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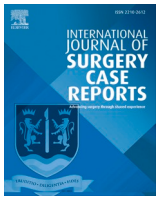

Case report

A Case Report of Recurrent Subaortic Membrane in a 3-Year-Old Saudi Child

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ABSTRACT

Introduction: While only a few cases have been reported in pediatrics, subaortic stenosis (SAS) is a gradually progressive disorder rarely seen at birth and infancy, however, it is the most common type of aortic stenosis. It obstructs the blood flow across the left ventricular outflow tract (LVOT). Although the cause is still not well known, different etiologies have been suggested by the literature. While surgical resection is the definitive treatment, recurrence is observed in many patients, nonetheless, LVOT gradient usually progresses over years of follow-up.

Case presentation: We report the clinical and diagnostic course of a 41-months-old Saudi boy, asymptomatic child who was found to have progressive recurrent subaortic stenosis within a few months which required two redo sternotomy for sub-aortic membrane resection throughout a period of two years.

Discussion: SAS is usually detected incidentally in asymptomatic patients requiring an echocardiogram to assess other accompanying congenital heart defects (CHD), or rather potentially arising after repair of CHD. Patient close monitoring is important aspect given the nature of disease progression, re-operation for recurrence demonstrate significant increase over years, re-resection rate was 0 % after one year, 6 % after five years, and 8 % after 10 years.

Conclusion: Recurrence of LVOT obstruction following sub-aortic membrane resection is common. Long-term follow-up care in postoperative patients is crucial. Majority of patients will need re-operation for recurrence at certain point during course of the disease.

1. Introduction

Subvalvular aortic stenosis (SAS), also referred to as subaortic stenosis, is a gradually progressive disorder rarely seen at birth and infancy. However, it is the most common type of aortic stenosis. It is characterized by a membrane below the aortic valve which obstructs the blood flow across the left ventricular outflow tract (LVOT) [1]. The cause is still not well understood. Genetic factors and hemodynamic abnormalities seen in other cardiac diseases, and surgical intervention are possible etiologies that have been suggested in the literature [2]. Epidemiologically, it accounts for 1 % of all congenital heart defects (CHD) (8 in 10,000 births) and 15 % to 20 % of all fixed left ventricular outflow tract obstructive lesions. In up to 60 % of cases, it is associated with other congenital cardiac defects, commonly ventricular septal defects (VSD) and patent ductus arteriosus (PDA). The majority of patients are asymptomatic, and it is discovered incidentally while evaluating for other cardiac disease; however, some cases can present with dyspnea on exertion, angina, effort syncope, orthopnea, and sudden cardiac death. [1]. Echocardiography is the test of choice for diagnosing SAS. It is treated by surgical membrane resection with or without septal myectomy. Recurrence is observed in many patients, however, LVOT gradient usually progresses over years of follow-up [3]. Presented at King Faisal Cardiac Center hospital, a 41-months-old boy with SAS post VSD, PDA, and coarctation of aorta (CoA) repair, who underwent sternotomy twice

Abbreviations: SAS, subvalvular aortic stenosis; LVOT, left ventricular outflow tract; CHD, congenital heart defects; VSD, ventricular septal defect; PDA, patent ductus arteriosus; CoA, coarctation of aorta; PG, peak gradient; AVSD, atrioventricular septal defect.

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for subaortic membrane resection due to rapid progression in LVOT gradient within a few months.

This work has been reported in line with the SCARE 2020 criteria [4].

2. Case presentation

A 41-months-old boy was referred to the cardiac clinic at 27-day-old due to a large outlet VSD of 8 mm with inlet extension, small aortic isthmus peak gradient (PG) 20 mm Hg, PDA, and CoA, all of which were successfully treated surgically. At the age of 5-month-old, during routine follow-up echocardiogram revealed an incidental finding of severe sub-aortic membrane with peak LVOT PG of 85–95 mm Hg and mean gradient of 44 mm Hg (Fig. 1). The patient underwent redo sternotomy and sub-aortic membrane resection, and was discharged home in good condition after 7 days of admission. In the follow-up appointment, at the age of 10-month-old, patient's subsequent echocardiogram revealed a recurring sub-aortic membrane, peak LVOT PG of 70 mm Hg and mean gradient of 42 mm Hg.

The pediatrics cardiac team planned to arrange a second operation after one month, however, the surgery was postponed due to COVID-19 pandemic situation. The echocardiography at this time showed disease progression, severe LVOT obstruction, peak LVOT PG reached 85 mm Hg (Fig. 2). At an age of 27-months-old, the patient underwent his second redo sternotomy, this time with extended resection of sub-aortic membrane. Apart from tension pneumothorax that was resolved with chest drain, the surgery went smoothly with no other post-operative complications.

After six months later, the echocardiogram showed an increased subaortic valve velocity with peak LVOT PG of 42 mm Hg. Additionally, the patient had no symptoms and was growing normally throughout the course of the disease.

Currently, at 41-months-old, an echocardiography revealed that the subaortic valve velocity had attained its maximal peak LVOT PG of 67 mm Hg; mean gradient of 40 mm Hg. Echocardiography also showed mild tricuspid regurgitation, good left ventricular function, and with no pleural effusion. The patient was asymptomatic, active, not cyanotic, and had no dysmorphic traits when examined. Developmentally, he was up to his age, and both of his weight and height were in the 90th percentile. Cardiac auscultation revealed a 3/6 harsh ejection systolic murmur that was most audible in the right second intercostal area. Upon examining other systems, there were unremarkable findings. Supine chest X-ray was done and showed mild Cardiomegaly, and healthy-appearing lungs. Case was discussed during pediatric cardiac team meeting, and the patient was planned and booked for Modified Konno Operation.

3. Discussion

SAS in most cases (up to 60 %) is associated with congenital heart defects, such as VSD, atrioventricular septal defect (AVSD), or conotruncal anomalies [1]. It may also develop after repair of a perimembranous or malaligned VSD or AVSD, as seen in our patient [3].

The pathophysiological theory explaining the development of SAS lesions was first described by Roenqist et al. that an increase in mitral aortic separation could contribute to the mechanism of SAS development. Other explanations included preexistent morphological abnormality or possible genetic components that have yet been well understood [3].

Patients with mild to moderate obstruction are typically

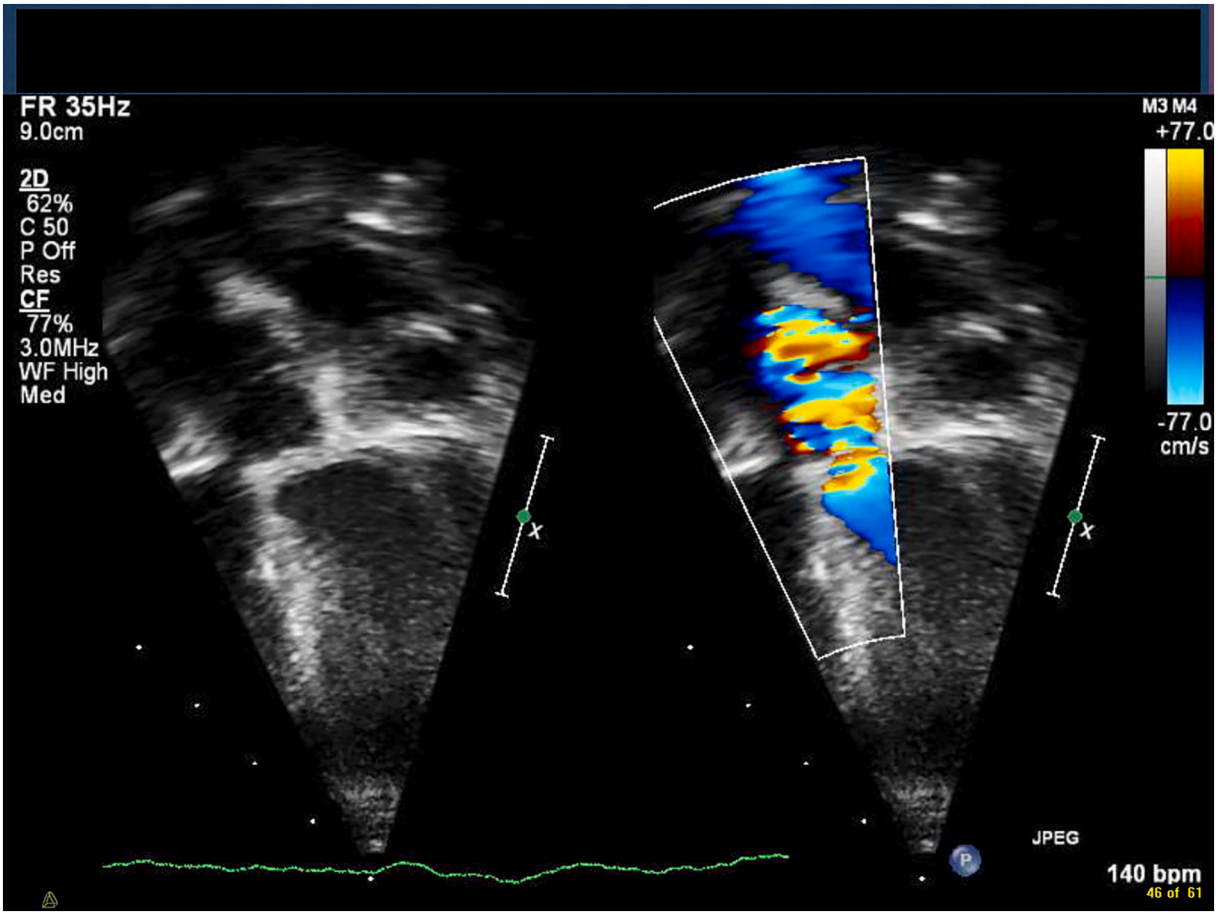

Fig. 1. Last echocardiogram done before first procedure; showed subaortic membrane with peak LVOT PG of 93 mm Hg; mean of 44 mm Hg.

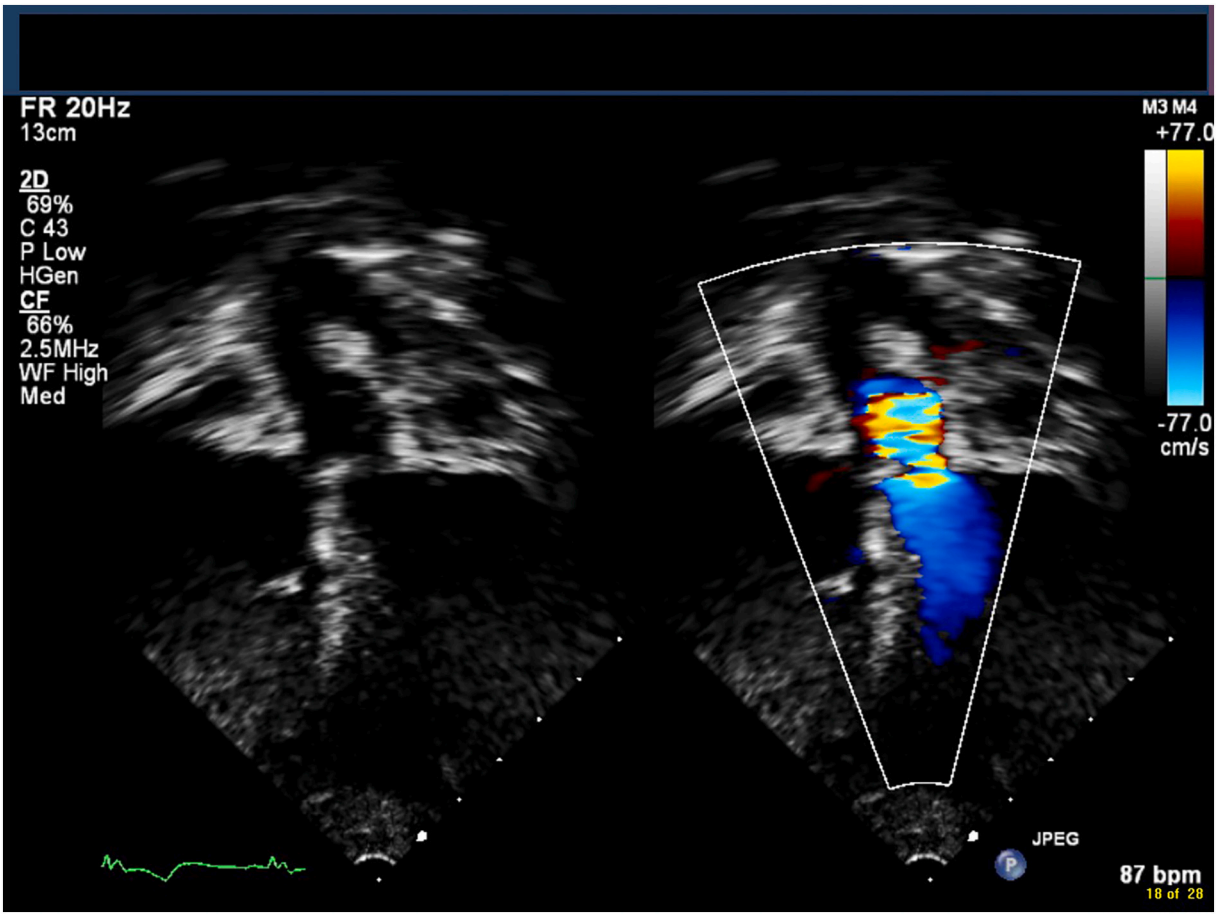

Fig. 2. Last follow-up echocardiogram before second operation; showed subaortic membrane with severe LVOT obstruction, peak LVOT PG of 85 mm Hg.

asymptomatic [1,3]. As in our patient, the lesion was incidentally found by echocardiogram for follow-up evaluation after the surgical repair of VSD and CoA.

As for the diagnosis, echocardiography is the current modality of choice to establish a diagnosis of SAS. It is used to characterize the subaortic lesion, and estimate the gradient and therefore the extent of obstruction across the LVOT [3]. Signs of left ventricular hypertrophy are usually seen in an electrocardiogram, and cardiac catheterization is not usually performed [1,3].

SAS surgical intervention is the definitive therapy that can involve simple membrane resection with or without septal myectomy, or a Konno procedure that is performed by ventricular septum incision and patch augmentation. The mortality rate is reported to be very low after surgery [1,5].

A retrospective chart review of a single center of 104 patients from 1991 to 2015 who have had surgical resection of sub-aortic membrane stenosis found that 3 patients required pacemaker, 9 patients required repeat resection for recurrence of sub-aortic membrane. The findings of transthoracic echocardiogram were studied and then follow-up echocardiogram was done. The median time of follow-up was 8.5 years. The surgical approach that was followed was LVOT via oblique aortotomy then blunt and sharp resection of the sub-aortic membrane. Re-resection rate was 0 % after one year, 6 % after five years, and 8 % after 10 years. [6]. However, even in recurrent cases, repeated surgery can be well tolerated and may be accomplished with no risk of complications, as observed in our case.

Furthermore, regarding the outcome in children operated for membranous subaortic stenosis, a retrospective review of patients followed at a pediatric cardiology clinic of university hospital during the period January 2002 till December 2013 compared those with membrane resection plus aggressive septal myectomy vs membrane resection alone. The total number of patients who went through subaortic membrane surgery was found to be 46. 19 cases had membrane resection and aggressive septal myectomy, and 27 patients had membrane resection alone. The Preoperative diagnosis was at the pediatric cardiac clinic of Addia Ababa University Hospital, but the surgery was done in different organizations. An echocardiogram was done on all patients. The surgical approach for subaortic fibromembranous in all cases was aortotomy incision. No postoperative complications were identified in any of the groups. The mean follow-up period was 2–11 years in the group of membrane resection and 1–6 years in the group that had membrane resection plus aggressive septal myectomy group. Moreover, 9 patients out of the group who had membrane resection-only showed significant regrowth of the sub-aortic membrane. However, no cases of recurrence were reported in those who had membrane resection plus aggressive septal myectomy. LVOT obstruction relief and aortic valve function improvement were also seen in patients who underwent membrane resection plus aggressive septal myectomy [7]. Moreover, a retrospective cohort study of patients who had resection of subaortic membrane at Boston Children's Hospital from 1984 till 2016 studied the reintervention of recurrent subaortic stenosis post discharge. The study formulated a prediction rule of recurrence that was based on multiple factors which include patient's age, preoperative LVOT, peeling of mitral valve membrane, distance from the membrane to the aortic valve, prior complex surgery, and other left heart lesions [8]. Furthermore, a retrospective study from 2006 till 2014 in Philadelphia Children's Hospital discussed the echocardiographic predictors of recurrent LVOT obstruction in children who have undergone subaortic stenosis resection. The criteria for recurrent LVOT obstruction was defined as increase in mean pressure gradient by more than 15 when compared to the most

recent follow-up echo or having a value of more than or equal to 20. Thirty patients were studied and 10 of them had recurrent LVOT obstruction. The drawn conclusion of the study is that LVOT obstruction recurrence after subaortic resection is common, residual LVOT obstruction may either increase or stay the same over time, and the closer the LVOT obstruction to the aortic valve the higher the risk of recurrence [9].

4. Conclusion

Subvalvular aortic stenosis is a gradually progressive disorder that consists of a fibrous membrane in the LVOT. Recurrence of sub-aortic membrane is not uncommon and has been seen in many patients over years of follow-up. However, as reported, our patient had rapid progression of sub-aortic membrane and frequent recurrence over a short period of time which required sternotomy twice. The case underlines the importance of close monitoring of LVOT gradient in patients with SAS for evaluation of surgical management. Recurrence and re-operation remain high, in which close long-term follow-up is warranted, and detailed knowledge of surgical procedures and their efficacy in reducing the rate of recurrence is desired.

Consent

Verbal consent obtained from the patient's guardians.

Ethical approval

An ethical approval was obtained prior to manuscript writing from the Bioethics Committee in King Abdullah International Medical Research Center (approval no. NRJ21J/274/11). Furthermore, patients' data were secured and used for the study purposes only.

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Guarantor

1) Loujen O. Alamoudi
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Research registration number

1. Name of the registry: Nil
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CRediT authorship contribution statement

LA, MA, and EA collected, assembled, and clinically evaluated the patient's interpreted data and wrote the manuscript. SA, AG did the final edit of the manuscript and initiated submission for publication. RW helped critically in revising the manuscript. All authors read and approved the final manuscript.

Conflict of interest

The authors certify that they have NO affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

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