Castleman disease of the parotid gland: A case report and a review of the literature

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Abstract
Castleman disease is a lymphoproliferative disorder with unknown cause and a characteristic hypervascular lymphoid hyperplasia. Commonly the disease is located in the mediastinum. Head and neck is the second most common localization for the disease. However involvement of parotid gland is extremely rare. We present a 29 year old female patient with parotid gland Castleman’s disease. Although it is rarely seen in the parotid gland, it should be considered in the differential diagnosis of cases with mass in the parotid gland.

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1. Introduction

Castleman’s disease (CD) is a lymph node hyperplasia which was first described by Benjamin Castleman in 1956.1 It was known by different names including lymphoid hamartoma, giant lymph node hyperplasia, angiofollicular lymph node hyperplasia, angiofollicular lymph node hamartoma and benign giant lymphoma.1 Commonly the disease is located in the mediastinum.2 However, it may also be seen in other areas of the body, where lymph nodes are normally found.3 Although the head and neck area accounts for 14% of cases, nearly 90% of these are located in the neck and the involvement of parotid gland is extremely rare.3,4 This article presents a patient who was found to have CD in the parotid region. Additionally, review of the literature on this entity is discussed.

2. Case report

A 29-year-old female presented with a painless mass with 2-year history, in the right neck region. She had no additional symptoms, including fever, fatigue, weight loss and night sweating. The mass was stationary in course. She had no other comorbid disease. Rest of the history was unremarkable. Physical examination revealed a solid, well marginating mass in the right parotid region. Facial weakness was not observed. General examination revealed no other swellings elsewhere. Laboratory tests including blood glucose, kidney function tests,
liver function tests, cell blood count were in normal limits. Chest X-ray was normal. Magnetic resonance imaging showed a 30 × 18 mm in size, well marginated mass in the parotid region (Fig. 1). Fine needle aspiration biopsy revealed polymorphic lymphoid population. No definitive diagnosis could be achieved. Patient underwent a superficial parotidectomy operation. Histopathological examination of the specimen was reported as 'Castleman disease, hyaline vascular type' (Fig. 2). After 3 months follow-up the patient remains free of disease.

3. Discussion

Castleman disease is a lymphoproliferative disorder with an unknown cause and a characteristic hypervascular lymphoid hyperplasia, that was first described by Benjamin Castleman et al. in 1956. Although this disease has been reported at all ages, it is very rare in children. The age at presentation ranges from 2 months to 71 years. It is more commonly found in young adults. It has no sex predilection. Our case was a 26-year-old woman.

Castleman disease has been referred to by many names, including giant lymph node hyperplasia, angiofollicular lymph node hyperplasia, follicular lymphoreticuloma, and lymphoid hamartoma. It has been commonly found in mediastinum (60%), head and neck (14%), retroperitoneum (11%) or axilla (4%). Nearly 90% of the head and neck cases, are located in the neck and the involvement of parotid gland is extremely rare with only 25 cases reported in the English language literature. Other localizations in the head and neck region are, floor of the mouth, submandibular gland, larynx, palatal region and parapharyngeal space. In the head and neck region, Castleman disease most commonly presents as a solitary mass under the sternocleidomastoid muscle or arises as an extension of a mediastinal mass. In our case, Castleman disease was located within the parotid region. The majority of the cases of Castleman disease of the parotid arise within the para-glandular or intraglandular lymph nodes.

Depending on the site, Castleman disease can be classified into two groups: localized, unicentric or focal Castleman disease and disseminated or multicentric Castleman disease. Localized CD presents as a slow growing mass and has a benign prognosis. Multicentric CD manifests with diffuse lymphadenopathy within at least one regional group of lymph nodes and has a poor prognosis. Castleman disease is divided into three subtypes depending on histopathological examination. The first type is hyaline-vascular type, which is characterized by concentrically arranged small lymphocytes (onion skinning) around the numerous small, follicle-like structure with prominent vascular proliferation and hyalinization is more common in young adults and older children. It is usually found in a localized form without constitutional symptoms and has a more favorable progress than the other subtypes. Histopathological examination of our case was consistent with this type.

The plasma cell type is characterized by sheets of mature plasma cells in the interfollicular spaces and larger hyperplastic follicles with less vascular proliferation. Twenty-two percent of the localized and majority of the multicentric Castleman disease is plasma cell type. It is more frequently associated with constitutional symptoms such as fever, fatigue, weight loss and increase of erythrocyte sedimentation rate, thrombocytosis, anemia, hypalbuminemia and polyclonal hyperglobulinemia. These are more common in children. Systemic chemotherapy including cyclophosphamide, vincristine sulfate, doxorubicin and glucocorticoids may be required in this type of Castleman disease. Radiotherapy is an alternative in cases of unresectable lesions, incomplete excision, recurrence, and poor surgical candidacy. The mixed type is rare and is histologically a mixture of two other types, and symptom of this type is associated with cell component.

The origin of this entity is still unknown. Some authors favor a theory of lympho-proliferation due to chronic antigenic stimulation by a virus or chronic inflammation while others consider it to be a lymphoid hamartoma. Systemic chemotherapy including cyclophosphamide, vincristine sulfate, doxorubicin and glucocorticoids may be required in this type of Castleman disease. Radiotherapy is an alternative in cases of unresectable lesions, incomplete excision, recurrence, and poor surgical candidacy. The mixed type is rare and is histologically a mixture of two other types, and symptom of this type is associated with cell component.

The origin of this entity is still unknown. Some authors favor a theory of lympho-proliferation due to chronic antigenic stimulation by a virus or chronic inflammation while others consider it to be a lymphoid hamartoma. Clinically Castleman disease is not common and the diagnosis is challenging due to its rarity and the absence of specific signs. Most patients presented with slowly enlarging
 unicentric or multicentric masses in the parotid and/or neck region without any obvious symptoms as seen in our case. The clinical manifestations of this disease, are not specific and are always similar to the benign lesions of the parotid and neck region. The unicentric lesion in the parotid gland is difficult to differentiate from Warthin tumor, pleomorphic adenoma, schwannoma. Multicentric disease, may be associated with polyneuropathy, organomegaly, endocrinopathy, anemia. Imaging examination can be useful for the diagnosis and the differential diagnosis. Ultrasonography is efficient to evaluate the hypervascular nature of the disease. Computed tomography scan with contrast shows a densely enhancing, well-circumscribed, homogenous mass which reflects the hypervascularity of these lesions. However, to date no radiological study is diagnostic. Magnetic resonance imaging can give a useful information, such as rich vascular bundles from local blood vessels, because the masses of Castleman disease are always observed with a rich blood supply during the surgical operation. The mass showed low signal in T1 weighted sections and in T2 weighted sections it was with high signal intensity.

The treatment of choice for this disease is surgical excision. The excision is both diagnostic and therapeutic. Recurrences are rare in the hyaline vascular type; however, follow-up is required in plasma cell type lesion.

In conclusion, Castleman disease is a rare entity, especially in the parotid region. Though it has a benign course, differential diagnosis from other head and neck masses may be challenging and awareness of the physician is required.

References