
Transthoracic oesophagectomy in a patient with Fanconi's anaemia

Sir,

Fanconi's anaemia (FA) is a rare autosomal recessive disease characterised by chromosomal fragility, congenital abnormalities, bone marrow failure and a predisposition to solid tumours.^[1]

A 14-year-old boy [Figure 1] was diagnosed with FA after stress cytogenetic studies showed increased sensitivity to mitomycin C. He presented with dysphagia and was diagnosed as carcinoma of the oesophagus and transthoracic oesophagectomy was planned. On assessment, his height was 142 cm (short for age) and weight 38 kg. He had café au lait spots, sensori-neural deafness and small genitals.

Screening for vertebral defects, anal atresia, cardiac abnormalities, tracheoesophageal fistulas, renal defects and limb anomalies defects^[2] showed no developmental malformations. His serum cortisol and growth hormone levels were normal. His blood picture was megaloblastic. He was on oral stanozolol and folic acid. He was initiated on high-protein diet and incentive spirometry.

Due to fever, thrombocytopenia and neutropenia in the immediate pre-operative period, surgery was deferred and he was optimised with one dose of granulocyte cell stimulation factor (G-CSF) 150 µg and piperacillin-tazobactam 2.25 g intravenously every 6th hourly.

After pre-medication with 60 µg intravenous fentanyl, anaesthesia was induced with intravenous 30 mg lidocaine, 60 mg propofol and 30 mg rocuronium. A 32-Fr left-sided double lumen endobronchial tube was inserted, anaesthesia was maintained on 30%

oxygen/air mixture with desflurane titrated to one minimum alveolar concentration with intravenous fentanyl (30 µg/h) infusion. Right internal jugular vein and radial arterial access were obtained.

The procedure lasted 345 min with estimated blood loss of 400 ml. Two units packed cells and one unit single donor platelets were transfused. Patient received intercostal blocks and bilateral subcostal block. Neuromuscular block was reversed and trachea was extubated when the child was fully awake. He received paracetamol 500 mg 6 hourly and fentanyl 15–20 mg/h to maintain VAS score of <2.

The immediate post-operative period was uneventful; on post-operative day (POD) 4, his oxygen requirement increased and lung ultrasonography showed bilateral basal atelectasis. This was managed with bronchodilators, incentive spirometry and bronchoscopic extraction of mucus plugs. On POD 6, the patient became thrombocytopenic with a platelet count of 35,000/mm³ and needed three units of single donor platelets. At the time of discharge on POD 14, the patient was orally able, afebrile with normal haematology.

Disease specific concerns include pancytopenia and a predisposition to sepsis. We avoided drugs such as aspirin, antihistamines, non-steroidal anti-inflammatory drugs and nitrous oxide due to their potential myelosuppressive effect. Central neuraxial anaesthesia was avoided in view of thrombocytopenia. Hepatorenal derangement can be a side effect of androgen therapy, and agents such as halothane, long-acting muscle relaxants, non-steroidal anti-inflammatory agents, morphine and



Figure 1: Patient with Fanconi's anaemia

pthidine were avoided.^[3] As endocrinopathies such as growth hormone deficiency, cortisol insufficiency, hypothyroidism and insulin resistance are common, pre-operative endocrine screening was done. Renal and urinary tract abnormalities, ear malformations and hearing loss are often present. Our patient had hearing loss hence ototoxic and nephrotoxic drugs were avoided.

Patients with FA are sensitive to ionising radiation and oxygen-derived free-radical-induced DNA damage, which increases the risk of developing malignancies.^[4] We used air-oxygen mixtures during anaesthesia to decrease the FiO₂ and avoided radiographic exposure by the use of point of care lung ultrasonography. Other concerns include the side effect of G-CSF^[5] treatment such as serous cavity effusions, capillary leak syndromes, interstitial pulmonary oedema and hypoxemia.

Anaesthetic concerns of FA are myelosuppression, potential for blood loss and pulmonary complications. Knowledge of associated congenital defects makes the anaesthesiologist vigilant and tailor anaesthetic management to avoid potential organ damage.

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.

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We wish to acknowledge that the father of our patient was very willing to let us take pictures and gave us complete consent to present the details of his son course in the hospital.

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Conflicts of interest

There are no conflicts of interest.

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