Anaesthesia in Turner's syndrome

V.M. Divekar MD DA (Lond),
M.D. Kothari MD DA DGO, B.M. Kamdar MD DA

A patient with Turner's syndrome anaesthetized on several occasions for laparoscopy, vaginoplasty and change of dressings is presented. Intubation was difficult due to webbing of the neck and a shortened tracheal length. Other problems in patients with this syndrome include difficulty in venipuncture, delayed excretion of drugs, associated cardiovascular anomalies and contraindication to regional blockade, in view of spinal deformities.

Key words

GENETIC FACTORS: Turner's syndrome,

COMPLICATIONS: Turner's syndrome, intubation.

In 1938 Turner described a syndrome of gonadal dysgenesis or ovarian agenesis in which the patients are of short stature though not actual dwarfs, the trunk is muscular, there is a webbed short neck, notable cubitus valgus and poorly developed breasts. Exaggerated epicanthic folds might be present. The vagina and uterus, if present, are under or mal-developed, hence the patient characteristically presents with primary amenorrhoea and sterility. Other gross congenital abnormalities may be present, such as coarctation of the aorta. Deformities of the digits may also be seen. Roentgenographic studies may detect cardiovascular or renal abnormalities, scoliosis, *spina bifida occulta*, or shortening of the fourth metatarsal and metacarpal bones. The incidence of Turner's syndrome is variously reported as 1:3000 to 1:10,000 live female births. The nucleus has only 45 chromosomes, i.e., 22 pairs of autosomes plus one instead of two sex chromosomes. A rare form of Turner's syndrome has also been described in the male.^{1,2}

From the Department of Anaesthesiology, B.Y.L. Nair Ch. Hospital, T.N. Medical College, Bombay 400 008, India, where correspondence should be addressed to Dr. Divekar.

Case report

A 22-year-old female with Turner's syndrome was scheduled for laparoscopy. She had a short webbed neck, shield-like chest, well developed secondary sex characteristics, right thumb deformity and absent right radial artery pulsation. The cervix and vagina were absent. Routine laboratory investigations were within normal limits, except for the electrocardiogram which showed right atrial enlargement, left axis deviation and clockwise rotation. X-ray of the chest showed scoliosis, cervical hemivertebrae and crowding.

Following premedication with atropine, a sleep dose of thiopental was administered for induction of anaesthesia. Ventilation by mask was possible without difficulty. The trachea was intubated following succinylcholine administration and the lungs were ventilated with nitrous oxide and oxygen using an Ambu-E-valve. Anaesthesia was supplemented by intermittent doses of meperidine and promethazine. It later became difficult to ventilate the lungs. On laryngoscopy it was seen that the upper border of the cuff was lying at the level of the epiglottis. An attempt to advance the tube resulted in diminished air-entry in the left lung, while slight withdrawal resulted in extubation. The tracheal tube length, from bevel to the teeth was only 13.5 cm whereas this distance is normally 20–22 cm. Laparoscopic findings were: absence of uterus, undescended right ovary and a streak-like left ovary. Postoperatively, air-entry to the left lung was reduced and a chest x-ray showed collapse of the left lung (Figure). The trachea was reintubated and suction yielded copious thick secretions. Following bronchoscopic suctioning, air-entry to the left lung improved clinically as well as radiologically.

The patient was further investigated before further surgery was undertaken. Intravenous pyelography showed an absent right and ectopic left kidney. The chest x-ray cleared. The tracheal bifurcation was seen to be at the level of the sternoclavicular joint, which is usually opposite the second thoracic

FIGURE Chest x-ray showing collapse of left lung and crowding of cervical vertebrae.

vertebra. General anaesthesia was administered for vaginoplasty. Following thiopentone and succinylcholine administration the trachea was intubated and respiration was controlled. The course of anaesthesia was uneventful.

On three subsequent occasions anaesthesia for change of vaginal dressings was uneventful.

Discussion

Patients with Turner's syndrome may present for anaesthesia for correction of the deformities of the reproductive system or for incidental surgery. The clinical features of importance to the anaesthetist are primary hypertension, short stature, high palate, micrognathia, short webbed neck and cardiovascular anomalies such as coarctation of the aorta and aortic stenosis. Urinary tract anomalies such as an absent kidney and peripheral lymph oedema may also be present.

The features encountered in our patient were short stature, primary amenorrhoea, sterility, short webbed neck, short right fifth metacarpal, absent right radial artery pulsation, cervical hemivertebrae

and crowding, scoliosis, shield-like chest, absent right kidney and ectopic left kidney. The uterus, cervix and vagina were absent, the right ovary was undescended and there was a streak-like left ovary. During the first anaesthetic there were two problems; difficulty in securing the tracheal tube to achieve bilateral air-entry, and the presence of troublesome secretions. The first problem was due to the anatomical abnormality of a short webbed neck, crowding of cervical vertebrae and bifurcation of the trachea at the level of the sternoclavicular joint. The problem of secretions and subsequent lung collapse may have been due to the atropine premedication or other agents rendering the secretions tenacious.³

References

- 1 Goodman RM, Gorlin RJ. The face in genetic disorders. St. Louis. The C.V. Mosby Co. (1970) pages 168-9.
- 2 Vaughan VC, McKay RJ, Behrman R. "Nelson's Text book of Paediatrics" 11th ed. Philadelphia, W.B. Saunders Co. (1979) pages 366-7.
- 3 Thornton HL. Emergency Anaesthesia, 2nd ed. (1974) p. 445.

Résumé

Une malade porteuse du syndrome de Turner a été anesthésiée à plusieurs occasions pour laparoscopie, vaginoplastie et changement de pansements. L'intubation a été rendue difficile pour le ptyrygion du cou et la brièveté trachéale. Les autres difficultés rencontrées comprennent la veinopuncture, l'excrétion médicamenteuse retardée, les anomalies cardiovasculaires et les difformités rachidiennes qui rendent impossible le blocage régional.