Development of Pulmonary Hypertension After Ventriculoatrial Shunt Implantation

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ABSTRACT

Pulmonary hypertension is known as one of most serious complication of ventriculoatrial shunts. There is a long latency period between ventriculoatrial shunt implantation and development of pulmonary hypertension. Because of this latency and relative rarity of pulmonary hypertension in the general population, clinicians may misdiagnosis these patients. A young male patient was referred to our clinic with suspect of an atrial septal defect from an outside hospital. He had a ventriculoatrial shunt because of hydrocephalus. We tought the clinical diagnosis as pulmonary hypertension due to ventriculoatrial shunt according to the his clinical history, ECG and echocardiography findings. The pulmonary perfusion scintigraphy findings supported the diagnosis. His shunt was replaced to the ventriculoperitoneal shunt by Brain Surgery Clinic.

Key words: *Ventriculoatrial shunt, pulmonary hypertension, pulmonary embolism*

Ventriküloatriyal Şant İmplantasyonu Sonrası Pulmoner Hipertansiyon Gelişimi

Ventriküloatriyal şant komplikasyonların en ciddi olanlarından biri de pulmoner hipertansiyondur. Ventriküloatriyal şant yerleştirilmesi ile pulmoner hipertansiyon gelişmesi arasında uzun bir latent dönem vardır. Bu uzun latent dönem ve pulmoner hipertansiyonun genel popülasyon içinde nispeten nadir görülen bir durum olması sebebiyle bu hastalar yanlış teşhislerle takip edilebilirler. Genç bir erkek hasta başka bir merkezden kliniğimize ASD ön tanısı ile sevk edildi. Hastanın hidrosefali sebebiyle ventriküloatriyal şantı mevcuttu. Klinik öyküsü, EKG ve ekokardiyografi bulgularına göre hastada ventriküloatriyal şanta bağlı pulmoner hipertansiyon düşündük. Akciğer perfüzyon sintigrafisi tanıyı destekledi. Hastanın şantı Beyin Cerrahi Kliniğince ventriküloperitoneal şanta çevrildi.

Anahtar kelimeler: *Ventriküloatriyal şant, pulmoner hipertansiyon, pulmoner emboli*

INTRODUCTION

Since ventriculoatrial shunts were widely used in the past, clinicians may be faced with patients having pulmonary hypertension due to ventriculoatrial shunt. In this case report, we presented a case with pulmonary hypertension after ventriculoatrial shunt implantation. Our aim is remind that patients with ventriculoatrial shunts have an increased risk of developing pulmonary hypertension, which can occur decades after shunt implantation.

CASE

A 30-years old, male patient was applied to an outside hospital complaining severe dyspnea last 2-3 days but gradually increasing since that is 6 months. Transthoracic echocardiography was performed to the patient and revealed that right cardiac chambers were wide and pulmonary artery pressure was high. His electrocardiography was showed right axis deviation and incomplete right bundle branch block. (Figure 1) The patient was referred to our clinic for further evaluation with suspect of an atrial septal defect. In history of the patient, ventriculoatrial shunt was performed to treat hydrocephalus which was occurred after a traffic accident. Transthoracic echocardiography revealed dilatation of right cardiac chambers and second degree tricuspid valve regurgitation. Pulmonary artery systolic pressure was 85 mmHg (70+15mmHg).(Figure 2) and there was a shunt catheter in the right atrium. Transesophageal echocardiography showed a patent foramen ovale but not atrial septal defect and doppler echo demonstrated flow across the interatrial septum in a right to left direction. Development of pulmonary hypertension was considered after ventriculoatrial shunt implantation. On pulmonary Tc-99m MAA (Macro Aggregated Albumin) perfusion scintigraphy, segmental and subsegmental perfusion defects were observed. These scintigraphy findings were reported as the most likely results of pulmonary thromboemboli. The patient was recommended to apply to the Brain Surgery Clinic and shunt was replaced to the ventriculoperitoneal shunt. Warfarin medication as anticoagulation therapy was given to the patient and discharged with recommendation of clinical follow-up .

DISCUSSION

Ventriculoatrial shunt implantation was first used for treatment of hydrocephalus in late 1940s (1). After demonstrations of ventriculoatrial shunt implantation as an efficient and survive improving medication in treatment of hydrocephalus, it was widely used in 1950s. However, several ventriculoatrial shunt related complications were observed including shunt infections, infective endocarditis, septicemia, meningitis, arrhythmias, myocardial damage, obstruction of vena cava, intracardiac thrombus, pulmonary emboli and pulmonary hypertension in 1970s. Therefore, ventriculoatrial shunt implantations were mostly abandoned (2,3) and ventriculoperitoneal shunts were preferred to use because of their ease and lower level complications (4). Since ventriculoatrial shunts were widely implanted in the past, it is possible to be faced with these patients in clinics. Kluge et al. retrospectively evaluated 575 patients having pulmonary hypertension between 1999 and 2006, and reported findings of 6 patients with ventriculoatrial shunt (5). Mean age of these patients were 42.5+/-8.3 years, pulmonary arterial pressure was 53.3+/-14.9 mmHg and the median interval from shunt implantation to the diagnosis of pulmonary hypertension was 16.5 years. On ventilation perfusion scintigraphy, bilateral multiple perfusion defects were observed in all patients but pulmonary thromboembolism was observed in only two patients on pulmonary angiography and thorax CT.

Bonderman et al reported that ventriculoatrial shunts increased the likelihood of chronic thromboembolic pulmonary hypertension 13-fold (6). Pathogenetic process underlying pulmonary hypertension in these patients remains unclear. Several mechanisms have been proposed, such as the presence of shunt infection causing the persistent activation of clotting factors with recurrent thromboembolism (5,7) or a reaction of the pulmonary endothelium to some yet undefined contents of cerebrospinal fluid, possibly brain thromboplastin (5). Seratonin is a component of the cerebrospinal fluid and its strong vasoconstrictor feature may have a critical role in severe pulmonary hypertension development (5)

There is a long asymptomatic embolization period before development of severe pulmonary hypertension in most cases. In cases that were reported by Kluge et al., the mean period was 16.5 years for diagnosis of pulmonary hypertension from shunt implantation and the mean time from the first symptoms to diagnosis was 9.5

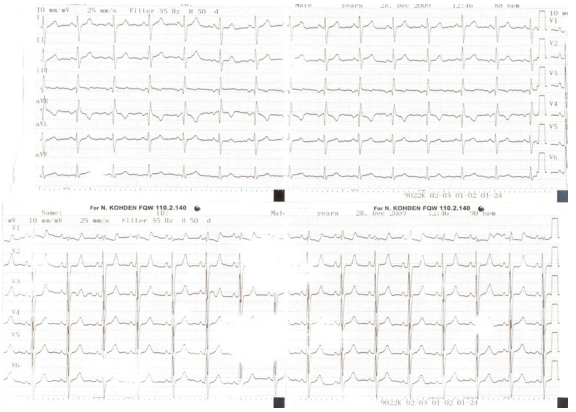

Figure 1*. ECG shows right axis deviation and incomplete right bundle branch block.*

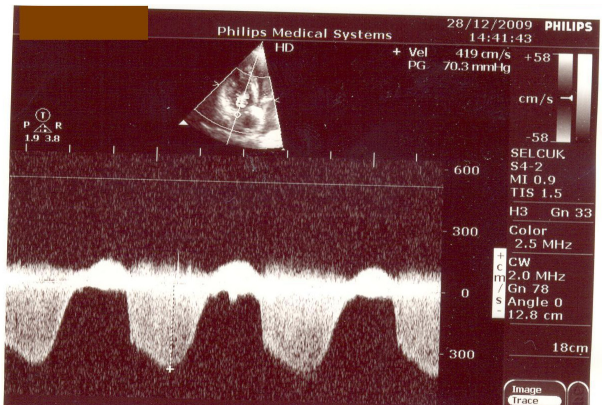

Figure 2*. Transthoracic echocardiography shows severe pulmonary hypertension*

months (1.5-12 months). These periods were 15 years and 6 months in our case, respectively. A serious time lag between the first symptoms and diagnosis has critical importance in these cases. Probable reasons for such delays may be pulmonary hypertension cases are rarely seen in population and the lack of awareness that ventriculoatrial shunts cause pulmonary hypertension.

In summary, patients with ventriculoatrial shunts should be kept under careful evaluation for symptoms of progressive dyspnea on exertion because of having increased risk of pulmonary hypertension. After diagnosis, shunt should be removed and replaced with another type of shunt if necessary. These patients should receive lifelong anticoagulation adjusted to a target INR between 2 and 3.

REFERENCES

1. *Nulsen FE, Spitz EB. Treatment of hydrocephalus by direct shunt from ventricle to jugular vein. Surg Forum 1952;2:399-403.*
2. *Machinis TG, Fountas KN, Hudson J, Robinson JS,T Roup EC. Accurate placement of the distal end of a ventriculoatrial shunt with the aid of real-time transesophageal echocardiography.Technical note. J Neurosurg 2006;105(1):153-6.*
3. *Yavuzgil O, Ozerkan F, Ertürk U, Işlekel S, Atalay Y,Buket S. A rare cause of right atrial mass: thrombus formation and infection complicating a ventriculoatrial shunt for hydrocephalus.Surg Neurol 1999;52(1):54-60.*
4. *Vernet O, Rilliet B. Late complications of ventriculoatrial or ventriculoperitoneal shunts. Lancet 2001;358:1569-70.*
5. *Kluge S, Baumann HJ, Regelsberger J, et al. Development of pulmonary hypertension in adults after ventriculoatrial shunt implantation.Respiration 2009;78:30-35.*
6. *Bonderman D, Jakowitsch J,Adlbrecht C, et al. Medical conditions increasing the risk of chronic thromboembolic pulmonary hypertension.Thromb Haemost 2005; 93(3):512-16.*
7. *Kumar A, Rose JS, Reid CL, Perera PF, Michael TA, Chandraratna PA. Echocardiographic demonstration of pulmonary embolism as it evolves through the right heart chambers. Am J Med 1985 ;79(4):438-540.*