

A Report Case of Sudden Cardiac Death in a Young Adult Male from Northeastern Part of Thailand with Mitral Valve Prolapse

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A case of sudden unexpected natural death in a young adult male from the north-eastern part of Thailand with clinical non Lai-Tai disease, but pathologic feature of mitral valve prolapse is discussed. The approach to the postmortem examination of the mitral valve is reviewed. Because of the sudden nature of this death, this entity is more commonly seen in Medico-legal Medicine populations than in hospital autopsies.

Keywords : Non Lai Tai disease, Mitral valve prolapse, Autopsy

J Med Assoc Thai 2004; 87(4): 446-9

Mitral valve prolapse syndrome has many other names, including floppy mitral valve, myxoid mitral valve, billowing, ballooning, hooding, and mid-systolic click syndrome. It is a fairly common condition and may be found in 5 to 15 % of the general population^(4,5), is more common in women (female to male ratio 3:2) between 20 and 40 years of age⁽³⁾. The syndrome is characterized by symptoms, signs, electrocardiographic changes, various imaging technique abnormalities and pathologic changes found in the heart⁽³⁻⁵⁾.

Sudden death in mitral valve prolapse occurs in adults where the findings on autopsy may be ballooning or thickening or myxoid degeneration and elongation of the chordae tendinae. The victims may or may not have a history of a mid-systolic click on auscultation of the heart and electrocardiographic changes. In the past, this condition has been ignored by the Medico-legal Medicine doctors, who did not believe it to be significant⁽⁶⁾. There is also evidence that other Medico-legal Medicine doctors have misidentified this entity as non Lai Tai disease or Sudden Unexplained Nocturnal Death in Southeast Asia⁽²⁾.

Only about 70 cases of sudden death in mitral valve prolapse have been reported^(8,9). A number of authors have indicated that the incidence of sudden death in mitral valve prolapse is probably underreported^(6,8-11). The authors report the first case of sudden death in mitral valve prolapse in Thailand.

Case Report

A healthy 38 year-old Thai male was found dead in his house on the bedroom floor in the morning by his spouse. He was born in Ubonrajathani (North-eastern part of Thailand). He had complained of chest pain a few days previously and had been feeling ill for the week before his death. The investigator from the Police Hospital gave a short statement of "suspicious of non Lai Tai disease".

At autopsy, the body weighed 70 kg. and measured 166 cm. in length. The heart weighed 370 grams (predicted normal weight 310 grams). The mitral valve showed ballooning of the cusp. The valve was thickened with myxoid degeneration. The chordae tendinae were elongated and delicate. Retrograde infusion of water into the left ventricle through the aorta showed the posterior mitral valve leaflet to balloon into the left atrium and leak water. There was mild subendocardial fibrosis of the tips of the papillary muscles and of the interventricular septum beneath the anterior mitral leaflet. The aortic, tricuspid, and pulmonic valves were normal. The thickness

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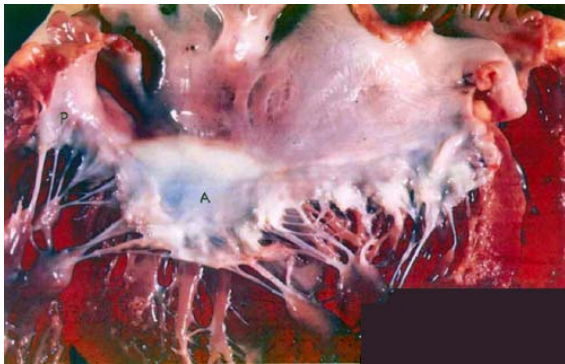


Fig. 1 Mitral valve of the heart opened in the conventional manner. The posterior leaflet (P) showed diffuse thickening and fibrosis. A gelatinous consistency was noted on the cut edge of the valve. The chordae tendinae are thin and delicate. There was fibrosis on the tips of the papillary muscles

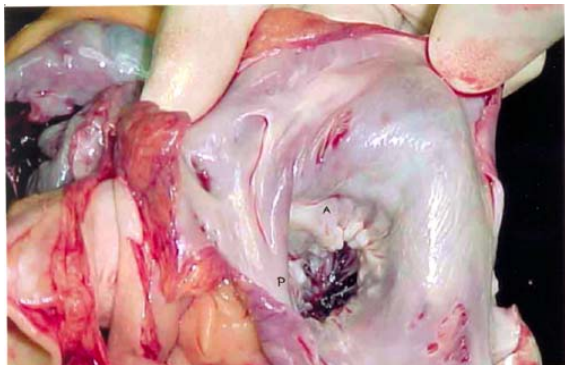


Fig. 2 Mitral valve of the heart viewed from left atrium showed markedly billowy anterior (A) and early change of posterior (P) valve leaflets with moderate interchordal hooding into the left atrium

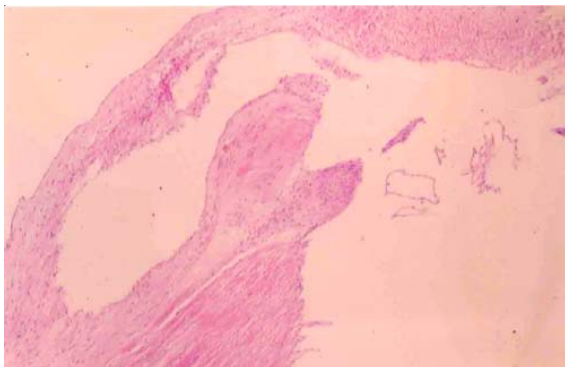


Fig. 3 Microscopic section of the mitral valve showed myxoid degeneration with fibrosis

of the right and left ventricles was normal. The coronary arteries pursued their usual anatomic course and displayed no atherosclerosis.

The myocardium revealed unremarkable changes. There was no evidence of any infiltrative cell in the myocardium of the heart. The lungs weighed 970 grams and were moderately congested and edematous. The liver weighed 2050 grams; spleen 150 grams; kidneys 330 grams; brain 1340 grams. All organs were minimally congested.

Discussion

Clinical features of mitral valve prolapse

The majority of people with mitral valve prolapse are asymptomatic and never seek medical attention⁽¹²⁾. A family history of mitral valve prolapse or sudden death or both may be helpful in suspecting the entity. There is a wide constellation of signs and symptoms related to mitral valve prolapse. These include light-headedness, syncope, chest pain, palpitation, dyspnea, and tachycardia⁽¹²⁾. Auscultatory findings usually reveal a characteristic mid-systolic click. Electrocardiographically documented arrhythmias; a trial fibrillation or flutter, ST-T abnormalities, prolongation of the QT interval, atrial or ventricular tachycardia and ventricular fibrillation have been reported. Echocardiography is a non invasive procedure used to track the motion of the atrial and ventricular walls and the cardiac valves. The usual finding in mitral valve prolapse is an abrupt, mid-systolic billowing motion of the mitral valve.

Pathology

The underlying pathologic process in mitral valve prolapse is myxomatous proliferation in the spongiosa layer of the involved leaflet that invades and disrupts the fibrosa layer. In the early stage of mitral valve prolapse, the valve leaflets are delicate, and transparent. Later the affected cusps become opaque with an increase in surface area. The leaflets become more fibrotic and thickened. As the process continues aneurismal dilatation of the leaflet areas between the chordae tendinae occurs. This process is known as interchordal hooding and gives the valve a parachute appearance. Thickening and fusion of the chordae tendinae may be seen. They are usually more evident on the posterior leaflet. Sudden death may occur at any stage^(7,13).

Examination of the heart

The diagnosis of mitral valve prolapse may be made on the basis of a medical history, the gross pathologic changes seen in the heart, and the exclusion of other causes of death. A bird's eye view through the left auricle at the mitral valve cusps before conventional opening of the heart and a retrograde infusion of water by hose through the aorta into the left ventricle is a simple way of demonstrating mitral valve prolapse at autopsy⁽¹³⁾. Microscopic examination should confirm the myxoid proliferation in the spongiosa and disruption of the fibrosa of the affected valve leaflet⁽¹⁴⁾.

Incidence of sudden death

The incidence of sudden death in mitral valve prolapse syndrome has varied from 5-15% in long term studies^(15,16). Young adult females appear to be at higher risk for sudden death in mitral valve prolapse⁽¹⁷⁾. Other risk factors include frequent premature ventricular contractions, ST-T changes on electrocardiogram, marked prolapse on echocardiography, mitral regurgitation and a family history of sudden death. The mechanism of death in the present report case is felt to be ventricular arrhythmias.

Conclusion

Mitral valve prolapse is a significant and probably underdiagnosed cause of sudden death, especially in adult Thai female and male. Because of the sudden unexpected nature of these deaths, many will come to the attention of the Medico-legal Medicine physicians or Forensic Medicine doctors. It is important that Forensic Medicine doctors maintain a high index of suspicion that mitral valve prolapse may be a possibility and diligently search for the pathologic changes found in this entity.

Acknowledgment

The authors wish to thank Ms. Manee Boonkerm and Mr. May-t Buachoo for their assistance in preparing the photographs used in this article.

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รายงานการชันสูตรศพชายหนุ่มไทยอีสานเสียชีวิตกะทันหันด้วยลิ่มหัวใจด้านซ้ายปลิ้น รายแรก
ของประเทศไทย

ธำรง จิรจรียาเวช, ปานใจ โวหารดี, วิชาญ เบี้ยวรัมย์

รายงานผู้เสียชีวิตชายไทยอีสานวัยกลางคนซึ่งทางตำรวจสงสัยว่าเป็นโรคหลอดเลือดหัวใจที่พบค่อนข้างบ่อย
ในชาว ลาว เขมร เวียดนาม และไทยอีสาน แต่จากการชันสูตรผ่าศพ พบว่าพยาธิสภาพของผู้เสียชีวิต มีลิ่มหัวใจ
ด้านซ้ายปลิ้น จึงได้มีการวิเคราะห์ ตรวจสอบ และวิจารณ์ถึงการพบของโรคนี้ ซึ่งเป็นโรคที่พบได้ค่อนข้างบ่อย
ในคนหนุ่มสาวที่เสียชีวิตอย่างปัจจุบันทันด่วน และไม่คาดคิด โรคหัวใจด้านซ้ายปลิ้นนี้เป็นโรคที่พบได้ค่อนข้างบ่อย
ในวงการนิติเวชของประเทศทางตะวันตก รายงานนี้เป็นรายงานแรกที่พบในประเทศไทย ที่เสียชีวิตนอกโรงพยาบาล
