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ANAESTHETIC IMPLICATIONS IN PATIENT WITH MAYER-ROKITANSKY-KUSTER-HAUSER SYNDROME

Medicine

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ABSTRACT

The mayer-rokitansky-kuster-hauser syndrome (MRKH) is a congenital condition consists of vaginal aplasia with other abnormalities. It occurs due to underdevelopment of Müllerian ducts during gestational period. Previously it was considered a sporadic Anomaly but now genetic etiology is being considered though specific gene is yet to be discovered. Patients usually undergoes puberty with normal thelarche and adrenarche. We present a case of 24-year-old female with the complaint of primary amenorrhea to be operated for vaginoplasty. We successfully managed the patient under spinal anaesthesia.

KEYWORDS

MRKH, primary amenorrhoea, vaginoplasty.

INTRODUCTION

Patients with Mayer-rokitansky-kuster-hauser syndrome presents a challenge to the anaesthesiologist as it may present with multiple malformations. It is a condition presenting with absence of uterus and vagina congenitally, it may be isolated (type 1) or it may present with other malformations (type 2)[1]. Type 2 MRKH syndrome may also be termed as MURCS (Mullerian duct aplasia, renal dysplasia and cervical somite anomalies). Incidence of MRKH syndrome is roughly around 1 in 4500 female births[2-4]. It is an autosomal dominant condition. Secondary sexual characters of the patient are normal and a normal 46, XX karyotype. Most commonly patient present with the complaint of primary amenorrhoea. These patients have normally functioning ovaries and normal external genitalia[5,6]. Patients may have associated renal malformations (unilateral renal agenesis, ectopic kidneys, or horseshoe shaped kidney)[7,8], skeletal abnormalities (asymmetric or fused vertebra, cervical scoliosis, klippel-feil associations, restricted neck movements, face and limb malformations or rib malformations)[9], auditory defects or deafness may be seen in some patients, its incidence is reported to be 10 to 25%[10,11]. MRKH syndrome may be associated with the cardiac anomalies such as aorto-pulmonary window, atrial septal defects, pulmonary stenosis or tetralogy of fallot[12]. Cardiac associations are less common but lethal. Psychological support is very important in such patients.

Case report

Twenty-four-year-old female suffering from MRKH syndrome posted for vaginoplasty. On Pre-anaesthetic check-up patient had history of asthma from 5 years for which patient was on Tab Montelukast and Foracort inhaler. Patient had history of depression from two and half years for which patient was taking Tab Aripiprazole, Tab trihexyphenidyl, Tab clonazepam. Other than this patient had a history of dyspnoea at two flight of stairs. Patient was able to carry out her routine chores. On examination there was facial asymmetry and cervical scoliosis. Chest wall deformity was present. Radial pulse in both hands were feeble on palpation and brachial pulse were of good volume. Patient had Mallampati grade 2. Neck extension was restricted but flexion could be done. Spine in lumbar region was comparatively less deviated than cervical-thoracic region. Breath holding time was 15 sec. Pulse rate was 84/min, regular and her blood pressure was 140/80 mm hg in left arm in sitting position. On auscultation bilateral air entry was present with no added sound. Heart sounds were heard normally with no added sounds. On investigations complete haemogram, renal function tests, coagulation profile were within normal limits (Hb-9gm%, PC-2.4 lac, INR-0.88, Na-137, K-3.9, BU-19, S.creatinine-0.8). On CECT abdomen she had ectopic fused kidney in left side. On echocardiography she had trace TR and trace MR with ejection fraction of 55%. Chest x ray showed bony cage deformity, cervical scoliosis and tracheal deviation to right side. Pulmonary function test showed FEV-1.33 L, FVC-1.71L and FEV1/FVC ratio of 78% post bronchodilator therapy. Patient was asked to continue her anti-depressant medications and was advised Tab Ranitidine 150 mg in HS and early morning. She was advised to stay nil per oral for 8 hours prior to surgery. Informed written high risk consent was taken. Patient was planned for surgery under spinal anaesthesia. On the day of surgery preparation for general anaesthesia was done. Difficult airway cart was kept ready. Surgeons presumed operative time was less than one hour. All the routine monitors were attached. Patient was cannulated with 20 gauze cannula. She was pre-loaded with 500ml of Ringer lactate. Under all aseptic precautions spine was palpated and 2ml of 2% lignocaine was instilled in skin and subcutaneous tissue in L3-4 space. Twenty three Gauze Quincke spinal needle was introduced in the same interspace but it was not successful as there was slight deviation of the spinous processes, then L2-L3 space was chosen and successful lumbar puncture was done, 2.6 ml of 0.5% Bupivacaine heavy was given. One milligram Midazolam IV was given to the patient. Oxygen was insufflated via face mask at 4 L per minute and etco2 monitoring was done. Spinal anaesthesia level was maintained at T10. Total fluid given intraoperatively was 1 L. Surgery was done in 45 minutes. No significant blood loss was present. Vitals were within normal limits intraoperatively. Patient was shifted to post anaesthesia care unit. Spinal anaesthesia regressed to Bromage score 1 postoperatively.

DISCUSSION

MRKH syndrome may present with multiple anomalies like in the above discussed patient. Hence one must always be aware of all the associated malformations. Effort tolerance of the patient must always be evaluated. In case of decreased effort tolerance pulmonary function test should be advised. Echocardiography should be done to rule out any cardiac defect. USG or CECT abdomen should be done to rule out ectopic or horseshoe kidney. A thorough examination of the spine should be done as cervical scoliosis may be present and fused wedged vertebra may be found in Lumbo-thoracic region. Lumber puncture may be difficult in these patients as was in this patient. Airway examination should be done properly. Under spinal anaesthesia proper pre loading and co loading of fluid should be done and any episode of hypotension should be avoided as patient may land up in pre-renal failure. Preparation for general anaesthesia with difficult airway cart should be kept ready. An anaesthetist should always be aware of possible malformations in the patient otherwise he may land up in unanticipated circumstances.

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