The manifestation of normal blood pressure on admission and a quick recovery of consciousness in this case argue against neurological disease. Another disease known as acute subarachnoid hemorrhage may also cause sudden death. It has been confirmed at autopsy that 20 percent of patients with sudden death died of subarachnoid hemorrhage and most of the victims were young persons. This patient lacked the aforementioned diagnostic criteria, therefore subarachnoid hemorrhage was ruled out. Considering the patient's medical history, the manifestation of peripheral hypoperfusion, together with her previous history of hypertension will form the pathologic basis for the formation of aortic dissection, especially with fluctuation of blood pressure, so aortic dissection is highly suspected.

Dr. YAN Muyang: This was an elderly woman who had a rapid onset of symptoms and died within 3 minutes. The possible cause was concentrated on ruptured dissecting aortic aneurysm or acute massive pulmonary thromboembolism. Factors in favor of the former were as follows: "cutting" pain accompanied by dyspnea and profuse sweating, the blood pressure being zero in arms, sudden death within 3 minutes, three episodes of paroxysmal hypertension during the night, history of hypertension and diabetes mellitus, broadening of upper mediastinum and calcification of the aorta. All these conditions led to the suspicion of aorta dissection type A, which may rupture and track into pericardium, resulting in pericardial tamponade. Evidences for suspicion of massive pulmonary thromboembolism included the following conditions: senility; prolonged immobilization without anticoagulation; long history of hypertension and diabetes mellitus, which might have the effect of damaging the arterial and venous intima; ambulation after long time period of immobilization, which might evoke detachment of emboli; dyspnea and cyanosis before death. Nevertheless, there were some other evidences against it such as no persistent peripheral edema, no response to diuretics, which often hints deep venous thrombus(DVT), being responsible for 80 percent of pulmonary thromboembolism, and no history of lower limb fracture, trauma or tumor. The "dragging" severe pain other than "pressing" pain and hypo-translucence on chest film were also the counterevidences. Therefore, massive PTE is questionable while minor PTE may not be excluded. In summary, the diagnosis is considered to be ruptured dissecting aortic aneurysm.

Pathological Discussion

Dr. LIU Xiaobing: Anatomical diagnoses: ① pericardial tamponade. Five hundred ml of bloody fluid and some blood coagula were found in pericardial cavity; ② ruptured dissecting aortic aneurysm. An irregular cleft about 2.5 cm in diameter and 2.8 cm away from the free margin of aortic valve in the intima of posterior wall of the ascending aorta was found, being parallel to the longitudinal axis of the aorta. Down from the cleft, dissection filled with large amount of blood coagula was found spreading from aortic root to ascending aorta, covering an area of 20.5cm×7.0cm. There were also hematomas inside the external layer of aortic root and around pulmonary root. Microscopic examination found intimal fibroplastic proliferation with hyaline degeneration in some areas and macro-phage foam cells aggregation and deposition of atherosclerotic substances in some other areas. Medial fibrosis with mucous degeneration and broken elastic fibers could also be seen. On gross examination, it could be seen that the separation was located in media close to adventitia. Pathologic diagnoses were: ① ruptured dissecting aortic aneurysm leading to pericardial tamponade; ② atherosclerotic arterial disease, atherosclerosis grade Ⅳ, complex lesion stage; ③ pulmonary congestion, emphysema with locatized pulmonary collapse; ④ marked multiorgan congestion including gastrointestinal tract, liver, spleen, adrenal glands and kidneys.

Dr. GAO Dewei: This patient, transferred from the emergency department to intensive care unit in respiratory department, was preliminarily diagnosed as respiratory failure. Supported by mechanical ventilation through artificial airway, the patient's condition got better with successful weaning from ventilator and extubation. On the fifth day after extubation, the patient had a sudden chest pain and dyspnea and died without any response to various emergent managements. The diagnosis was verified to be ruptured dissecting aortic aneurysm by autopsy. Dissecting aortic aneurysm has long been regarded as rare. Physicians other than cardiologists have little knowledge about it and are therefore little aware of it. In this case, chest film illustrated obvious aortic calcification and broadening of upper mediastinum shadow. If cardiac ultrasound study was performed just after extubation, direct evidence would be available. We should learn a lesson from this case in which the patient died without confirmatory diagnosis owing

to incomplete investigation. In addition, in this case since the only symptoms the patient presented on admission were drowsiness, weak breathing and respiratory and metabolic acidosis, the predisposing condition of respiratory failure was worthy of exploring. In my opinion, ruptured aortic dissection already existed when the patient was admitted, and caused cerebral ischemia due to its extension to carotid artery and produced such clinical sympotoms as drowsiness or coma. This case reminded us especially doctors in emergency department to enhance awareness of such disease. According to medical literature, dissecting aortic aneurysm should be thought of when the following conditions especially hypertension present: ① sudden onset of severe chest, abdomen or back pain described as tearing and migratory; ② asymmetry of pulses or blood pressure; ③ severe pain but with mild signs; ④ chest pain with progressive worsening of left heart failure; ⑤ symptoms of multi-organ dysfunction such as angina pectoris, syncope, renal failure and jaundice which can not be explained by a disease alone. Diagnostically, cardiac ultrasound study is recommended. Computed tomography (CT) or magnetic resonance imaging (MRI) is also a diagnostic option if possible.

Dr. LI Bojun: The natural prognosis of dissecting aortic aneurysm is devastating as evidenced by its very high mortality in patients without timely treatment. It has been reported that 25 percent died within 24 h, 50 percent within a week, 75 percent in one month and 90 percent by one year and rupture is the commonest cause of sudden death. This patient died of ruptured dissecting aortic aneurysm type Stanford A resulting in fatal acute pericardial tamponade. The lesion in this case was suitable for surgical intervention if the clinical course was not so rapid and critical. The widely used surgical technique is the replacement of aorta with valve assisted by extracorporeal circulation. In case of chronic type Stanford A, the same surgical method is chosen as dealing with acute cases. For cases of chronic type Stanford B with dissection limited to descending aorta, still the same technique is used because blood supply for aortic branches may come in part from blood in false lumen. Here is the general surgical procedures. After successful construction of left ventricular bypass, blood flow in descending aorta is blocked, with the proximal end of incision located a little distant to subclavian artery. Then a wedge-shaped part of the septum between true and false lumen is cut from the proximal end, followed by fixing the remaining septum on the external wall of aorta and ligating its two ends to construct a new closed false lumen. After that, a tube-shaped artificial blood vessel is anastomosed with the false lumen so that blood can enter either true or false lumen to maintain blood supply of extremities. In cases of type Stanford B in which tearing part is located near descending aorta, endovascular repair is feasible.

Dr. YU Senyang: The patient with previous history of chronic respiratory failure presented drowsiness on physical exertion. Evidences including abnormal values of arterial blood gases, high white cell count and increased lung markings on chest radiograph led to the diagnoses of respiratory and metabolic acidosis as well as respiratory failure worsened by pulmonary infection, which were also proved by the improvement of the patient's condition after the use of antibiotics and mechanical ventilation. The main cause of sudden death was ruptured dissecting aortic aneurysm causing pericardial tamponade and subsequent heart failure. Echocardiography is considered to be very specific and accurate in diagnosing aortic dissection, though trans-thoracic echocardiography had poor display of aortic dissection limited to aortic arch and descending aorta. With the clinical application of trans-esophageal echocardiography, diagnostic rate has been greatly improved. Digital subtraction angiography (DSA) remains the gold standard in diagnosing aortic dissection, but its use is limited owing to its interventional characteristics and the use of contrast medium. In clinically unstable patients with hypersensitivity to iodine or with liver or kidney dysfunction, this investigation is contraindicated. CT, spiral CT (SCT) and magnetic resonance imaging are among the useful diagnostic methods. SCT scanning can further identify involved aortic branch vessels and therefore can provide more effective information for the surgical management.

(Translator CHEN Shujuan)

1 例 83 岁女性夹层动脉瘤

(第 2 例)

1 病历摘要

患者,女性,81 岁。因昏睡伴大小便失禁半天于 2001 年 2 月 11 日入急诊室。发病时血压 120/80 mmHg(1mmHg = 0.1333kPa),血糖 16mmol/L。入急诊室后患者口唇紫绀,嗜睡,白细胞(13×10^9/L)及中性粒细胞(0.90)增多,血气分析提示失代偿性酸中毒,Ⅱ型呼吸衰竭。经过气管插管、机械通气及纠正酸中毒后,病情略好转入院。既往有高血压病、2 型糖尿病及糖尿病性肾病、慢性支气管炎、阻塞性肺气肿、慢性呼吸衰竭病史。入院查体双肺底可闻及少许细湿性啰音,余正常。胸片示肺纹理增多,肺门影不大,主动脉迂曲,主动脉结突出、钙化。入院后继续机械通气,同时抗感染、扩冠、降压、降糖等治疗,病情渐好转,全身情况改善。心肌酶由 CK 964U/L,逐渐下降,至 2 月 16 日心肌酶及血气分析结果均转为正常,心电图也正常。2 月 18 日出现双下肢水肿,胸片示肺门模糊,右肺透光度减低,上纵隔增宽。胸腔 B 超提示右侧胸腔少量积液。经利尿治疗后水肿消失。21 日成功拔管撤机,之后病情稳定 4 日。25 日血气分析结果为 pH7.4, $PaCO_2$ 59.3mmHg, HCO_3^- 31.7 mmol/L, $PaCO_2$ 80.1mmHg。2 月 26 日患者精神好,白天与家人谈话时间较长,晚 20:20 卧床休息时突感胸痛,呈刀割样,伴呼吸困难,口唇紫绀,大汗淋漓,随即意识丧失,血压测不到,呼吸 8 次/min,心率 65 次/min,立即抢救治疗,20:23 呼吸、心跳停止,行气管插管机械通气等抢救治疗无效死亡。

2 临床讨论

这是一个猝死病例。老年女性患者,有长期高血压、糖尿病病史,发病时表现为突发的刀割样胸痛,伴呼吸困难及紫绀,血压测不到,抢救治疗无效,患者在 3 min 内死亡。分析其猝死原因,主要考虑为主动脉夹层动脉瘤破裂,诊断依据如下:① 患者既往有高血压的病史,本次住院期间血压有大幅度的波动,提示存在引起主动脉夹层的病理基础;② 患者发作当时表现为典型的刀割样疼痛伴呼吸困难、大汗,血压测不到,而呼吸、心率尚存;③ 入院后的检查曾发现上纵隔影增宽,主动脉有明显钙化。根据上述资料推测发病过程可能为:A 型主动脉夹层动脉瘤破裂,血液进入心包,造成心包填塞,引起循环衰竭而死亡,其次要考虑的是急性大面积肺栓塞,患者有长期高血压,糖尿病等易损害血管内膜而促进血栓形成的危险因素,加之卧床 10 余天未进行抗凝治疗,为血栓形成进一步创造了条件,而卧床后的正常活动可能成为血栓脱落的诱因。结合发作时表现为胸痛和呼吸困难,胸片提示右侧胸水、左膈肌抬高等特点均不能排除肺栓塞的诊断。存在的疑点是栓子来源不明,深静脉血栓形成的证据不足。另外要排除脑血管疾病。入院前患者有昏睡、大小便失禁,提示有脑血管病存在的可能,但随后的所有资料均未提供进一步证据,可以排除。

(参加讨论的医师:郑秋甫,解恒革,晏沐阳)

3 病理讨论

解剖发现:① 心包填塞;② 主动脉夹层动脉瘤形成伴破裂,夹层起始于升主动脉后壁,终止于降主动脉,解离处位于中膜近外膜侧。

病理诊断:① 主动脉夹层动脉瘤形成,动脉瘤破裂继发心包填塞;② 主动脉粥样硬化症,主动脉粥样硬化Ⅳ级,复合病变期;③ 双肺淤血,肺气肿伴局部肺萎缩;④ 全身多脏器(胃肠、肝、脾、肾上腺、肾)显著淤血。

主动脉夹层动脉瘤一直被认为是少见病,非心血管病医师对此认识不足。结合本例患者及有关资料,提醒医师对于一个高血压病患者若伴有突发胸、腹、背部撕裂样疼痛,胸痛剧烈而相应体征少而轻、血压、脉搏不对称及难以用单病因解释的多系统损害等情况时要想到该病的可能。超声心动图检查有较高的诊断特异性及准确性,而数字减影血管造影术(DSA)仍是诊断的金标准,但对于有碘过敏史或有肝、肾损害者,CT,SCT 及 MRI 等检查均是可靠选择。主动脉夹层的预后凶险,死亡率高,主要原因是瘤体破裂,一旦发生,即应行紧急外科干预,有望挽救部分患者生命。对于确诊的慢性稳定期患者,可酌情选择主动脉夹层内修复治疗或主动脉置换术。

(参加讨论的医师:刘小兵、高德伟、李伯君、俞森洋)

(高德伟整理)

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