SEVERE LARYNGOSPASM IN A CASE OF ARTHROGRYPOSIS MULTIPLEX CONGENITA UNDERGOING GENERAL ANAESTHESIA

Khalid Mahmood, Naseem Ahmed, Sohail Muzammil

Combined Military Hospital Pano Aqil

INTRODUCTION

Arthrogryposis Multiplex Congenita (AMC) is characterized by multiple, congenital, non progressive joint contractures. There are reports of anomalies in other organ systems as well. Patients with AMC frequently require anesthetic care to correct orthopedic deformities. The multi-system involvement and the neuromuscular elements of the disorder lead to perioperative concerns with such patients including the potential for difficulties with airway management.

CASE REPORT

A baby girl, aged 14 months, suffering from Arthrogryposis Multiplex Congenita, reported for Ponseti plaster application for correction of telepes equino varus .The baby cried inconsolably and was uncontrollable, therefore general anaesthesia was planned. IV line was established and the baby was anaesthetized by giving inhalational agents (Oxygen, Nitrous oxide and Isoflurane).She developed mild laryngospasm during induction that was managed by increasing the depth of the anaesthesia and intermittent positive pressure ventilation. Intra operative course remained smooth and eventless. During recovery however she developed severe, life threatening laryngospasm. Her oxygen saturation dropped to 30 to 40% and she became cyanotic. Intermittent positive pressure ventilation (IPPV) was given with 100% O2 for about 10 minutes. Gradually her Oxygen saturation improved and she regained adequate spontaneous ventilation. She made a complete recovery from the effects of General anaesthetics.

On subsequent weekly changes of plaster, she was anaesthetised with intravenous ketamine, avoiding volatile inhalational agents. Her subsequent four anaesthetics with intravenous ketamine remained uneventful.

Correspondence: Maj Khalid Mahmood, Classifief Anaesthetist, CMH Pano Aqil
Email: khalid@1224@gmail.com
Received: 29 Jan 2009; Accepted: 10 Nov 2009

DISCUSSION

AMC was first described in 1841 by Otto, who referred to it as congenital myodystrophy. The current term was suggested by Stern in 1923 from two Greek words meaning "crooking of joints". AMC has a monogenic mode of inheritance and most likely due to homozygous autosomal recessive genes. It may also be seen in a number of states that immobilize the fetal limbs at some period during or shortly after the formation of joints1. A prominent feature of AMC is presence of joint contractures (mostly flexion type) and the muscles at the affected joints are weaker than those at the normal joints. There is scant subcutaneous tissue and muscle2. AMC may present as a myopathy or neuropathy, with associated hypotonia and wasted muscle mass. Fibrous and fatty degeneration of the muscle, as well as fatty infiltration between muscle bundles, can be seen. There can be involvement of the spine leading to hyperlordosis or scoliosis. Central nervous system can have degeneration and a reduction in the number and size of anterior horn cells. The diameter of the spinal cord is decreased, usually in the cervical and lumbar regions. About 10% of affected patients have associated congenital heart disease including patent ductus arteriosus, aortic stenosis, coarctation of the aorta and cyanotic congenital heart disease.

Problems arising during anaesthetic management include difficulty in tracheal intubation due to microganthia and a short neck, laryngospasm, bronchospasm, hypotension, diminution of muscle tone and prolonged action of various inhalational and injected anaesthetic drugs 3,4. Respiratory problems include alveolar hypoventilation, microatelectasis, a restrictive respiratory pattern, a decreased ability to cough and an increased incidence of aspiration.

Administration of inhalational anaesthetics may further aggravate the hypoxia due to counteraction of the hypoxic pulmonary vasoconstriction and the added central depression of respiration, especially in spontaneously breathing patients5.

Ketamine provides a better choice as a general anaesthetic as it is not associated with significant respiratory or cardiovascular depression. In these patients, a high incidence of malignant hyperpyrexia (MH) has been reported. Ketamine has been used safely in MH susceptible patients.

CONCLUSION

We conclude that AMC patients undergoing general anaesthesia pose a serious management challenge to the anaesthesiologist due to underlying complex multisystem problems. These patients may have hypersensitive, difficult airways, a high potential to react adversely to inhalational anaesthetics and muscle relaxants and a susceptibility to malignant hyperthermia. They require specialized perioperative care and management plans. Low dose ketamine is a safe anaesthetic in a patient with AMC.

REFERENCES

1. Jago RH. Arthrogryposis following treatment of maternal tetanus with muscle relaxants. Arch Dis Child 1970; 45:277-8.
2. Baines DB et al. Anaesth Intensive care, 1986, 14,370; Oberdi G. S.et al. Can. J. Anaesth.1988,35, 288-290 & 612-614
3. Jones AEP, Pelton DA. An index of syndromes and their anaesthetic implications. Can Anaesth Soc J 1976; 23:
4. Lee's Synopsis of anaesthesia, 1999, 12,355
5. Bjertnaes LJ. Hypoxia-induced pulmonary vasoconstriction in man: inhibition due to diethyl ether and halothane anaesthesia. Acta Anaesthesiol Scand 1978; 22: 570-88.