Case Report

Castleman’s disease: an entity forgotten in ENT

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ABSTRACT

Castleman’s disease (CD) usually presents as localized or systemic lymphadenopathy or as an extra nodal mass. The usual site of presentation are mediastinum, retroperitoneum, axilla and mesentery. Only 3 cases of CD have been reported in retro pharyngeal space. We report a case of 20 year old male patient with retropharyngeal mass. He presented with difficulty in swallowing, change in voice and respiratory distress. The mass was removed in-toto transorally after performing elective tracheostomy. The histopathological findings were consistent with hyaline vascular type of CD. He was decannulated after two day and postoperative period was uneventful. Postoperative CT imaging confirmed the complete excision of tumor and patient is on follow up, with no signs of recurrence. The presentation of tumour in the retropharyngeal space which is a rare site of occurrence add to the uniqueness of this case. Unicentric CD has an excellent prognosis and surgery is the management of choice. Its clinical features, histological subtypes, treatment modalities and prognosis are discussed.

Keywords: Retropharyngeal mass, Trans oral approach, Castleman’s disease

INTRODUCTION

CD is an uncommon clinicopathological entity which usually presents with lymphadenopathy or an extra nodal mass. It is a rare form of massive lymph node hyperplasia and is histologically characterized by angiofollicular lymph node hyperplasia. In 1956, Castleman et al first described this entity in a group of patients with solitary masses in the mediastinum.1

This study reported one of the rare sites of presentation of CD and its successful surgical management.

CASE REPORT

A 20 year old male presented to ENT outpatient department with complaints of difficulty in swallowing for 6 months. He also complained of difficulty in breathing which worsened on exertion for past 1 month. The patient didn’t give any history of weight loss, hemoptysis, hematemesis, malena, dyspepsia or any addiction.

Endoscopic evaluation of oropharynx and hypopharynx revealed a large sub mucosal globular swelling arising from posterior pharyngeal wall and bulging over laryngeal inlet, obscuring view of endo larynx. Computed tomography showed a well-defined homogenous non-enhancing mass lesion of retropharyngeal space extending from second to fifth cervical vertebra level, projecting over supraglottic larynx with airway attenuation (Figure 1).
The MRI reported a large, lobulated, homogenously enhancing mass lesion in the retropharyngeal space extending from C2-C5 vertebral levels, measuring 4.1×6.5×5.5 cm in retropharyngeal space, projecting into hypo pharynx and supraglottic larynx with airway attenuation. The lesion appeared isointense on T1W1 and heterogeneously hyper intense on T2W1 with near homogenous contrast enhancement.

After doing an elective tracheostomy, patient was taken up for direct laryngoscopic evaluation, which revealed a huge sub mucosal smooth mass arising from posterior pharyngeal wall and covering its entire breadth, extending superiorly from the level of base of tongue and inferiorly reaching just above cricopharynx. Biopsy was taken but was inconclusive. Patient was then taken up for tumour excision biopsy via intra oral approach. After positioning a Boyle Davis mouth gag, the tumor was exposed adequately. A midline vertical mucosal incision was given over posterior pharyngeal wall up to the level of constrictor muscle. Tumor was dissected by bipolar cautery, blunt dissection and sharp dissection by tenotomy scissors to free it from surrounding tissues (Figure 2). Rigid endoscopy were used to dissect in area of hypopharynx. Laterally plane was created between the mass and bilateral carotid artery by careful dissection. The mass was freed and delivered out in toto with minimal blood loss. He was put on Ryle’s tube feeding for a week.

Postoperative period was uneventful and he was decannulated after two days. Histopathology showed vascular channels of variable size with inter-communication along with skeletal muscle, nerve bundle and lymphoid follicles which was consistent with hyaline vascular type of CD (Figure 3). Postoperative CT imaging confirmed the complete excision of tumor and patient is on follow up, with no signs of recurrence.
DISCUSSION

Giant lymph node hyperplasia, angiofollicular hyperplasia, angiomatous lymphoid hamartoma and follicular lymphoreticuloma are the synonyms of same entity.

Clinically the disease can be of localized or multicentric type, out of which the former is more common. Based on histological pattern, the main types are hyaline vascular type and plasma cell variant. A third type showing a mixed picture can also be seen in multicentric type.

Localized CD is usually asymptomatic and is often discovered incidentally. Although most commonly involved site is the mediastinum (60%), other sites include retroperitoneum, neck, axilla, mesentery, nasopharynx, leptomeninges and parotid. Diagnosis requires histological analysis of lymph node biopsy.

Multicentric CD on the other hand is usually symptomatic and present as asthenia, weight loss, fever or polyadenopathy with a mean of four site involvement and is often associated with hepatosplenomegaly.

In a study conducted by Yoshizaki et al it was observed that there was an abrupt drop in interleukin 6 levels and resolution of symptom following removal of CD mass. A similar study by Nishimoto et al on multicentric CD showed that treatment with IL-6 receptor antibody relieved symptoms and signs of the disease. This indicates a possible role of IL-6 in the pathogenesis and symptomatology of CD.

Although the etiology of this disease is not well understood, HHV-8 which infects B lymphocytes and macrophages appears to be causally associated with multicentric CD. Quantitative analysis of HHV 8 DNA suggests a predominance of frequent lytic replication in multicentric CD. Also its association with Kaposi sarcoma which develops in the clinical course of most HIV positive cases has been reported. Another associated condition seen with multicentric CD is coexisting POEMS syndrome characterized by peripheral neuropathy, organomegaly, endocrinopathy and monoclonal plasma proliferative disorder and skin changes. Various other risk factors identified include multicentric disease, plasma cell variant, organomegaly, low albumin and peripheral neuropathy.

In a study by Dispensieri et al in a group of patients of multicentric CD, it was observed that those patients who had POEMS syndrome and osteosclerotic lesions had the best outcomes in comparison to patients who had POEMS syndrome without osteosclerotic lesions. A similar study by Shin et al suggested the presence of splenomegaly and age of the patient to be the most important prognostic factors for multicentric CD.

Mallik et al reported that large atypical cells with crumpled tissue paper like chromatin with nuclear indentations and nuclear grooves is the most consistent clue to cytological diagnosis of CD, although it’s very difficult.

Hyaline vascular variant of CD is the most common subtype of CD. Microscopically, the characteristic features of hyaline cell variant include increased number of scattered large follicles at different levels of maturity and marked vascular proliferation of their germinal centers with or without hyalinization of blood vessels. Based on relative morphologic changes in follicular and interfollicular region, two sub categories are described. Follicular hyaline vascular variant show predominantly follicular abnormalities which compose more than 50% of the lymph node. Whereas stroma rich subtype show predominantly interfollicular changes like presence of numerous sclerotic blood vessels. Interfollicular region with proliferating blood vessels constitute more than 50% of lymph node. Arrangement of lymphocyte in the mantle zone gives an onion skin or stadium seating appearance.

The plasma cell variant on the other hand has plasma cells, eosinophils, immunoblasts and monocytes in the interfollicular area surrounding larger germinal centers and has significantly lesser vascularity compared to hyaline variant.

CD is often misdiagnosed due to its histopathological similarity with follicular lymphoma, mantle zone lymphoma, AIDS related complex and thymoma.

Unicentric CD has an excellent prognosis and surgery is the management of choice. Excision of the lesion need not be followed by any further treatment. In our case, we opted for a trans oral pharyngotomy approach for the retropharyngeal mass with consent to change to transcervical approach if required. This approach provides direct, but limited field of access; therefore may be inappropriate for larger, ill-defined vascular tumors where there might be life-threatening hemorrhage. This necessitates a detailed preoperative radiological evaluation for appropriate selection of patients.

Trans Oral Robotic Surgery (TORS) provides an attractive alternative as it is minimally invasive and obviates the need for external conventional pharyngotomy or mandibulotomy thereby maintaining the anatomical framework of laryngopharynx which is necessary for deglutition. The added advantages include shorter hospital stay and lesser morbidities.

Management of multicentric CD includes high-dose chemotherapy with peripheral blood stem cell transplantation, rituximab, and anti-interleukin-6. The introduction of rituximab in patients with HIV positive, HHV 8 positive multicentric CD has improved the general outcome and reduced the risk of transformation
into lymphoma. The recent introduction of monoclonal antibody targeting IL-6 treatment including tocilizumab or siltuximab needs to be followed up for the results.

**CONCLUSION**

Although a rare entity, CD should be considered as a differential diagnosis in cases of systemic and localised lymphadenopathy. The presentation of tumour in the retropharyngeal space which is a rare site of occurrence add to the uniqueness of this case. Unicentric CD has an excellent prognosis and surgery is the management of choice.

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**REFERENCES**
