Case Report

A 20 year old patient reared as a male presented to endocrinology OPD with history of left inguinoscrotal swelling of 1 month duration and gynaecomastia (Figure 1) of 6 months duration. At the age of 2 years, patient’s mother had noticed two streams of urine. He underwent two corrective surgical procedures, between 8 and 10 years of age following which he became asymptomatic. There was no significant contributory family history. Due to poor scholastic performance he discontinued studies after class 7. His gender identity, role and sexual orientation were like a male. There was no history nocturnal penile tumescence.

On examination: Vitals were normal. Height: 175 cm, Arm span: 184 cm, Upper segment: 86 cm, Lower segment: 89 cm, US: LS: 0.87, Weight: 60 kgs, BMI: 19.60 kg/m2. SPL – 8 cm, 2 opening visible on ventral aspect, right testicular volume – 8 ml. Left inguinoscrotal swelling: Two separate swellings palpable–Upper swelling 2 x 2 cm, soft, non-tender, extending from left inguinal region up to base of scrotum with positive cough impulse. Lower swelling 1x1 cm, fluctuating, non-tender. His SMR A3, P4. He had grade 4 gynaecomastia (Figures 1 and 2). Systemic examination was within normal limits.

Investigations

CBC : Hb : 12.4 gm/dl, TLC : 9610 (4000-11,000), DLC : Normal ESR : 23 mm at the end of 1 hour (0-20), Platelet count : 3,24,000 /mm3 (1,50,000-4,00,000), FBS : 80 mg/dl (70-110) PPBS : 85 mg/dl (70-140)

Hormonal profile: T3 : 1.32 ng / dl (1.23-3.23), T4 : 87.63 ug / dl(59-135), TSH : 0.899 uIU / ml (0.5-4.3), FSH : 10 mIU/ ml (1.5 – 12.4 ), LH : 12.06 mIU/ ml ( 1.7 – 8.6 ), Testosterone : 3.85 ng /ml ( 1.8 – 7.63), Estradiol : 40.49 pg / ml (< 39.8 )

USG pelvis: Well-defined hypoechoic area measuring 31 x 15 mm in left scrotum suggestive of ovary, Right scrotal sac shows testis measuring 25 x 11 mm. A well-defined cystic area adjacent to ovary measuring 11 mm.

MRI pelvis: Testes is seen in the scrotal sac on right side, inguinal hernia on left side with herniation of structure that looks like ovary. Inferiorly loculated fluid collection that measures 19 X 20 X 12 mm suggestive of encysted hydrocele. Uterus not identified; prostate not visualized (Figures 3 and 4).

Chromosomal and FISH analysis: Chromosomal analysis of cultured peripheral blood from male patient revealed female 46 XX chromosome complement (Figure 5). Patient sample confirmed presence of two X chromosomes and absence of SRY (Sex determining Region on Y chromosome) – Negative for SRY gene

Patient underwent surgery for left inguinoscrotal swelling. Surgical repair of hernia was done. Hernial sac examination revealed uterus, fallopian tube and ovary (Figure 6). He has been advised reduction mammoplasty for gynaecomastia.

Histopathology

Gross examination: Uterus with ovary with fallopian tube were seen. Uterus measured 5 x 2 x 2 cm ; endometrial thickness was 1 mm ; Left ovary measured : 3.5 x 1.5 x 1 cm ; fallopian tube was measuring 2 cm.

Microscopic examination: Sections from the uterus showed tubular endometrial glands. Sections from the ovary showed predominantly showed fibrotic areas. The fallopian tube showed mucosal plicae.

Discussion

Ovotesticular DSD is rare disorder. It has 3 types: 1. Lateral-(20%) have a testis on one side and an ovary on the other 2. Bilateral - (30%) have testicular...
and ovarian tissue present bilaterally, usually as ovotestes. 3. Unilateral-(50%) have an ovotestis present on one side and an ovary or testis on the other. It is a rare condition, with a frequency of 1:20,000-25,000 male newborns and was first described by La Chapelle and cols. in 1964. 2 On basis of the analysis and detection of SRY gene, 46, XX male patients can be clinically divided into SRY-positive and the SRY-negative group. SRY region was thought to be necessary for determining maleness. However, presence of normal males without SRY region on X chromosome proves that other factors apart from SRY also contribute to maleness of a person. 4

SRY-positive individuals: 90% of 46 XX ovotesticular DSD are SRY positive. Patients in this group have normal male genitalia, small azoospermic testes and hypergonadotropic hypogonadism. 3 Most carry the SRY gene translocated to X chromosome. 4 Diagnosis is achieved in adulthood during infertility investigation.

SRY- negative individuals – 10 % of 46 ovotesticular XX DSD are SRY negative. This group includes patients with ovotesticular DSD: presence of both testicular and ovarian tissue in the gonads of the same individual in absence of SRY on X chromosome. 5 Development of the testis and normal male genitals in SRY-negative 46,XX males suggests existence of other autosomal or X-linked genes in the sex-determining pathway. The maleness in the absence of SRY gene is explained by several mechanisms. The up-regulation or super-expression of some members of SOX family (SRY-related HMG-box) has been proposed as one of the mechanisms. McElreavey et al. (1993) proposed that SRY acts by inhibiting a regulatory autosomal recessive gene, termed ‘Z’, whose product normally inhibits the male pathway. 6 Two different studies have shown the duplication of a region of the long arm of chromosome 22 in a XX true hermaphrodite. 7

In this particular case, the diagnosis was done only when patient had inguinoscrotal swelling and gynecomastia at an adult age. In a recent case series of ovotesticular DSD from India 3 inguinoscrotal swelling and genital ambiguity before puberty and gynecomastia and cyclical hematuria after puberty were common presentations in ovotesticular DSD cases. Ovotesticular DSD should be considered as one of the differential diagnoses in any case of ambiguous genitalia with nonpalpable or asymmetrical gonads, pubertal gynecomastia, and cyclical hematuria.

Conclusion

46 XX ovotesticular DSD is a rare case. Such patients need timely diagnosis and management by a multidisciplinary team consisting of endocrinologist, pathologist, urologist, surgeon, psychiatrist and geneticist for better health.

Acknowledgements

We are sincerely grateful to Dr. K. M. Suryanarayana, Senior Professor and Head of Department of Endocrinology for evaluating this case.

References

Recurrent Prosthetic Pulmonary Valve Endocarditis in Repaired Tetralogy of Fallot

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Abstract
25 year old male who was a known case of repaired Tetralogy of Fallot with history of early prosthetic pulmonary valve fungal endocarditis in 2012 presented in 2016 with history of prolonged fever. On subsequent work up, he was diagnosed to have recurrent fungal prosthetic pulmonary valve endocarditis.

Introduction
Clinical diagnosis of prosthetic valve endocarditis is guided by the modified Duke criteria¹. Mostly prosthetic valve endocarditis in adults has been frequently described with prosthetic aortic or mitral valve. There is a paucity of data on prosthetic pulmonary valve endocarditis. We are presenting a case of recurrent fungal endocarditis of a Bioprosthetic pulmonary valve in an adult male who had a total correction of tetralogy of Fallot earlier in 1993. Cases of Prosthetic pulmonary valve endocarditis are increasing nowadays because of growing number of prosthetic valves being placed in repaired tetralogy of Fallot.

Case Report
25 year old male, presented to the Sir Ganga Ram hospital with One month history of Fever associated with 15 days history of Vomiting and pain abdomen. On examination there was presence of Pansystolic murmur in Tricuspid area with ejection Systolic Murmur and early Diastolic murmur in pulmonary area with splenomegaly.

He was a known case of Cyanotic congenital heart disease (Tetralogy of Fallot) which got total correction done in 1993. Patient had undergone Pulmonary valve replacement (Bioprosthetic) in 2012 due to development of free pulmonary regurgitation. Subsequently He developed early prosthetic valve fungal endocarditis. He was managed with I/V antifungals and fever subsided. Patient underwent echocardiographic examination which revealed vegetation attached to prosthetic pulmonary valve (Figure 1) and pulmonary regurgitation (Figure 2). Blood culture was positive for fungus Candida Albicans. Diagnosis of the prosthetic pulmonary valve endocarditis was made.

Discussion
Infective endocarditis means the infection of the cardiac valve or endothelium, which can be seen as vegetations. The common congenital heart anomalies predisposing to infective endocarditis are bicuspid aortic valve, Patent ductus arteriosus, Ventricular septal defect, Coarctation of the aorta, Tetralogy of Fallot etc.

Prosthetic valve endocarditis accounts for about 10% to 20% of all cases of infective endocarditis. The greatest risk of infection is in the first 6 months after valve implantation and appears to be similar in mechanical and bioprosthetic valves.

Fungi account for 10% to 15% of late prosthetic valve endocarditis cases and are associated with a higher mortality rate.² Prosthetic pulmonary valve endocarditis is a rare entity. In a large multicenter prospective 5 year observational study by Wang et al.³ 556 cases of prosthetic endocarditis were reported, out of which only 31(5.5%) cases had prosthetic pulmonary valve involvement. However, the increased number of cardiac surgeries followed by prosthetic valve implantation has led to their increasing incidence.

Conclusion
Prosthetic pulmonary valve endocarditis is a rare entity with an increasing incidence in the current era. High index of suspicion should be maintained for detecting it.

References

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Received: 15.11.2016, Accepted: 04.08.2018
An Unusual Site of Infective Endocarditis after Surgical Trauma-Evaluated by Three Dimensional Echocardiography

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Introduction

Infective endocarditis (IE) is an inflammation of endocardium and heart valves, associated with high risk of morbidity and mortality, prompt and early diagnosis and early treatment is essential. The age-specific incidence of endocarditis is 5 cases per 100,000 person-years among persons younger than 50 years to 15 to 30 cases per 100,000 person-years in the sixth to eighth decades of life.¹ Ventricular septal defect (VSD), patent ductus arteriosus (PDA) and bicuspid aortic valve (BAV) are common predisposing lesion for IE in adults. Isolated Atrial septal defect (ASD) associated with IE not reported in literature yet. We are reporting a rare case ASD associated with IE.

Case Report

A 43 year non diabetic, non-hypertensive, post-surgical closure of atrial septal (ASD) female patient presented with high grade fever from last one month. Her blood pressure was 130/86 mm of mercury and pulse rate was 104 minute, chest bilateral clear, no murmur, Abdomen and nervous system examination was normal. No significant abnormality detected in x ray chest PA view, total leucocyte count was 18000/cm³, 90 % neutrophil. All blood culture were negative. All other parameters were normal. Evaluation with two dimensional echocardiography (2D ECHO) reveals oscillating mass seen in right atrium (Figure 1A) attached to interatrial septum. which was not present in previous 2D ECHO before surgical closure. Trans esophageal echocardiography confirmed oscillating mass attached to opening of superior vena cava (Figure 1B) which was further confirmed by three dimensional echocardiography (Figure 1 C, D).

Discussion

Intact cardiac endothelium is resistant to bacterial invasion, damaged cardiac endocardium is strong stimulator for bacterial attachment leading to infective endocarditis. In VSD, PDA, BAV high velocity blood stream jet cause damage to endothelium in adult leading to IE. In literature most common site IE is valves (native or prosthetic), interventricular septum and intra-cardiac devices.² IE after surgery first reported by Taussig and associates in patients tetralogy of Fallot, assumed that unhealed suture line was the potential source of IE.³ Various case reports also described IE after surgical mitral commissurotomy.⁴⁻⁶ Hurst, Jones and Scott reported case of IE after PDA surgery.⁷⁻⁹ Uncomplicated ASD in adult never been reported in literature as associated with IE. But after surgical closure of ASD, normal endothelium could be receptive for bacterial adhesion due to unusual site surgical trauma, could be a source of prolonged fever. Prompt and early recognition and treatment result in excellent patient recovery.

Conclusion

Surgical trauma, could be a unusual site of infective endocarditis. Awareness of unusual site of infective endocarditis and early recognition result in excellent patient recovery.

References


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Received: 20.02.2018; Accepted: 14.08.2018
Myocardial Infarction following Organophosphorus Compound Poisoning

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Abstract
We report a 22 year old male who was admitted to our hospital with alleged history of consumption of monocrotophos poison and had presented with chest pain. His electrocardiogram (ECG) had showed ST segment elevation myocardial infarction and troponins were elevated. He also had low cholinesterase levels and was treated with pralidoxime and atropine and his condition improved. Cardiac catheterization showed patent coronaries. Acute coronary syndrome is a rare manifestation of organophosphorus compound (OPC) poisoning. The current case and subsequent review of literature tells us the need for close cardiac monitoring of all patients with OPC poisoning.

Introduction
OPC poisoning is very common in India where farmers form a significant proportion of the population who commonly use it as insecticides. OPC poisoning can cause cholinergic symptoms like salivation, lacrimation, urination and defecation. Nicotinic symptoms like neck muscle weakness, ocular weakness, proximal muscle weakness and respiratory muscle weakness can occur as a part of intermediate syndrome. ECG changes like transient ST-T wave changes, QT prolongation, atrial and ventricular arrhythmias can occur¹. Few cases of myocardial infarction (MI) after OPC poisoning has been reported. We report a young man who developed myocardial infarction after OPC (monocrotophos) poisoning.

Case
22 year old young man got admitted in our toxicology ward with alleged history of consumption of 15 ml of monocrotophos poison in his house. He was initially taken to the nearby private hospital where gastric lavage and activated charcoal was given. He had presented to the hospital with complaints of chest pain. Chest pain was left sided and diffuse and 8/10 in intensity. He also had shortness of breath at the time of presentation. No palpitation or syncope was noted. He also had increased salivation. Review of system was negative for other complaints. He had no significant past medical history. He was a nonsmoker and did not drink alcohol. He was not allergic to any medications. Physical examination revealed moderately built male. Cardiopulmonary examination was clinically normal. Abdomen was soft and he had bilateral constricted pupils on neurological examination.

Fig. 1: ECG of the patient showing ST-elevation in lead 2, 3 and AVF— Inferior wall myocardial infarction

Fig. 2: Coronary angiogram of the patient which reveals patent coronary vessels

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Received: 18.01.2017; Revised: 14.06.2018; Accepted: 24.05.2018
ECG which was taken revealed ST elevation in leads II, III, AVF (Figure-1). His vitals were stable. He was then referred to the government general hospital, Chennai. In our center, serum CPK-MB, troponins were immediately done which were elevated. Echocardiogram was done which showed regional wall motion abnormality in the inferior wall of the left ventricle. Serum cholinesterase levels were 1172 IU/dl which is low. Serum homocysteine levels, PT/INR, APTT, antithrombin, lupus anticoagulant and anticardiolipin antibodies were within normal limits. On the next day serum pro-NT BNP levels was done which was elevated. Patient was treated with pralidoxime, atropine, anticoagulant and antiplatelet drugs. Following this treatment, the patient’s serum cholinesterase levels improved, chest pain recovered. Coronary angiogram (Figure 2) was done the next day which was found to be normal. Patient’s medical condition improved and he was discharged.

Discussion

Cardiac complications often accompany poisoning with OPC. These may be serious and often fatal, being represented by cardiac arrhythmias, electrocardiographic abnormalities and conduction defects, as well as MI, a rarely reported complication of OPC poisoning. The extent and pathogenesis of cardiac toxicity from these compounds is not yet clearly defined. In literature we had few cases of MI occurring after OPC poisoning. Lionte C et al\(^1\) reported a 57 year old woman who developed anteroseptal MI and succumbed to death. Kiss Z et al\(^2\) reviewed 168 cases of OPC poisonings with special respect to frequent arrhythmias. In five patients a transient picture of MI was seen. Dayton S.B et al\(^3\) reported increased risk of MI among farm women exposed to pesticides. A rare case of MI due to parathion poisoning was reported by Yajneesh kidiyoor et al.\(^4\) The affected patient was a farmer from rural India who had succumbed to the complications of MI. Madhu Pankaj\(^5\) et al reported a 30 year old male who had taken chlorpyrifos and had presented with anterior wall myocardial infarction. Edibe Karasu\(^6\) et al also reported a 52 year old patient who had presented with inferior wall myocardial infarction after parathion ingestion. In patients with angiographically smooth coronary arteries, acetylcholine has been reported to produce both vasodilation and constriction. The development of vasoconstriction is likely to be an abnormality of endothelial function that precedes atherosclerosis or an early marker of atherosclerosis not detectable by angiography. This is a likely mechanism in our patient. Coronary vasoconstriction response in isolated perfused heart mediated by M 3 receptor has been reported in rats. The cardiovascular manifestations also reflect mixed effects on the autonomic nervous system. Increased sympathetic tone is often initially present and most patients manifest as sinus tachycardia and sometimes hypertension. As toxicity becomes more severe, bradycardia with a prolonged PR interval and atrio-ventricular blocks of various degrees occur because of excessive parasympathetic tone and possibly because of reduced coronary blood flow.

Conclusion

Cardiac complications often accompany poisoning with OPC, particularly during the first few hours. Hypoxemia, acidosis, and electrolyte derangements are major predisposing factors. Close monitoring in intensive or coronary care facilities with administration of antidotes in adequate doses early in the course of the illness will improve the outcome.

Conflict of interest

The authors of the paper declare that there is no conflict of interests involved regarding the publication of this paper.

Acknowledgement

We thank all the faculty of Institute of Internal Medicine, Madras Medical College for their kind help rendered in the evaluation of this case report.

References