

Multisystemic Erdheim-Chester disease: case report outlining anaesthetic considerations and management

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Background:

With only around 500 cases reported in scientific literature, Erdheim-Chester disease (ECD) is an extremely rare form of CD68 positive non-Langerhans cell histiocytosis, characterized by xanthomatous infiltration of tissues in various organs. From the anaesthetic point of view, these patients should be addressed globally, given the multisystemic involvement of their pathology.¹ The reported case describes a patient diagnosed with ECD (pulmonary, endocrine and vascular disease) referred for thyroidectomy after being diagnosed with a carcinoma. To our knowledge, the only existing case report describing the anaesthetic management of a patient with ECD was published in 2014.^{1,3}

Case report

- **69-year-old** patient referred for a **thyroidectomy** after a positive biopsy for papillary thyroid carcinoma
- **Previous diagnosis of ECD**
 - pathological evidence in an epiploon biopsy
 - central *diabetes insipidus*
 - diffuse vascular thickening in several territories such as cerebral, thoracic and abdominal
 - retroperitoneal fibrosis
 - pleural and pericardial thickening

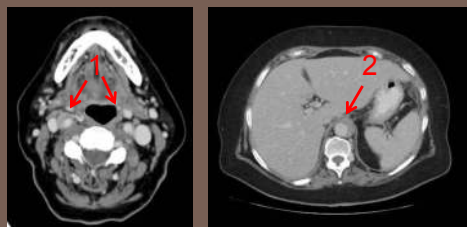


Figure 1. TC scan showing primitive carotids with incipient diffuse wall thickening (1) and periaortic fibrosis involving the whole aorta leading to a "coated aorta" appearance (2).

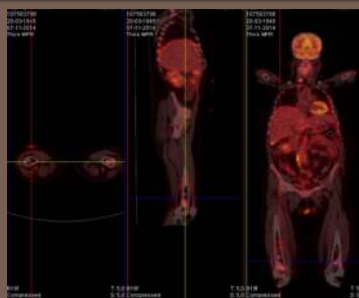


Figure 2. PET-scan showing inflammatory diffuse disease

ECD	
Histopathologic features	
CD68	+
CD163	+
CD1a	-
CD207	-
S100	- or weakly +
Factor XIIIa	+
Touton giant cells	+
Other characteristic features of lesional histiocytes	Xanthomatous features Fibrosis

Table 1. Histopathologic positive findings leading to ECD diagnosis.²

Anaesthetic management

1. Evaluation and optimization of the patient in the pre-anaesthetic period;
2. Basic monitoring and invasive blood pressure during surgery;
3. The chosen anaesthetic technique was a balanced general anaesthesia. Induction was made with propofol (2 mg/kg), fentanyl (20 mcg/kg) was used as an analgesic and rocuronium (0.6 mg/kg) as a muscle relaxant. Anaesthesia was maintained with desflurane. Reversal of neuromuscular blockade was achieved with suggamadex (2 mg/kg);
4. Monitoring and post-operative stay in intensive care unit.

Discussion

Given the pulmonary findings, endocrine and cerebrovascular disease and cardiovascular alterations, pre-anaesthetic evaluation and optimization was essential. Pleural thickening leads us to assume the presence of some degree of pulmonary fibrosis, even when is not evident in pulmonary function tests. Pericardial thickening presented by the patient could be associated with cardiac complications as constrictive pericarditis, pericardial effusion and cardiac tamponade, despite pre-operative echocardiogram did not showed such evidence. In the context of diabetes insipidus she maintained her usual therapy with desmopressin; that was vital to assure an adequate control of ADH's secretion and prevent complication such as hyponatremia and hypovolemia. It was also crucial to avoid periods of hypotension and hyperventilation in intra-operative period since hypocapnia is associated with reduction of cerebral blood flow and therefore increased risk of ischemic event. The main goals of the anaesthetic management were to achieve the best possible hemodynamic stability and to prevent any critical events.

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2. Eli L. Diamond, Lorenzo Dagna, David M. Hyman, et al. Consensus guidelines for the diagnosis and clinical management of Erdheim-Chester disease. Blood, 24 July 2014 ; volume 124, number 4

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