Available online at www.rajournals.in

RA JOURNAL OF APPLIED RESEARCH
ISSN: 2394-6709
DOI:10.47191/rajar/v7i9.08
Volume: 07 Issue: 09 September-2021

International Open Access

Impact Factor- 7.036

Page no.- 2532-2534

Dyspnea in an Adult with Atrial Septal Defect and Sjôgren's Syndrome: Who's to Blame?

Z. Mehssani MD[1], R. Fellat PR[2], N. Fellat PR.[3]

[1,2,3]Department of Cardiology, A/National Cardiovascular League, Ibn Sina University Hospital Center, Mohammed V University, Rabat, Morocco.

ARTICLE INFO

Published Online
25 September 2021

Corresponding Author:
Z. Mehssani MD

ABSTRACT

ASD and more frequently the ostium secundum type is a relatively common adult congenital heart disease. Sjogren's syndrome on the other side is the second most frequent auto immune disease, Involving the lungs in 9–20% of patients. We report the case of a 69 years old woman, complaining of a long-standing dyspnea and was diagnosed with Sjogren's syndrom. Her CT scan and pulmonary function tests were normal. That lead to investigate for pulmonary hypertension as a potential cause for her dyspnea. A TTE was performed and estimated her systolic pulmonary pressure at 30 mmhg, and showed an OS ASD with left to right shunt. Right heart catheterization calculated her mPaP at 20 mmhg. She is scheduled for percutaneous closure of her OS ASD. There are only few cases reporting an association between ASD and Sjogren's syndrome in polyendocrine syndrome, and even fewer without polyendocrine syndrome. Farook and Al. discussed a case of a women presenting OS ASD with SS complicated with PAH, suggesting an association between the two pathologies by inflammatory process, and Inoue et Al. presented a case of a women with polyartritis rheumatoid and Sjogren's syndrome complicated with PAH, inducing a right to left shunt through a patent foramen oval. In our case, the ostium secundum type ASD with left to right shunt with mild pulmonary hypertension is probably the reason for her dyspnea, as there was no interstitial lung disease with normal CT and PFT. Unfortunately, there is no sufficient data suggesting an eventual association between ASD and Sjogren's syndrome without polyendocrine syndrome. The learning point is the importance of screening for congenital heart disease in adult with unexplained dyspnea.

KEYWORDS: Adult Congenital Heart Disease; Atrial Septal Defect; Sjogren's Syndrome; Pulmonary Arterial Hypertension

INTRODUCTION

Atrial septal defect and more frequently the ostium secundum type is a relatively common adult congenital heart disease. Sjogren's syndrome on the other side is the second most frequent auto immune disease, involving the lungs in 9–20% of patients. We report in this case to describe the possible association and interaction of Sjogren's syndrome and atrial septal defect.

CASE REPORT

We report the case of a 69 years old woman, complaining of a longstanding dyspnea stage II NYHA, since the age of 12, and had seen multiple specialists and pulmonologist putting her under salbutamol with no significant improvement. 9 years ago, she started to present inflammatory joint pain at the knees and elbows and goes to a rheumatologist. At the physical exam he noticed and asked for dryness of the mouth and eyes which were present. Blood labwork was done which came back positive for antibody Ro, with normal sedimentation rate, CRP, and white blood. This was completed with labial biopsy and was consistent with Sjogren's Syndrom. She was then refered to a pulmonologist to search for the causing of her dyspnea, as it is known that Sjogren's syndrome causes pulmonary manifestations. Pulmonary functional test and the pulmonary CT scan came back normal, but the angio CT scan objectified the atrial septal defect.

She was then admitted in our cardiology department for evaluation and eventual closure of the atrial septal defect. At the physical exam, cardiac auscultation was normal with no mesosystolic murmur, no sign of right heart dysfunction, no clubbing, and pulmonary exam was normal. The EKG did not show a RBBB, no flutter or auricular fibrillation.

A transthoracic echocardiography was performed and estimated her systolic pulmonary pressure at 30 mmHg, and

showed an ostium secundum atrial septal defect with left to right shunt. The right heart catheterization found a mean pulmonary pressure at 20 mmHg, and Qp/Qs at 2.21, and there was no anomalous pulmonary venous connection. She is scheduled for percutaneous closure of the ostium secundum atrial septal defect.

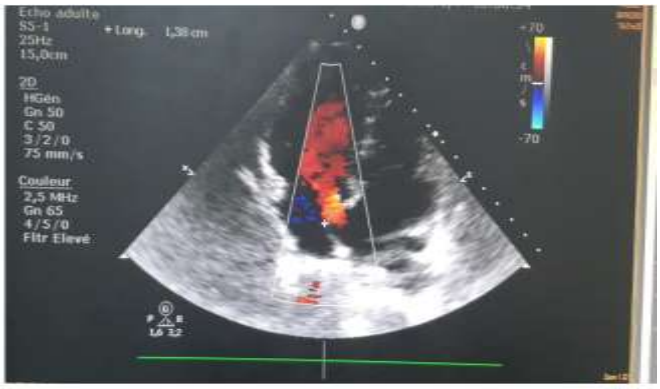

Figure 1: TTE with color Doppler showing the atrial septal defect.

DISCUSSION

The ostium secundum atrial septal defect is the most common type of atrial septal defect, accounting for 75% of all ASD cases, representing approximately 7% of all congenital cardiac defects and 30 - 40% of all congenital heart disease in patients older than 40 years. [1] Many patients with ASDs are free of symptoms for many years, although most will become symptomatic at some point in their lives. The age at which symptoms appear is highly variable and is not exclusively related to the size of the shunt. Exercise intolerance in the form of exertional dyspnea or fatigue is the most common initial presenting symptom. [2] On the other hand, primary Sjögren's syndrome was originally described in 1926 by GOUGEROT. Its prevalence is estimated at 0.5% with a female preponderance. It is the second most common multisystem autoimmune disease after rheumatoid arthritis, and is characterized by eye and mouth dryness and lymphocytic infiltration of the salivary glands. In addition to dryness, clinical presentation of Sjögren's syndrome generally includes asthenia and arthralgia. The disease can extend and systemic manifestations including vasculitis, lung, renal, cardiac or neurological involvement can occur. The cardiac manifestations are mostly pericardial effusion, left ventricular diastolic dysfunction, pulmonary artery hypertension, left atrium enlargement and mitral insufficiency. [3] The pulmonary manifestations of Sjögren's syndrome include airway abnormalities, interstitial lung disease (ILD) and lymphoproliferative disorders. Lung involvement occurs in about 9–20% of patients. Subclinical lung disease is even more frequent, including small airway disease and airway inflammation. The presence of ILD is associated with impaired respiratory function [4]. This is why in our case with the normal respiratory function has led to search for another cause of her dyspnea, and was not consistent with pulmonary manifestation of Sjogren's syndrome. There are only few cases reporting an association between ASD and Sjogren's syndrome in polyendocrine syndrome [5], and even fewer without polyendocrine syndrome.

Inoue et al. presented a case of a woman with polyartritis rheumatoid and Sjogren's syndrome complicated with PAH, inducing a right to left shunt through a patent foramen oval. Farook et al. discussed a case of a woman presenting ostium secundum atrial septal defect with Sjogren's syndrome complicated with pulmonary arterial hypertension, suggesting an association between the two pathologies by inflammatory process. Buyon et al. proposed the possible mechanism of an association of atrial septal defect in primary Sjogren's syndrome in infants. According to them, intracellular SSA/Ro antigens are translocated to the cell surface secondary to physiologic apoptosis of cardiocytes. This enable circulating antibodies, most commonly IgG, to bind to the antigen. Apoptotic cardiocytes are then phagocytosed by macrophages causing release of inflammatory cytokines favouring the differentiation of fibroblasts into myofibroblasts that promote irreversible scarring. Due to prolonged physiologic stress and myogenic muscular action of the heart, septal defect formation is possible. The mecanism proposed by Buyon et al. in infants with primary SS may have merits but its relevancy in adult patients of ASD with primary SS is unknown. [6] In our case, the ostium secundum type ASD with left to right shunt with mild pulmonary hypertension is probably the reason for her dyspnea, as there was no interstitial lung disease with normal CT and PFT.

CONCLUSION

Unfortunately, there is no sufficient data suggesting an eventual association between ASD and Sjogren's syndrome without polyendocrine syndrome. The learning point is the importance of screening for congenital heart disease in adult with unexplained dyspnea.

REFERENCES

1. Farook Ahmad, Muhammad Athar Sadiq, Kok Han Chee, Ahmad Syadi Mahmood Zuhdi and Wan Azman Wan Ahmad Atrial Septal Defect Coexistent with Sjogren's Syndrome Journal of the College of Physicians and Surgeons Pakistan 2014, Vol. 24 (6): 441-443
2. Gary Webb, MD; Michael A. Gatzoulis, MD, PhD Atrial Septal Defects in the Adult Recent Progress and Overview (Circulation. 2006;114:1645-1653.)
3. Yi-cong Ye, Yong Zeng, Wen-ling Zhu, Yan Zhao, Xiao-feng Zeng, Shu-yang Zhang, Quan Fang, Xiao-meng Li : Cardiac manifestations of Sjogren

Syndrome, Zhonghua Xin Xue Guan Bing Za Zhi. 2008 Apr, 36(4):327-31.

4. Thomas Flament, Adrien Bigot, Benjamin Chaigne, Helene Henique, Elisabeth Diot and Sylvain Marchand-Adam : Pulmonary manifestations of Sjogren's syndrome, Eur Respir Rev 2016; 25:&&0-123.
5. D.Y.AKSOY, MD; K.AG BAKT MD; O.HARMANCI MD; O.KARADAG MD; K.AYTEMIR MD; A.SUNGUR MD; A.M.ONAT MD; S.APRAS MD. B.O.YILDIZ MD; M.BAYRAKTAR MD; Autoimmune Polyendocrine Syndrome With Atrial Septal Defect, The American Journal Of The Medical Sciences, Feb 2008 Vol 335 Number 2.
6. JP Buyon, A Rupel and RM Clancy Neonatal Lupus Syndromes; Lupus (2004) 13, 705-712