Erdheim — Chester disease: Clinical pearls for the anesthesiologist

Sir,

Erdheim–Chester disease (ECD)[1] is an extremely rare non-Langerhan's form of histiocytosis, characterized by xanthomatous tissue infiltration with foamy CD68/CD1a histiocytes. Its prompt preoperative recognition and evaluation of all the involved systems is the key to successful perioperative outcome. Approximately half of these patients have extra-skeletal manifestations, including exophthalmos,[2] xanthelasma, interstitial lung disease,[3] retroperitoneal "fibrosis" with perirenal or ureteral obstruction, renal failure, diabetes insipidus, as well as central nervous system[4] and cardiovascular involvement.[5] The diagnosis of ECD can be difficult because of the rarity of the disease and the need to differentiate it from Langerhan cell histiocytosis[6] and Rosai–Dorfman disease.[7]

A 66-year-old male patient presented with right ankle fracture for open reduction and internal fixation. He was a diagnosed and treated case of ECD, with severe bilateral exophthalmos (right > left).

On preoperative evaluation, all routine blood investigations, including thyroid function test and coagulation profile were within normal limits. Electrocardiogram (ECG), chest X-ray, pulmonary function tests (PFT), and ultrasound abdomen were within normal limits. Whole body bone scan revealed increased radiotracer uptake in several long bones.

Biopsy of the eye mass done previously showed numerous foamy histiocytes and plasma cells. Immunohistochemical stains were strongly positive for CD 68. Both eyes show axial proptosis, with bilateral multinodular mass (noncompressible, painless, nonpulsatile) in inferotemporal part of the orbit, restricted ocular movements, bilateral cataract, and bilateral disc edema on fundus examination.

In cardiac evaluation, baseline echocardiography was normal (ejection fraction 55%). Stress test (dobutamine stress echocardiography) was negative for reversible myocardial infarction.

Intraoperatively, combined spinal–epidural anesthesia was administered under all asepsis. Epidural infusion was started with 0.125% bupivacaine and continued for postoperative analgesia. Special precautions were taken to protect the eyes, bony prominences, and pressure points. All vital parameters, including ECG and urine output were normal. The patient was shifted to surgical intensive care unit for observation and later shifted to ward. He was finally discharged after 2 days with advice regarding regular follow up with the orthopedic surgeon, ophthalmologist, and cardiologist.

In brief, the perioperative: precautions to be taken in a patient of ECD include preoperative PFT evaluation, postoperative incentive spirometry and physiotherapy, identification of the extent of renal function derangement and choice of anesthetic agents accordingly; preparedness to perform dialysis in the perioperative period if needed, close monitoring for conduction blocks and pericardial effusion or tamponade, strict monitoring of intake–output and early identification of diabetes insipidus, gentle handling of patient; padding of pressure points and bony prominences; evaluation of the spine for any deformity, especially before attempting regional blocks, preoperative ophthalmologic assessment to document degree of vision loss and optic nerve compression; and avoidance of raised intraocular pressure and eye injuries.

To conclude, the multisystem presentation of ECD can pose a challenge for the anesthesiologist, especially if not aware of its problems.

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Quick Response Code:	Website: www.joacp.org
	DOI: 10.4103/0970-9185.130127