Isolated Supravalvular Aortic Stenosis with Infective Endocarditis presenting as Pyrexia of Unknown Origin

Deepak Kumar Mishra¹, Vishal Khullar², Shalima Gautam¹, Tamanna Khullar³

Abstract
Supravalvular aortic stenosis is a less common form of left ventricular outflow tract obstruction (LVOTO); commonest being the valvular aortic stenosis followed by valvular and subvalvular forms respectively. Most of the supravalvular aortic stenosis is associated with Williams syndrome; isolated supravalvular aortic stenosis is further rarer. We present a case of isolated SVAS with infective endocarditis (1.6) as the cause of pyrexia of unknown origin (PUO).

Case Report
A 25 years female presented with ECG features suggestive of LVH. She was running fever since 6 weeks. Fever was moderate grade intensity and used to occur with chills. There was associated history of weight loss and decreased appetite. She was prescribed paracetamol and oral antibiotics outside without any improvement. Patient was admitted for evaluation. She was tachycardic with BP of 130/90 mm Hg in right arm and BP of 116/88 mm Hg in right upper parasternal region. S4 was palpable at apex. She had harsh ejection systolic murmur peaking late in systole with maximum intensity on right upper parasternal region and with more radiation to right side of clavicle as compared to left. There was no radiation to apex. There was an early diastolic murmur at the aortic area occupying more than half of diastole which was maximum in sitting with leaning chest position. S4 was audible at apex. ECG showed LVH. X-ray was normal. 2D echocardiography revealed severe LVH, mild LV dysfunction with EF 45 %, supravalvular aortic stenosis with peak gradient of 100 mm Hg. There was moderate valvular aortic regurgitation. Other valves were normal. She had a vegetation at the site of origin of brachiocephalic trunk. Blood investigations ESR was 50 mm for 1 hr. Blood counts were normal. Blood cultures grew streptococcus viridians with colony count of 50,000/ cu.mm. Other blood parameters were normal. CT angiogram showed narrowing of ascending aorta including the sinotubular junction and part of ascending aorta (Figures 1-3). The patient was started on inj. Ceftriaxone 1gm IV 12 hrly for 6 weeks and inj. Gentamicin 40 mg OD for 2 weeks. The patient responded well to the antibiotics course. Two weeks after the therapy conventional cardiac catheterization as done which confirmed the findings of echo and CT angiography. Coronaries were dilated but there were no luminal obstruction. She was taken up for definitive surgery: Dacron patch repair with aortic root enlargement (Figures 4 and 5) and aortic valve replacement (18 mm ATS valve) which went uneventful and was discharged on 7th postoperative day. The patient is doing well in follow-ups with INR in therapeutic range.

Discussion
PUO has varied definition depending on the duration of fever and the type of set up i.e nosocomial, neutropenic or in HIV / AIDS patients Commoner cardiac condition manifesting as fever are infective endocarditis, pericarditis, left atrial myxoma, Takayasu arteritis, cardiac sarcoidosis, myocardial tuberculosis/granulomas. Infective endocarditis is a dreaded disease with an incidence of 1.7 – 6.2 cases per lac patient year. Males are twice as commonly affected as females.¹² Common predisposing factors are rheumatic heart disease, congenital heart disease, prosthetic heart valve patients and intravenous drug users/ abusers. It has acute (fulminant) presentation or subacute presentation. The mortality as studied in Indian patients are as high as 23 % as mentioned by Garg et al¹ and Choudhary et al.¹

Criteria for diagnosing I.E have evolved since the Von Reyn criteria initially described for infective endocarditis followed by Duke’s criteria to recent modified Duke’s criteria. Serum procalcitonin level greater than 2.3 ng/ ml has been predicted to diagnose I.E with almost 85 % specificity.²

With the introduction of new guidelines for antibiotic use for the prevention of IE introduced by ACC/ AHA in 2008 all the recommendation which were class I were changed to class 2a and it clearly mentions that rheumatic valvular heart disease doesn’t require any antibiotic prophylaxis as long as antisepsis is not breached.

And these indications are :
1. Patients with previous prosthetic cardiac valve, prosthetic material used for cardiac repair.
2. Patients with previous IE.
3. Patients with congenital heart disease: unrepaired cyanotic, palliative shunts / conduits, completely repaired with prosthetic material, device closure for 6 months. Repaired congenital heart disease with residual defects.

¹Intervention Cardiologist, ²Consultant Cardiothoracic Surgeon, ³Consultant, Dept. of Non-invasive Cardiology, Bombay Hospital Institute of Medical Sciences, Mumbai, Maharashtra
Received: 29.11.2014; Accepted: 30.03.2017
4. Cardiac transplant recipients with valve regurgitation due to structurally abnormal valve.

The exact incidence of isolated supravalvular aortic stenosis (SVAS) is not known. SVAS in association with Williams-Beuren syndrome (chromosome 7) is commoner than the isolated forms. Williams syndrome is either familial or sporadic. Syndromic association include bicuspid aortic valve, Mitral valve prolapse, coarctation aorta, pulmonary stenosis, extracardiac manifestation like diabetes mellitus, hypercalcemia, hypothyroidism, typical Elfin facies (God like), denture abnormality, hearing abnormality, gastrointestinal problems, skin abnormalities, genitourinary problems, psychosomatic complaints and attention deficit hyperkinetic disorders. Our patient had no other feature suggestive of William syndrome except SVAS.

SVAS can present as breathlessness, angina, presyncope, Syncope. It can either progress (mostly in initial five years) or remain the same unlike the pulmonary stenosis component which usually regresses.7,8

One peculiar thing about SVAS is that the risk of sudden cardiac death is low unlike other forms of LVOTO. But this risk increases significantly if pulmonary stenosis is also present alongwith right ventricular hypertrophy.9

Treatment option for SVAS is mostly different forms of surgical correction depending upon the different types of obstruction: hourglass, multiple stenosis, diffuse hypoplasia. It can be either end to end resection, vertical incision with Dacron patch repair, etc. Balloon angioplasty has poor result in such pts. as media of aorta has plenty of smooth muscle in the disease (mutation of Elastin ELN gene on chromosome 7). On the contrary, balloon angioplasty has good result for other vessels like pulmonary arteries.

Our patient was treated with intravenous antibiotics: Inj. ceftriaxone 1 gm IV BD for 6 weeks and Inj. Gentamicin 1mg/kg for 2 weeks. The vegetation at brachiocephalic trunk was due to selective streaming of jet towards the right side of aorta and its branches (Coanda effect). This phenomena also describes the selective radiation of systolic murmur to right side of trunk in SVAS.
Cardiac catheterization (Figures 1, 2) was done to corroborate the 2D echocardiography and CT Angiogram (Figure 3) findings. The patient had severe aortic regurgitation also and severe left ventricular hypertrophy with mild left ventricular dysfunction (EF 45%). So patch repair of aorta (Figures 4, 5) along with prosthetic aortic valve size no 18 mm was done.

The operation was uneventful; so was the recovery and the patient was discharged on 7th postoperative day and is doing well in follow up.

**Conclusion**

Infective endocarditis complicating supravalvular aortic stenosis can be a rare but potentially treatable cardiac cause of PUO.

**References**