CASTLEMAN’S DISEASE OF THE PAROTID GLAND: A CASE REPORT

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Castleman’s disease (CD) was first identified in 1954 as a mass in the mediastinum resembling a thymoma [1]. CD is a rare, benign lymphoproliferative disorder of unknown cause. CD may be found in lymph nodes in any body regions, although over 60% of cases are located in the mediastinum. The head and neck is the second most commonly involved site, with up to 14% of cases. Of these, nearly 85% are located in the neck. These lesions rarely occur in the salivary glands. Clinically, CD can be divided into unicentric and multicentric forms. We report an unusual case with unicentric CD presenting as a parotid tumor in a 34-year-old woman. The lesion was found coincidentally during routine cranial magnetic resonance imaging in the absence of symptoms such as swelling or a mass in the parotid region.

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CASE PRESENTATION

A 34-year-old woman was admitted to our clinic for evaluation of an abnormal cranial magnetic resonance imaging (MRI) finding. Approximately 1 year earlier, she had undergone surgery for a cerebellar mass. The abnormal finding was seen in her routine cranial MRI scan, with a 10 × 15-mm mass in her left parotid gland (Figure 1). She denied any constitutional symptoms except tinnitus in her left ear lasting for 6 months. Physical examination revealed no mass or swelling in the left parotid region. The facial nerve functions were intact. There was no associated lymphadenopathy. Her complete blood count, chemistry profiles and chest X-ray findings were all within normal limits. Ultrasonography demonstrated a 14 × 15-mm mass in her left parotid gland with hypoechoic vascularity. Computed tomography (CT) scans showed a well-marginated, round, homogenous mass in the posterior region of the angulus mandible extending into the deep lobe of the left parotid gland (Figure 2). Ultrasound-guided fine-needle aspiration was performed and the
biopsy was non-diagnostic. The patient underwent a total parotidectomy with preservation of the facial nerve. After dissecting the superficial parotid lobe from the facial nerve, a $15 \times 15 \times 10$-mm round, solid grey mass was found in the deep lobe. The mass was in the anteroinferior location of the main truncus of the facial nerve. There was no adhesion between the mass and the surrounding gland parenchyme or facial nerve. The deep lobe containing the mass was carefully dissected. Histologic analysis of the surgical specimen showed the CD was of the hyaline-vascular type, and no further treatment was given (Figure 3). Postoperatively, the patient made an excellent recovery and continues to be free of disease 8 months after surgery.

**DISCUSSION**

CD is an uncommon benign lymphoproliferative disorder with unknown etiology. CD has been referred to by many names, including giant lymph node hyperplasia, angiomatous lymph node hamartoma, angiofollicular lymph node hyperplasia, follicular lymphoreticuloma and benign giant lymphoma [1–6]. This variable terminology reflects its uncertain cause, although the most tenable theories of pathogenesis are that CD is derived from hamartomatous or inflammatory processes. CD mainly affects young adults aged 15–35 years old, but shows no sex predilection [6].

The pathogenesis of CD is not well understood. Some authors have proposed a reactive lymph node hyperplasia theory in which the disease is an immunologic response whereas other authors have suggested it is derived from a benign tumor or hamartoma [1,7,8].

CD may occur at any lymph node in the body, although the mediastinum is the most common location of the disease, accounting for 60% of all occurrences, followed by the head and neck region accounting for 14% of occurrences [2]. Additional sites of occurrence include the axilla, retroperitoneum, mesentery, vulva, pancreas, and pelvis [5]. Within the head and neck, 85% of lesions occur in the neck, while salivary gland involvement is very rare.

 Clinically, there are two forms of CD, unicentric and multicentric. The unicentric form, as in our case, is the
most common form and usually presents in a lymphonodular region in the absence of other symptoms [1,2,9]. The multicentric form is often associated with fever, diaphoresis, fatigue, weight loss and disorders, e.g. anemia, hyperglobulinemia, nephrotic syndrome, elevated erythrocyte sedimentation rate and hepatosplenomegaly [9]. This form has also been associated with some malignancies, e.g. Kaposi’s sarcoma, non-Hodgkin’s lymphoma, Hodgkin’s disease and POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, M protein and skin changes) [1,9,10]. It is also associated with human immunodeficiency virus and human herpes virus [10,11]. Oral erosive lichen planus and pemphigus have also been reported to be associated with multicentric CD [12].

Histologically, there are two different types of CD, the hyaline-vascular type and the plasma cell type. The hyaline-vascular type, as in our case, is the most common type and accounts for 90% of all cases [4]. It usually affects a single lymph node. The histologic appearance of this type includes lymphoid follicles surrounded by concentric layers of small lymphoid cells with an “onion skin” appearance. The atrophic germinal centers are traversed by mantle zone lymphocytes. The plasma cell type is a relatively uncommon type of CD and is associated with systemic symptoms and abnormal hematologic values. Histologically, the plasma cell type is characterized by a dense infiltration of plasma cells and involves several lymph nodes. However, intermediate forms showing transition between the two types have been also described [13]. It has been suggested that the plasma cell type is an earlier stage of the hyaline-vascular type or that both types represent different responses of the same process [1,2,4,6,14].

Although CD may be included in the differential diagnosis of lymphoproliferative disorders in the head and neck, CD in the parotid gland poses considerable diagnostic difficulties owing to its unusual location and its tendency to mimic neoplasms. In our case, MRI and CT findings and the patient’s medical history suggested that the mass was a benign neoplasm.

Tests can occasionally be helpful for differential diagnosis. CT scans show dense, homogenous enhancement after intravenous contrast administration [2,15]. The hyaline-vascular type tends to show greater enhancement owing to its hypervascularity [5]. Although MRI features are not specific, CD should be considered in the differential diagnosis for a solitary neck mass showing low to intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images [7,15].

The definitive diagnosis of CD is based on histologic analysis [1,2,3,9]. The treatment choice for unicentric lesions is excision of the mass. If excision is complete, recurrence is uncommon. Although Sanchez-Cuellar et al recommended chemotherapy for the plasma cell type; most authors suggested surgical excision [1]. The systemic symptoms usually abate in 30–60 days. The multicentric form of CD has a poor prognosis and warrants aggressive therapy. Due to the multisystem involvement of multicentric CD, treatments may require surgery plus chemotherapy [2].

In conclusion, CD is a benign lymphoproliferative disease that rarely occurs in the head and neck region. CD should be included in the differential diagnosis when considering a lymphoproliferative lesion in the parotid gland. The hyaline-vascular type is more common than the plasma cell type. Surgical excision is the preferred treatment for unicentric CD, whereas the multicentric form requires more aggressive treatment and long-term follow up.

**References**


