
Castleman's disease: Implications for an anesthetist

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Abstract

Purpose: Castleman's disease is a rare disease of the lymphoreticular tissues characterized by lymph node swellings in the neck, groin, mediastinum etc. In its multicentric form, it is often associated with systemic diseases of immune nature including myasthenia gravis, nephrotic syndrome etc. An anesthetist should exert caution while encountering a patient with such a presentation. Though the exact incidence of the disease is not known, many patients can remain asymptomatic.

Clinical report: We report a patient with tonsillar growth and mediastinal lymphadenopathy taken for direct laryngoscopic examination and biopsy under general anaesthesia who was subsequently diagnosed as Castleman's disease.

Conclusion: This report should heighten the anesthetists' concern for adequate pre-operative assessment and careful planning for a patient of suspected Castleman's disease undergoing surgery or any other procedure under general anesthesia.

INTRODUCTION

Castleman's disease (also called giant lymph-node hyperplasia or angiofollicular lymph-node hyperplasia) is a highly heterogeneous clinico-pathological entity belonging to the lympho-proliferative disorders. Originally described in 1956 by Castleman and co-workers¹ as a large, benign, unique asymptomatic mass of mediastinal lymph nodes, it was shown in subsequent reports to include extra-mediastinal and multicentric forms.²

Although its etiology is not very clear, it has been suggested that abnormal production of a B cell growth factor, such as IL-6, in the hyperplastic lymph nodes causes lympho-proliferation and plasma cell differentiation.³

As Castleman's disease is considered to be a heterogeneous entity related to conditions of immune deregulation it is believed that various disorders of the immune system may be characterized by Castleman-like histological changes, such as infections (HIV) and primary autoimmune diseases (systemic lupus erythematosus, POEMS syndrome, etc.)²

Histologically CD is classified into two major types according to pathological findings: 1) the hyaline-vascular type with small hyaline-vascular follicles and interfollicular capillary proliferation; and 2) the plasma-cell type characterized by a massive accumulation of polyclonal

plasma-cells in the interfollicular region. There is also a third, mixed variant, with features of both hyaline vascular and plasma-cell type.^{4, 5}

Clinically, CD can be classified into either localized or systemic forms. The usual presentation for localized disease is a single enlarged lymph node or widened mediastinum. The systemic form of the disease, also referred to as multicentric CD (MCD), is characterized by diffuse lymphadenopathy, hepatosplenomegaly and constitutional symptoms. Localized CD of the hyaline vascular type is usually asymptomatic, whereas patients with the plasma cell variant often have systemic manifestations. Patients with MCD have significant lymphadenopathy and an aggressive, sometimes fatal, clinical course.⁴

Since the multicentric Castleman's disease can affect the lungs, kidneys, muscle, pericardium, central nervous system and vulva, a thorough pre-anesthetic check up is necessary to assess the associated pathologies.

An extensive literature search showed only one article on anesthetic considerations for Castleman's disease.⁶ Due to the rarity this disease and the possibility of associated conditions which may have anesthetic implications, we report this case.

CASE

A forty-five year old male, weighing 55 kg and height 158 cm presented with hoarseness of voice of 2 months duration. Indirect laryngoscopy revealed tonsillar swelling, and CT scan of the chest revealed solitary mediastinal lymphadenopathy. The patient was planned for direct laryngoscopy and biopsy under general anaesthesia as day care procedure.

During pre-anesthetic assessment the, physical assessment revealed mild glossitis and cheilitis. The mouth opening was normal and there was a non-tender palpable lymph node in the submental region. On auscultation, there was mild bilateral wheeze but the patient had no difficulty in breathing. The routine chest X ray and ECG were normal.

But since the CT chest had revealed a solitary enlarged lymph node, the possibility of Castleman's disease besides lymphomas etc. was kept in mind.

The blood investigations, CT head and MRI spine were normal.

Since the patient was a day case, he did not receive any premedication.

In the operating room, standard monitoring was established with continuous ECG, NIBP and SpO₂. Anesthesia induction was achieved with IV Propofol 120 mg and gas mixture of N₂O /O₂ (2:1) with sevoflurane. Relaxation was achieved with single dose of succinyl choline 50 mg and direct laryngoscopy was performed. There was growth in the right tonsillar region which was seen extending backward behind the palate. After obtaining the biopsy sample, patient was manually ventilated till the return of spontaneous respiration. The entire procedure lasted for less than ten minutes and the recovery was uneventful. The diagnosis of Castleman's disease was confirmed after obtaining the histopathological report of the biopsy sample.

DISCUSSION

The preoperative assessment of any patient with mediastinal lymphadenopathy and growth in the oral cavity should arouse suspicion for Castleman's disease. Although the majority of the cases are likely to be asymptomatic with localized mediastinal lymphadenopathy, a minority of cases may be associated with a variety of conditions like myasthenia gravis, nephrotic syndrome, thrombocytopenic purpura, POEMS syndrome (Takatsuki's syndrome), HIV, Kaposi's sarcoma, Rheumatoid arthritis, Sjogren's syndrome

etc.^{7,8,9,10,11,12}

One of the potentially fatal complications which can arise in an undiagnosed Castleman's disease is an anterior mediastinal mass. There can be respiratory tract obstruction or great vessels obstruction leading to circulatory collapse during induction of anesthesia. Careful planning about the anesthesia techniques with discussion with the surgical counterpart may lessen the risk of cardiorespiratory complications.

In our CT chest, we had seen an enlarged lymph node, but after prior discussion with surgeon and radiologist we had the option of emergency tracheostomy planned beforehand and consent for the same was also obtained.

Since neck is the most common site for extrathoracic presentation¹³, any combination of oral and perioral infiltration with steroid induced facial obesity can produce significant difficulty with endotracheal intubation in these patients. Appropriate measures including staff and equipment should be arranged beforehand.

Since sensorimotor neuropathy is the most frequent neurological manifestation in Castleman's disease¹⁴, preoperative assessment should include neurological examination and CT and/or MRI of the CNS as may seem necessary. Neuropathies should be considered as relative contraindications for spinal anesthesia whereas CNS infiltrations or masses as absolute contraindications for the same. When associated with myasthenia gravis¹¹, careful monitoring of muscle relaxation with titration of relaxant drugs becomes mandatory.

To conclude, any patient with diagnosed or suspected Castleman's disease requires adequate preoperative assessment to exclude wide ranging medical problems and their possible influence on anesthetic management.

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