Perioperative management of apical hypertrophic cardiomyopathy (Yamaguchi Syndrome) complicating pregnancy for emergency lower segment caeserean section

Sir,

Hypertrophic cardiomyopathy (HCM) is not uncommon in pregnancy but apical hypertrophic cardiomyopathy (AHCM) is a rare variety.[1]It is a benign, incidentally diagnosed condition with a 5% incidence.[2] A 22-year-old G2P1L1 presented to the emergency department (ED) with palpitations for one month, angina, breathlessness on minimal exertion (New York Heart Association grade III), and progressive limb oedema for 2 weeks, oliguria for 2 days and occasional dry cough for a few months. She had a history of similar complaints in her previous pregnancy 2 years back at the 8th month for which caeserean section was done under GA and postoperatively ventilated. The patient was lost to follow up. Her records were lost. Her family history was notable for sudden cardiac death in her grandfather at a young age.

At admission, the patient weighed 45 kg with a height of 150 cm (body mass index-20kg/m^2). She had bilateral pedal edema, elevated jugular venous pressure, heart rate of 116 beats/minute, respiratory rate (RR) of 20/min, blood pressure (BP) of 116/80 mm Hg, peripheral oxygen saturation (SpO2) of 75% on room air and 95% on O2 by mask (6 litres), loud first heart sound with a grade 3/6 mid systolic murmur, soft second heart sound and fourth sound, Decreased breath sounds on the left side and bilateral basal crepitation. Electrocardiography (ECG) revealed T wave inversion in leads I, II, V2-V6 [Figure 1]. Echocardiography revealed Hypertrophic Obstructive Cardiomyopathy (HOCM) with severe biventricular hypertrophy. The patient was initially managed by the obstetrician as HOCM by fluids and intravenous morphine. However, her condition worsened and anaesthesiologist and cardiologist opinion was sought. Our bedside echocardiography revealed HCM with asymmetrical septal thickness, predominant apical thickening more than 4 cm, left ventricular outflow tract (LVOT) gradient less than 10 mm hg, Mild tricuspid regurgitation, right ventricular systolic pressure 45 mm Hg, and grade 3 diastolic dysfunction [Figure 2]. The patient worsened and she was taken for emergency lower segment caeserean section because of fetal distress.

Preoperatively, she was administered a stat dose of IV furosemide 20 mg, ranitidine, and metaclopramide. Pre-induction arterial line and ultrasound-guided central venous line was inserted in the right internal jugular vein. On table, BP-130/80 mm Hg, PR-130/min, RR- 24/min, end-tidal CO2 – 60 mmhg, central venous

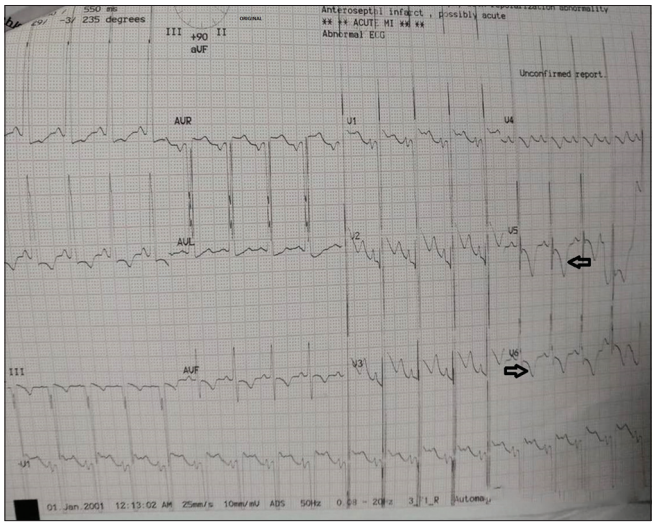

Figure 1: ECG: ECG shows deep T wave inversion in lead I, II and V2-V6 due to abnormal thickening of the cardiac apex and RVH and LVH strain pattern

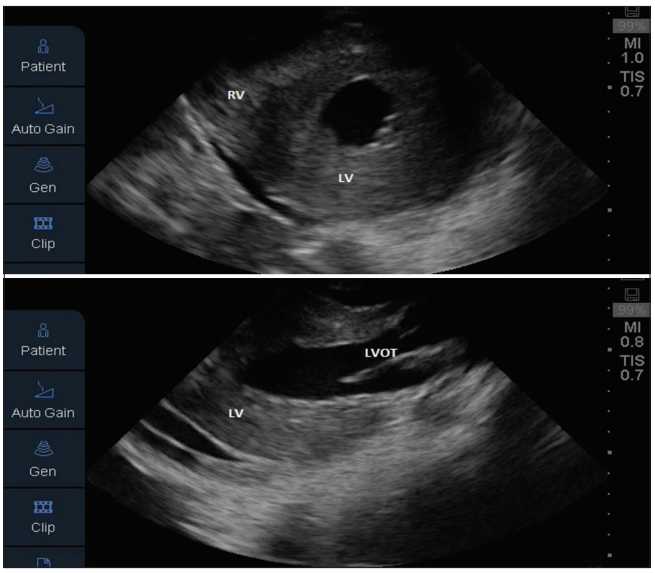

Figure 2: Top figure: Parasternal short axis view with LV hypertrophy with apical predominance. Bottom figure: Parasternal long axis view showing no signs of LVOT obstruction

pressure – 27 mm Hg. Preoperative arterial blood gas analysis showed severe uncompensated respiratory acidosis. The patient was preoxygenated with 100% oxygen for 3 minutes. Titrated doses of metoprolol up to 5 mg IV were given for ablation of intubation response. Modified rapid sequence induction (RSI) was done with intravenous fentanyl 50 μg, etomidate 15 mg, rocuronium 60 mg, intubated with 6.5mm endotracheal tube, connected to a ventilator with volume control mode tidal volume 350 ml, RR 20, inspiratory: expiratory ratio 1:2 and positive end expiratory pressure 5 cm. Peak pressure was 37 cm H_2O. Intravenous furosemide 20 mg was repeated. Maintenance was achieved with 100%O_2 and isoflurane 0.6%. Tachycardia was managed with metoprolol. After delivery, oxytocin infusion was started at 5 U per hour. Post-delivery air-oxygen ratio was kept at 40:60, isoflurane 0.5%, Inj.fentanyl 50 μg and Inj.midazolam 2 mg bolus was given. Peak pressure reduced to 22cm H2O and CVP was 12 mm Hg. No maintenance fluid was administered. Repeat ABG showed pH 7.54, pCO_2 32.5, pO_2 142.6, due to the hyperventilation provided. She was gradually weaned off ventilator support, extubated on postoperative day 2, and started on atenelol and intermittent non-invasive ventilation.

AHCM (Yamaguchi Syndrome), a rare variety of HCM with autosomal dominant inheritance pattern has a E101K mutation in the alpha actin gene.[2] Diagnostic parameters include asymmetrical left ventricular hypertrophy confined to apex, apical wall thickness ≥15 mm, ratio of maximal apical to posterior wall thickness ≥1.5, and ace of spade ventricular cavity.[3] AHCM can manifest as myocardial infarction, embolic events, AF, ventricular fibrillation, and congestive heart failure.[3] HCM presents due to diastolic dysfunction with well-preserved systolic function. The physiological changes during pregnancy worsen the diastolic dysfunction. Poor predictors for this patient included E/A >2 and CHF.[4] ECG changes in Yamaguchi syndrome include deep symmetrical T wave inversion which contradicted her prior diagnosis of HOCM. transthoracic echocardiography is a non-invasive modality for diagnosing AHCM, but is usually missed. Treatment goals include avoiding tachycardia, reducing after load, maintaining euvolemia, normal sinus rhythm, avoiding an increase in the contractility, and avoiding sympathetic stimulation.[2,5,6] Since she had presented with severe diastolic dysfunction, fluid restriction was advised whereas, in HOCM, liberal fluid therapy is required.[7] Because of tachycardia, metoprolol was given decreasing diastolic dysfunction and improving ventricular filling in non-obstructive HCM. Induction was done in this manner to avoid haemodynamic fluctuations. To prevent the worsening of pulmonary oedema, furosemide was given before delivery with fluid restriction. Treatment of refractory cases includes heart transplant and apical myomectomy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Noel Marie Pio Samy, Chitra Rajeswari Thangaswamy, Valliyoor Ramamoorthy Srivats, Thirumurugan Arikrishnan

Department of Anaesthesiology and Critical Care, Jawaharlal Institute of Postgraduate Medical Education and Research (JIPMER), Puducherry, India

Address for correspondence: Dr. Chitra Rajeswari Thangaswamy, Department of Anaesthesiology, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry, India.
E-mail: dr.chitrarajeswari@gmail.com

Submitted: 18-Jan-2020
Revised: 05-Apr-2020
Accepted: 11-Jun-2020
Published: 31-Jul-2020

REFERENCES

1. Payus A, Sholeh F, Mustafa N. Yamaguchi syndrome – A pseudoacute coronary syndrome of the young: A case report on apical hypertrophic cardiomyopathy. J Med Sci 2019;39:197-9.
2. Paluszkiewicz J, Krasinska B, Milting H, Gummert J, Pyda M. Apical hypertrophic cardiomyopathy: Diagnosis, medical and surgical treatment. Kardiochirurgia Torakochirurgia Pol Pol J Cardio-Thorac Surg 2018;15:246-53.
3. Diaconu CC, Dumitru N, Fruntelata AG, Lacau S, Bartos D. Apical hypertrophic cardiomyopathy: The ace-of-spades as the disease card. Acta Cardiol Sin 2015;31:83-6.
4. Moon J, Shim CY, Ha J-W, Cho IJ, Kang MK, Yang WI, *et al*. Clinical and Echocardiographic predictors of outcomes in patients with apical hypertrophic cardiomyopathy. Am J Cardiol 2011;108:1614-9.
5. Elliott PM, Anastasakis A, Borger MA, Borggrefe M, Cecchi F,

Charron P, *et al*. 2014 ESC guidelines on diagnosis and management of hypertrophic cardiomyopathy the task force for the diagnosis and management of hypertrophic cardiomyopathy of the European society of cardiology (ESC). Eur Heart J 2014;35:2733-79.
6. Juneja R, Nambiar P. Cardiomyopathies and anaesthesia. Indian J Anaesth 2017;61:728-35.
7. Choudhury M. Neuraxial anaesthesia in parturient with cardiac disease. Indian J Anaesth 2018;62:682-90.

Access this article online

Quick response code

Website: www.ijaweb.org

DOI: 10.4103/ija.IJA_14_20

How to cite this article: Samy NM, Thangaswamy CR, Srivats VR, Arikrishnan T. Perioperative management of apical hypertrophic cardiomyopathy (Yamaguchi Syndrome) complicating pregnancy for emergency lower segment caeserean section. Indian J Anaesth 2020;64:725-7.