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Neck Localisation of Castelman's Disease Mimicking A Metastatic Lymph Node: A Case Report and Review of Literature

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Case report

Castleman's Disease (CD) was first described by Benjamin Castleman et al, who reported 13 cases of mediastinal lymphadenopathy in 1954 [1]. It is an uncommon disease of lymph node hyperplasia and is also known as angiofollicular lymph node hyperplasia, giant lymph node hyperplasia, and angiofollicular lymphoid hamartoma [2].

Since then, the clinical and pathological reports about CD cases throughout the whole body have been increasingly reported in the literature [2-3,5].

Our understanding of CD has progressively evolved with time [3,4,5,]. This clinical entity is not common and usually occurs in adults but rare in children [6,7]. The most common site of CD is the mediastinum, followed by the neck, retroperitoneum, and axilla [3].

Clinically, these diseases can be categorized into two main forms: Unicentric CD (UCD) and Multicentric CD (MCD). The UCD typically presents as isolated lymphadenopathy and usually be asymptomatic, whereas the latter often involves multi-

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ple lymph nodes and organs, and also associates with systemic symptoms such as recurrent fever, anemia, night sweats, malaise, and hepatosplenomegaly [10].

Castleman's disease rarely involves the head neck region and is extremely uncommon in the oral maxillofacial region [10,11].

The diagnosis of this rare disorder largely depends on pathological findings and can be histologically classified into three main subtypes: hyaline-vascular, plasma-cell, and mixed type.

Generally, most UCD cases are identified as the hyaline vascular type and treated by surgical excision with favorable prognosis [5]. However, patients with MCD are plasma-cell or mixed subtypes and usually require systemic therapy, but with inferior prognosis [3,4,11].

In this article, we describe a case of CD in the neck mimicking a metastatic lymph node with an unusual presentation, management and pathological characteristics of this disease are discussed, together with a literature review.

Case report

This is a 28 year old, single, unprofessional patient with no medical and surgical history, who consulted in our depatement for a swelling of the soft palate gradually increasing in volume, painful associated with lateral cervical lymphadenopathy all evolving in a context of apyrexia and conservation of the general state.

Clinically, the patient is conscious, in good general condition, stable on the hemodynamic and respiratory levels, non-pyretic.

Examination of the oral cavity finds a firm bilobed tissue mass occupying the soft hemi-palate, about 5cm with a healthy mucosa facing without bleeding or superinfection.

The cervical examination finds a cervical lymphadenopathy of the left area III, mobile with respect to the two superficial and deep planes, of about 3cm, the rest of the lymph nodes are free.

We completed with a nasofibroscopy in search of a secondary location, which objectified normal and healthy mucosa on the nasopharyngeal and laryngeal stage.

There is also the absence of an infectious dental or pharyngeal focus explaining the presence of cervical lymphadenopathy.

The examination was completed by the following by a computed tomography and cervico-facial magnetic resonance imaging which were in favor of a tumor process at the expense of the soft palate, lateralized to the left oval, well limited measuring 43 * 40 * 26 mm , T1 hyposignal, T2 intermediate signal, slightly raised after injection of contrast medium.

It comes into contact with the hard palate and the base of the tongue, the pterygoid process and the medial pterygoid muscle, with loss of the fatty border in places, reducing the lumen of the oropharynx.

Associated with a 22 mm benign jugulo-carotid lymphadenopathy with a few nodes under the angulo-mandibular, and infra-centimetric spines.

The decision was taken for exeresis of the palatal tumor with an exploratory cervicotomy followed or not by a lymph node dissection according to the extemporaneous examination.

The extempanous examination of the lymphadenopathy was in favor of a benign mass without signs of malignancy and the final examination is in favor of an angiofollicular hyperplasia compatible with a Castelman disease of the hyalino-vascular type.

The pathological examination is in favor of a pleomorphic adenoma of the palate. The follow-up was marked by good development and good healing.

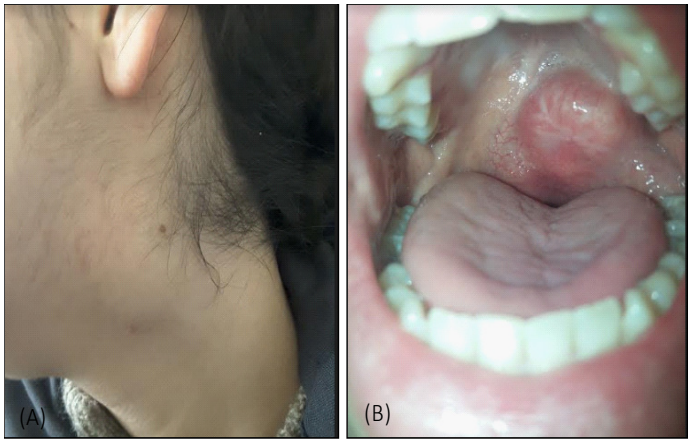


Figure 1: Preoperative aspect of the palatal tumor **(A)** and cervical lymphadenopathy **(B)**.

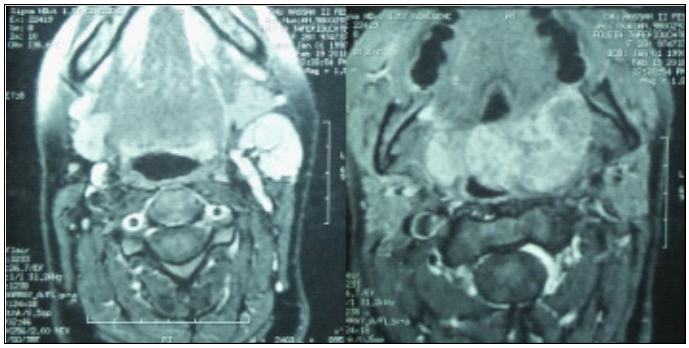


Figure 2: MRI image showing the palatal tumor process and the homolateral cervical lymphadenopathy.

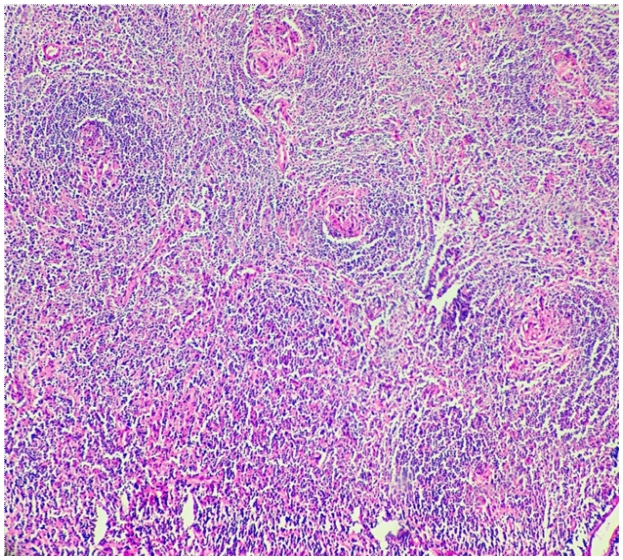


Figure 3: Castelman disease Lymph node tissue with lymphoid follicles with a small atrophic germ center, containing one or more capillaries with a hyaline wall and concentric deposits of lymphocytes in an "Onion Bulb".

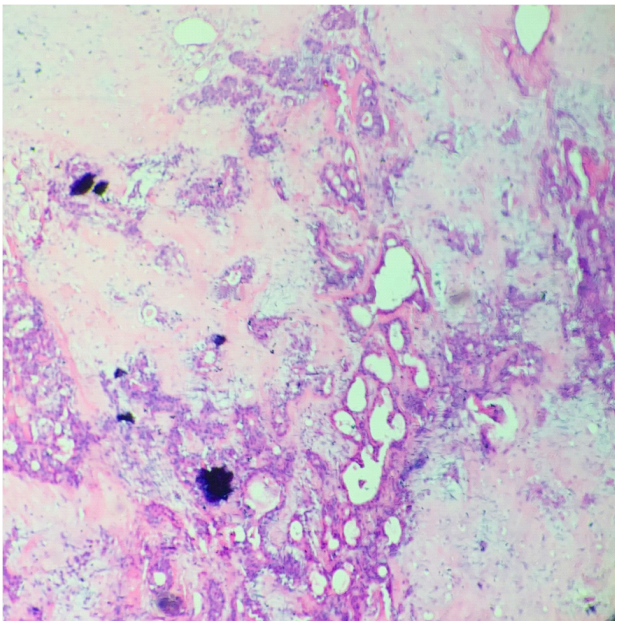
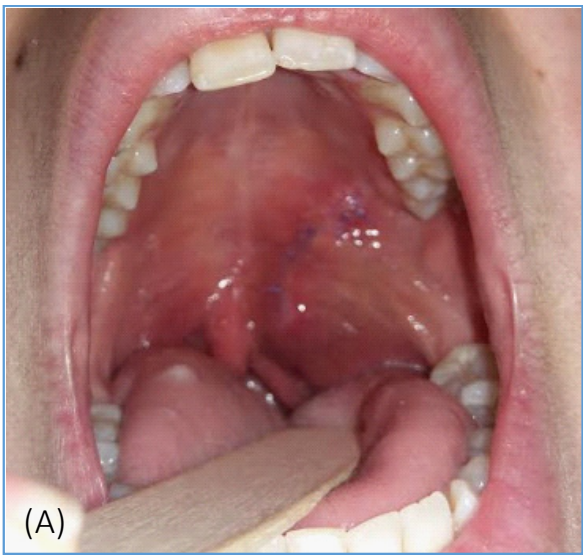


Figure 4: Pleomorphic adenoma Benign tumor proliferation with a double mesenchymal contingent, within a chondromyxoid and fibrohyaline stroma.



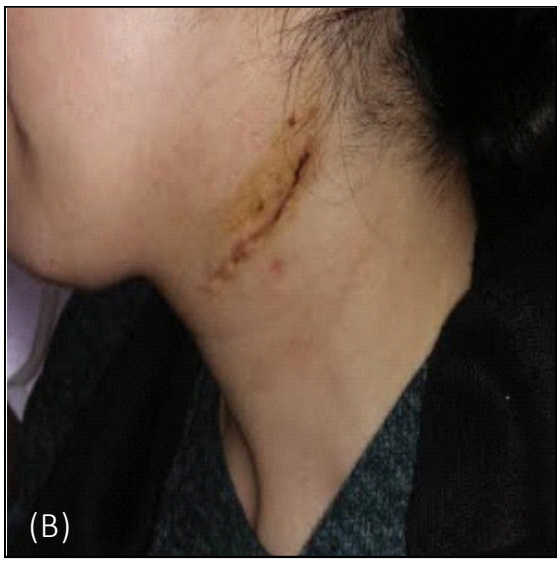


Figure 5: Post-operative aspect.

Discussion

CD is a rare disorder of the lymphoid tissue. Since the first report in 1954, just over 500 cases have been reported, of which 57 were localized to the cervical region. Patient ages ranged from two months to 76 years, with a peak incidence in the second and third decades [1].

No predilection for either gender noted. Cervical CD is very uncommon in children, with only 24 cases reported previously in the literature, comprising only 23% of CD in children [1,2].

The etiology of CD remains unclear, but there is evidence that certain viral infections or a chronic inflammatory reaction will induce the disease [13,14]. It has attracted attention because of its association with the human immunodeficiency virus and human herpesvirus 8 [15,16].

There are 2 histologic subgroups of Castleman's disease: Hyaline vascular (80–90% of cases) and plasma cellular type.

The hyaline vascular type is characterized by small lymphoreticular follicles scattered in hypervascular hyalinized stroma. The plasma cellular type is rare but more aggressive, characterized by mature plasma cell clusters between lymph follicles. The follicles are round and variable in size, surrounded by a cuff of small lymphocytes arranged in concentric "Onion Skin" layers, with germinal centers frequently demonstrating atrophy with radically penetrating blood vessels [17].

The pathological findings in our patient were compatible with the hyaline vascular type.

Casper suggested an algorithm for the evaluation and management of patients suspected to have Castleman's disease in 2005 [18].

Excisional biopsy is still an essential procedure for diagnosis. CT is another useful tool for ruling out Castleman's disease from lymphoma or thymoma 6. The lesions of Castleman's disease demonstrate enhancement on CT scan, but lymphoma and thymoma do not.

Surgery is the treatment of choice in cases of giant lymph node hyperplasia, whereas chemotherapy, radiotherapy, and steroids are proposed for the multiple forms. No recurrences have been reported in the literature after complete resection of the hyaline vascular type.

Sanz et al [14] reported the only case of the plasma cell type in the head and neck, which recurred after 11 months. Our case showed no progression or recurrence over 24 months of follow-up.

The Multicentric form of CD (MCD), characterized by multiple systemic symptoms including hepatosplenomegaly, recurrent fevers, night sweats, and lymphadenopathy, was later described.

The most common therapy for Multicentric CD is high dose steroids, chemotherapy alone, or combined therapy. Because of the nature of disseminated lymphadenopathy, complete resection is rarely possible. Radiotherapy is another choice but only 23% of patients have favorable responses [20].

Alternative therapies including anti-IL-6 monoclonal antibodies interferon-alpha, antiviral therapy and high-dose melphalan with autologous bone marrow transplantation have been reported.

The prognosis of multicentric disease is poor, with a median survival range from 14 to 30 months.

The main causes of mortality are infections which cause sepsis, multi-organ system failure, and malignancies. According to some studies, multicentric disease may have a risk of progressing to lymphoma and Kaposi's sarcoma [19].

Conclusion

CD is an uncommon disease of unknown etiology that induces reactive lymph node hyperplasia. Histopathological evaluation is the only way to make a definite diagnosis.

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