LETTERS

Renal transplantation in a patient with Bardet-Biedl syndrome, situs inversus totalis and bifid epiglottis: Anesthetic management

Dear Editor,

Bardet Biedl syndrome (BBS) is a multisystem autosomal recessive disorder, characterized by retinal dystrophy, central obesity, polydactyly, cognitive impairment, and gonadal and renal dysgenesis. Classification of the clinical manifestations of BBS is complex and ambiguous due to phenotypic variability of the disorder poses. The later poses great challenges to the management of such patients. Bifid epiglottis and disturbance of tooth formation have been reported, while situs inversus and Hirschsprung disease are considered as uncommon manifestations of the syndrome. Contrariwise, renal impairment is frequent and an important cause of death in these patients.[1]

A 13 year old female patient was referred to our Transplant unit for kidney transplantation from a healthy related living donor. The patient had retinitis pigmentosa, congenital horizontal nystagmus, hypogonadism, central obesity, situs inversus totalis, low growth and learning disabilities. She had also undergone two repair operations at the age of 3 and 6 for postaxial polydactyly of the upper and the lower extremities respectively, which were performed under general anesthesia without complications. One hour before surgery the patient was given ranitidine and ondasetrone i.v. and slow i.v. infusion of antibiotic prophylaxis with sublactam/ampicillin was started. Anesthesia was induced with lidocaine 60 mg i.v, propofol 2.5 $mg.kg^{-1}$ i.v., fentanyl 3.5 $\mu g.kg^{-1}$ i.v. and succinylcholine 2.5 $mg.kg^{-1}$ i.v. and was maintained with continuous delivery of sevoflurane, continuous infusion of remifentanyl and intermittent doses of atracurium as needed. Laryngoscopy revealed a Cormack-Lehane III class airway with a bifid epiglottis. Nevertheless, tracheal intubation proved unexpectedly easy as the tracheal tube passed through the two "leaflets" of the epiglottis. Heart rate and E.C.G. were monitored continuously using a 5-lead system with the patches placed reversely to the standard E.C.G. Arterial pressure , cardiac output and stroke volume variation were monitored directly through left radial artery cannulation and a central line was inserted through RJV. Transcranial cerebral oximetry was also obtained with near infrared spectroscopy. The patient was haemodynamically stable during the operation that lasted 260 minutes. After the operation the young girl was transported to the transplant unit where steroid-free immunosuppression with basiliximab was initiated. Her overall postoperative course was uneventful and she was discharged from the hospital four days later.

Differential diagnosis of BBS includes Alstöm syndrome, Meckel-Gruber syndrome, McKusik-Kaufmann syndrome and other ciliopathies. Renal impairment is the leading cause of death in BBS and includes both morphological and functional disorders. Bifid epiglottis is a rare anomaly, often associated with other anomalies, particularly polydactyly and Pallister-Hall syndrome [2]. There are only two reports in the literature of bifid epiglottis in patients with BBS [2]. Situs inverus totalis is present in 50% of individuals with Kartagener syndrome, but it's a very rare feature of BBS[3]. Dilated cardiomyopathy and severe osteodystrophy have also been reported[3].

Considering the involvement of multiple major organ systems, anesthetic management of patients affected by the syndrome poses a considerable challenge to the anesthesiologist. This case has given us the insight of this rare condition with some new findings.

References:

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