Case Report

Castleman’s disease presenting as parotid mass: a case report

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ABSTRACT

Castleman’s disease is a rare, benign, lymphoproliferative disorder of unknown etiology. It can involve any lymph node group in the body with mediastinum being the commonest site. Salivary glands are affected rarely. We report a 35 year old male patient who presented with slowly progressive, painless right sided parotid swelling for 3 years. Clinical examination showed a 4×3 cm single, firm, non-tender, non-pulsatile swelling with smooth surface and normal overlying skin present in the right parotid region. Magnetic resonance imaging (MRI) revealed a well-defined hyper-intense mass lesion on T2 measuring 3.9×3×2.8 cm and Fine needle aspiration cytology (FNAC) showed intense lymphoplasmacytic infiltrates. No conclusive diagnosis could be made on the basis of FNAC. So, the excision of parotid mass with facial nerve preservation was done. The final histopathology confirmed the diagnosis as Castleman’s disease. Although Castleman’s disease in the parotid gland is rare, clinicians should consider it as the differential diagnosis of any solid tumors that exhibit non-specific presenting characteristics and surgical excision is preferred treatment for unicentric disease.

Keywords: Castleman’s disease, Parotid, Head and neck, Salivary gland

INTRODUCTION

Castleman’s disease is a rare, benign, lymphoproliferative disorder, first described in 1956 by Dr. Benjamin Castleman.¹ It is known by different names including lymphoid hamartoma, giant lymph node hyperplasia, angiofollicular hamartoma and benign giant lymphoma.² The etiopathology of Castleman’s disease (CD) is still unknown and current hypothesis suggest antigenic stimulation, chronic low grade inflammation, hamartomatous process, viral infection as Epstein-Barr, herpes virus and immunodeficiency state.³ The disease may present in either unicentric or multicentric forms. The unicentric form usually present as an asymptomatic mass whereas the multicentric form presents with generalized lymphadenopathy, malaise, hepatosplenomegaly and follows an aggressive clinical course.

The parotid gland involvement of CD is rare and from our literature review, fewer than 32 cases of Castleman’s disease involving the parotid gland have been reported up to date. In this report, we present a rare case of unicentric CD presenting as a mass in right parotid gland.

CASE REPORT

A 35 year old male patient presented to Otorhinolaryngology outpatient department with complaints of right sided parotid swelling for past 3 years. The swelling was insidious in onset and slowly progressive in past 1 year with no history of trauma and without any...
suspiciously related lesions elsewhere. The swelling was not associated with any postmeal increase in size and it was painless. On physical examination; there was a 4 × 3 cm single, firm, non-tender, non-pulsatile swelling with smooth surface and normal overlying skin present in the right parotid region. There were no signs of inflammation, no palpable lymphadenopathy and no evidence of facial nerve involvement. Rest of the head & neck and systemic examination was unremarkable.

Magnetic resonance imaging (MRI) of the parotid and neck was done which revealed a well-defined hyper-intense mass lesion on T2 measuring 3.9×3×2.8 cm occupying the superficial lobe of parotid gland with enlarged lymph nodes (Figure 1). Fine needle aspiration cytology (FNAC) was performed which showed intense lymphoplasmacytic infiltrates, epithelial cells, giant cells and occasional acinar cells.

The patient underwent right sided excision of parotid mass with facial nerve preservation. The histopathology revealed Castleman’s disease of hyaline vascular subtype (Figure 2). The patient recovered well following surgery with normal facial nerve function and is under follow-up for one year with no evidence of recurrence.

**DISCUSSION**

Castleman’s disease was first described by Castleman et al in 1956 as a benign solitary lesion under the title of “localized mediastinal lymph node hyperplasia resembling thymoma”. It can occur at any age but most of the cases are young adults between 15 and 35 years of age; with no sex predilection.

Although pathogenesis of this disease is still unknown, several theories have been suggested. The most supported theory is excessive lymphoproliferation due to chronic stimulation by a virus or chronic inflammation. A strong association between Castleman’s disease and viral infections: EBV, HIV and HHV-8 have been reported in literature. Another strong theory proposes the significance of the interaction between interleukin 6 and tumor necrosis factor alpha and systemic presentation of multicentric CD.

The disease usually presents as a solitary mass, commonly found in mediastinum (60-86%), head and neck involvement is rare (6-14%). Within the head and neck region, CD most commonly presents as a solitary mass under the sternocleidomastoid muscle or arise as an extension of mediastinal mass. In the parotid, it probably arises in the para-glandular lymph nodes related to the parotid capsule or intra-glandular lymph nodes within the gland parenchyma. In our case the lesion was located superficially mimicking a parotid gland tumor.

CD is classified clinically into unicentric or localized and multicentric or generalized types. The unicentric form, as the name suggests, has a more benign process. It usually presents as a symptomatic palpable enlarged lymph node /mass. On the other hand, multicentric type of CD is more aggressive and presents with systemic symptoms, including fever, loss of weight and splenomegaly and can be associated with syndromes such as nephrotic syndrome and POEMS (polynuropathy, organomegaly, endocrinopathy, monoclonal protein, skin changes) syndrome.

Histologically, CD has 3 distinct subtypes: hyaline vascular, plasma cell and mixed type. The hyaline vascular CD is the most common type, accounting for 80-90% of the cases. This subtype has a benign character and is characterized by prominent proliferation of small blood vessels within the interfollicular areas that often have thick, hyalinized vessels walls and numerous small atrophic follicle centres. Peripheral concentric layering of lymphocytes results in expansion of the mantle zone with an “onion-skin” appearance. The plasma cell subtype is marked by diffuse plasma cell proliferation between follicles. Twenty two percent of the localized and majority of multicentric Castleman’s disease is plasma cell type. The mixed type is rare and is histologically a mixture of the two types.

**Figure 1:** T2 weighted MRI showing well defined hyper-intense mass lesion in right parotid gland (a) axial; (b) coronal.

**Figure 2:** Microscopic images showing (a) diffuse proliferation of plasma cells in the interfollicular region; (b) follicle with abnormal germinal centres with prominent hyalinization around vessels. The follicle is surrounded by concentric layering of lymphocytes giving “onion skin” appearance.
CD of the head and neck, especially parotid is often a diagnostic challenge. The diagnostic problem is due to paucity of signs and symptoms and the ability of CD to mimic other neoplasms. Imaging can be useful for differential diagnosis. Ultrasonography is efficient to evaluate the vascular nature of the disease. Computed tomography scan with contrast shows a densely enhancing, well circumscribed, homogenous mass which reflect hypervascularity of these lesions. On magnetic resonance imaging, Castleman’s disease shows a linear hypointense signal in a stellate or arborizing pattern, especially on T2-weighted sequences. However, the definitive diagnosis is only possible with histopathologic examination.

Total excision is the standard treatment for unicentric CD in head and neck region with 100% control rate for the hyaline vascular type and no recurrences in the unicentric variants. However, the non-operable cases are managed with radiotherapy although excision has a more preferable prognosis. On the contrary and due the aggressiveness of the multicentric form, it is controlled by palliative treatment only. Some patients require corticosteroid therapy with occasional chemotherapy (cyclophosphamide) in non-responders to steroids.

**CONCLUSION**

Although Castleman’s disease in the parotid gland is rare, clinicians should consider it as the differential diagnosis of any solid tumors that exhibit non-specific presenting characteristics. The hyaline vascular type is the most common histologic type and surgical excision is the preferred treatment for unicentric disease. On the other hand, multicentric form requires more aggressive treatment and long term follow-up.

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**REFERENCES**
