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Unroofed Coronary Sinus Syndrome (Report of one case)

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〈국문초록〉

관상정맥동천정 결손증 (치험 1례)

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관상정맥동천정 결손증이란 선천성 심장기형의 일종으로 관상정맥동과 좌심방사이의 중격 (관상정맥동천정)이 부분 혹은 완전 결손되는 매우 휘귀한 질환이다.

이 질환은 관상정맥동의 결손과 잔존 좌측 상공정맥의 좌심방내 개구를 특징으로 한다. 우-좌 단락과 혈구과다증 등으로 야기되는 중추신경계의 합병증을 예방하기 위하여 좌상공정맥의 결찰 혹은 우방쪽으로의 우회술은 중요한 일이다. 수술치료는 대개 심방내 baffle을 이용하여 3개의 공정맥을 우방쪽에 오도록하여 심방중격을 폐쇄하는 방법으로 한다.

본 교실에서는 최근 술전 진단에서 좌상공정맥과 부분 심내막상결손증으로 진단되었으나 수술시 관상정맥동 천정결손증, 좌상공정맥, 단심방, 승모판 cleft로 확진된 7세의 여아에서 Dacron 포를 이용한 심방내 baffle로 수술하여 좋은 경과를 취한 1례를 경험하였다.

— Abstract —

The unroofed coronary sinus syndrome is a spectrum of cardiac anomalies in which part or all the common wall between the coronary sinus and the left atrium is absent. This defect is part of a developmental complex which incliudes absence of the coronary sinus and termination of a persistent left superior vena cava in the left atrium. Recognition of this complex is important so that interruption or diversion of the left superior vena cava may be done to prevent subsequent centeral nervous system complications. Surgical correction uses an intraatrial baffle to divert flow from the left superior vena cava to right atrium and to close the atrial septal defect.

This reoprt describes a 7 years old female patient in whom the left superior vena cava was identified preoperatively and the complex (unroofed coronary sinus syndrome, common atrium, mitral valve cleft) recognized at the time of operation. Surgical correction, following repair of cleft mital valve, utilized a Dacron patch baffle to route the left caval blood to the right atrium and included closure of the atrial septal defect

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Case

A 7-year old female was referred for evaluation of cardiac murmur detected in several clinics for frequent URI and exertional dyspnea. There were no specific family and past histories. Activity was restricted because of easy fatigability and dyspnea (NYHA II). There was no history of cyanosis and painting.

The blood pressure was 90/60 mmHg and the pulse was 80/minute and regular. There were no cyanosis or clubbing. The first heart sound was increased slightly in intensity, and the second heart sound was split during expiration. A grade 4/6 systolic ejection murmur was present over the upper and midleft sternal border, and there was a grade 2/6 blowing systolic murmur at the apex. A faint, low-pitched sound was heard in diastole at the lower left sternal border.

The EKG showed left axis deviation and a slightly prominent R was in Lead V_1(Fig. 1). A chest X-ray revealed prominent pulmonary vascularity. The overall heart size was normal with slight prominence of the left atrial appendage(Fig. 2).

In echocardiogram, large ASD and mitral valve cleft and paradoxical movement of ventricular septum were noticed.

Cardiac catheterization and angiography was done from the right femoral artery and vein. A significant increase in oxygen saturation of sampled blood from the right atrium was demonstrated. All right heart pressures were normal. From the right the left atrium and ventricle were entered easily, suggesting an ostium primum type of atrial septal defect. when an attempt was made to advance the catheter to the pulmonary artery, the tip was noted to pass up the left sternal border superior to the main pulmonary artery. The cather was retracted before blood samples were obtained, but the pressure monitoring indicated a low-pressure vessel.

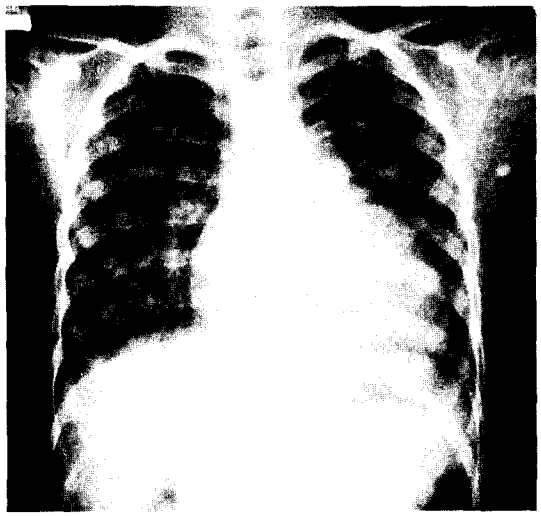

Fig. 2. The preoperative chest X-ray revealed some prominent pulmonary vascularity. The overall heart size was normal with slight prominence of the left atrial appendage.

This was assumed to be a persistent left superior vena cava. An attempt to pass the catheter form the right to the left subclavian vein was successful. In left heart catheterization, cleft mitral valve with

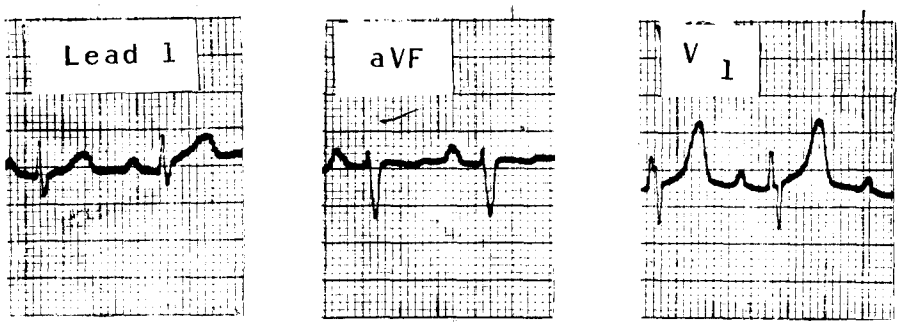

Fig. 1. The preopertaive EKG, showing left axis deviation and a slight prominent R wave in V_1.

Table 1. Cardiac Catheterization data

Site	Pressure(mmHg) Systolic/diastalic(mean)	O_2Sat. (%)
SVC(Right)	9/3 ($\bar{6}$)	63.0
IVC		63.3
RA(High)		90.9
RA(Midle)	8/3 ($\bar{4}$)	92.7
RA(Low)		89.7
RV	30/4	91.0
PA(main)	24/15($\overline{21}$)	93.4
PA(left)	24/15($\overline{19}$)	91.5
LA	7/2 ($\bar{4}$)	90.0
LV	96/16	90.6
Aorta	90/60($\overline{70}$)	90.8

SVC : superior vena cave, IVC : inferior vena cava RA: right atrium, RV : right ventricle, RA : pulmonary artery, LA : left atrium, LV : left ventricle

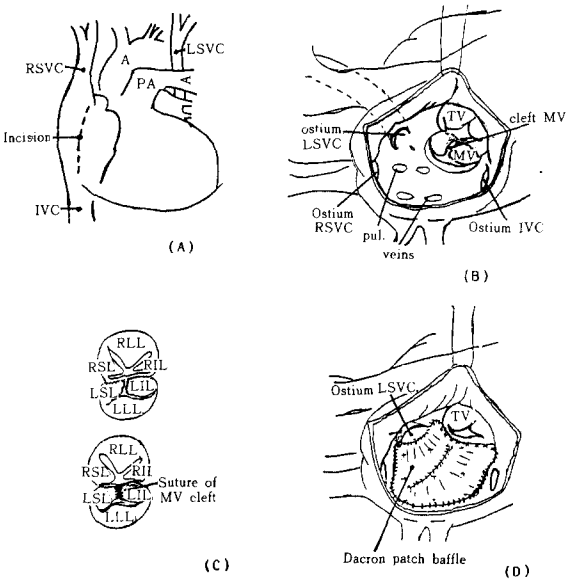

Fig. 3. The schemata of operation. (A) Persistent LSVC and no connection between the right and left SVC was noted. (B) On opening the RA, nearly completely absence of atrial septum (common atrium), cleft of MV anterior leafleat, ostium of LSVC in LA just anteriorly the ostium of left auricle were noted. (C) Suture repair (5 interrupted suture with 6-0 Prolene) of mitral valve cleft was done. (D) Dacron patch (5 cm×5 cm) baffle was used for formation of atrial septum and diverting the LSVC blood to RA. (A: aorta, PA: pulmonary artery, RSVC: right superior vena cava, LSVC: left superior vena cava, IVC: inferior vena cava, TV: tricuspid valve, MV: mitral valve, RSL: right superior leafleat, RLL: right lower leafleat, RIL: right inferior leafleat, LSL: left superior leafleat, LLL: left lowre leafleat, LIL: left inferior leafleat, RA: right atrium, LA: left atrium)

mitral regurgitation was noticed(Table 1).

The preoperative diagnosis was ostium primum atrial septal defect with a probable persistent left superior vena cava. The patient underwent operation utilizing cardipulmonary bypass. Opening the pericardium, identified persistent left superior vena cava and absence of communication between the right and left superior vena cavae. External diameter of both side SVC was 1.0 cm, IVC was 2.0 cm. Size of aorta was 1.5 cm and main pulmonary artery was 2.5 cm. Right ventricular hypertrophy was more prominent than left side and severe right atrial enlargement were also noted(Fig. 3–A). Upon exploration of the right atrium nearly complete absenc of atrial septum (common atrium) was identified. complete cleft was noted in the anterior leafleat of the mitral valve. The coronary sinus was absent, and dark blood was noted to be entering the left atrium through multilpe small thebesian veins. The left superior vena cava was similar in size to the right and entered the left atrium just medial to the left atrial appendage. These findings were all compatible with unroofed coronaray sinus syndrome with common atrium, mitral valve cleft (Fig. 3–B).

Because no connection between the left and right superior vena cavae had been identified prior to operation, a third venous cannula was introduced to the left superior vena cava through the interior of the atrium and snared externally as other vena vava (Fig. 4). The mitral cleft was repaired by 5 interrupted sutures of 6–0 Prolene (Fig. 3–C), a large Dacron (5 cm×5 cm) patch baffle was used to from the atrial septum remaing all three vena cavae in right atrium to tunnel blood from the left superior vena cava to the right atrium. Coronary venous drainage was ignored (Fig. 3–D).

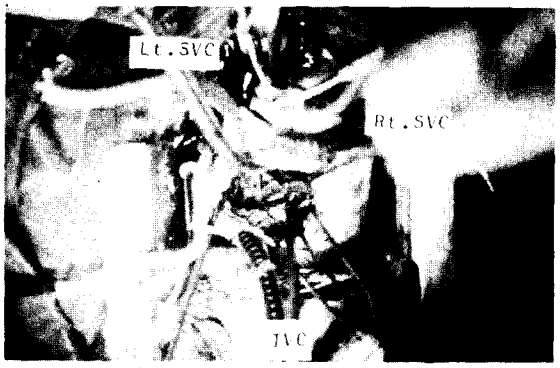

Fig. 4. Real operative findings, showing a third venous cannula inserted to the left superior vena cava through the interior of the atrium. (Lt. SVC: cannula of left superior vena cava, Rt. SVC: cannula of right superior vena cava, IVC: cannula of inferior vena cava)

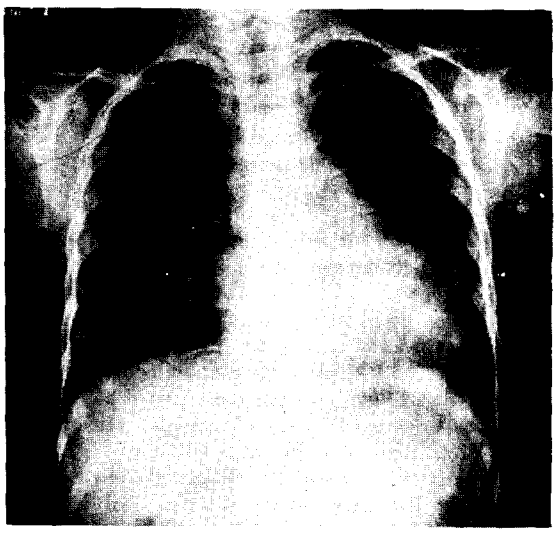

Fig. 5. The postoperative chest X-ray findings, showing decreased heart size and pulmonary vascurality.

Postoperative recovery was excellent. A postoperative chest X-ray showed a decrease in heart size and pulmonary vascurality (Fig. 5). She was discharged 3 weeks later with the good result.

Comments

The unroofed coronary sinus syndrome did not come to the attention of cardaic pathologists before the era of cardiac catheterization and cardiac surgery. Winter provided a pathologic paper in 1954 that mentions persistent left superior vena cava (LSVC) attached to the left atrium[1], and Friedlich and colleagues reported four instances of LSVC entering the left atrium, as identified at cardiac catheterization[2]. An isolated case was also presentented by Tuchman and colleagues in 1956[3]. Campbell and Deuchar, in 1954, referred to instances of LSVC attached to the left atrium, although they did not have such an example in their own series of LSVC[4]. They appreciated that in such cases there were no ture coronary sinus. The understanding of the morphology of the syndrome came from the classic paper by Raghib, Edwards, and coleagues in 1965[5]. The descriptive phrase "unroofed coronary sinus" was first used by Helseth and Peterson in 1974[6].

The embryological abnormality causing this developmental anomaly has been described in detail by Raghib and colleagues[5]. All the features of the complex result from faulty development in the sinoatrial region of the heart. The left atriovenous fold and left cardinal vein normally contribute to formation of the posteroinferior portion of the atrial septum and coronary vein. Anomalous development results in a posteroinferior defect in the atrial septum and absence of the coronary sinus. The left superior vena cava persists and, together with the thebesian veins, drains directly into the left atrium. They, therfore, described a developmental complex characterized by (1) union of the left superior vena cava with the left atrium, (2) absence of the coronary sinus, (3) an atrial septal defect in anticipated location of the coronary sinus ostium.

The unroofed coronary sinus is complete or incomplete. In one from of this the coronary sinus does not exist because the common wall between it and the left atrium is absent. A persistent LSVC, which normally become continous with the coronary sinus, connects to the left upper corner of the left atrium. The place of connection of the LSVC to the left atrium appears to be constant

and lies between the opening of the left atrial appendage anteriorly and slightly superiorly and the opening the left pulmonary veins posterioly and inferiorly. the pulmonary veins may enter the left atrium more superiorly than usual in this form of the syndrome[7].

The atrial septal defect (ASD) is seperated from the atrioventricular valve ring by a remnant of atrial septum (in contrast to an ostium primum ASD), while its inferior margin is formed by the atrial wall where it joins the inferior vena cava. There may be seperate forame ovale ASD or a single large confluent ASD, formed by cofluence of both defect. The coronary sinus ASD may be confluent with an ostium primum ASD, or there may be a common atrium. As the coronary sinus does not exist, the individual coronary veins drain seperately into the inferior aspect of the left atrium. Some also drain into the right atrium[7].

In our case, completely nuroofed coronary sinus was combined with common atrium, persistent LSVC and mitral valve cleft. We, therefore, also considered this as incomplete endocardial cushion defect with unroofed coronary sinus syndrome.

Usually, the diagnosis of unroofed coronary sinus syndrome is only suspected, not confirmed, before operation. However, the demonstration of an LSVC by catherter passage or cineangiogram provides suspicion of the diagnosis, which is confirmed, if the LSVC can also be shown to drain into the left atrium. Konstam and colleagues have pointed out that radionuclide angiography can be used to make the diagnosis, since intravencus injections into the left arm will show larger right-to left shunting than those into the right arm[8]. The EKG is not likely to be helpful in identifying the develcpmental complex of atrial septal defect and persistent left superior vena cava. Of the 8 patients reported by Raghib and his colleagues, 4 had a normal frontal QRS complex. Two patients with incomplete A-V communis had the EKG usual for that condition, indicating some dergree of left axis deviation. In 2 patients with associated VSD, the EKG demonstrated right axis deviation and right ventricular hypertrophy[4]. The chest X-ray may show an unusually straight left heart and mediastinal border.

Cyanosis from the right-to-left shunting dominates the clinical picture of isolated completely unroofed corcnary sinus with persistent LSVC and determines its natural history. The cyanosis has been mild in UAB (University of Alabama Medical Center, Birmingham) cases, all of whom were less than 17 years old, but was severe in some patients reported in the literature. Cerebral embolization and cerebral abscess complicate the life history in 10 %-25 % of such patients. This situation is similar to that in other type of right-to-left shunting. Presumably, life expectancy is considerably reduced by these complications and the other problems associated with increasing cyanosis and polycythemia[9].

In cyanotic patients with a communication between the left and right SVC, the LSVC was first tied off by Hurwitt and colleag ues in 1955[10] and then by Davis and colleagues[11]. Taybi and colleagues, in 1965, reported a ligation and mentioned "transferring the left SVC to the right atrium", but presumably this was unsuccessful, since no futher details are given[12]. The first report of successful repair was done by Rastelli and colleagues in 1963[13]. In this case, a tunnel was constructed from the posterior wall of the left atrium. In the same Jorunal[14], they described the insertion of a large pericardial atrial baffle that totally corrected anomalous connection of both the superior vena cava and inferior vena cava to a left-sided atrium. This procedure was also used by Helseth and Peterson, in 1974[5].

Of considerable surgical importance is the fact that the left innominate vein is absent in 80-90 % of cases with the unroofed coronary sinus syndrome and LSVC[9,13]. The right SVC is frequently small and it may be absent. The inferior vena cava not infrequently crosses to the left side below the diaphragm to enter the left hemiazygos vein, wh-

ich joins the LSVC. The hepatic veins usually enter the inferior aspect of the right atrium, but they too may enter a morphologically left atrium, total anomalous systemic venous connection is present[7].

The operaion may be done routinely with cardiopulmonary bypass at 25℃ and the usual direct caval cannulation; the return from the LSVC is picked up with a pump-oxygenator sump sucker. Alternatively, in infanacy a single venous cannula and repair during profoundly hypothermic total circulatory arrest may be used in GLH (Green Lane Hospital, University of Auckland)[7].

When the diagnosis of completely unroofed coronary sinus with persistent LSVC is made, operation is advisable because of arterial desaturation, the risk of cerebral emboli, and the good results of the rate isolated completely unroofed coronary sinus without persistent LSVC (coronary sinus ASD) and the same as other types of ASD[15]. When unroofed coronary sinus is associated with other major cardiac anomalies, the associated anomaly usually presents a clear indication for operation.

The risk of repairing unroofed coronary sinus syndrome, either isolated or combined with other specific malformations, is low, 1 death(4 %) having occurred in the 23 patients in the UAB experience and the GLH experience combined[7].

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