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Dr. Keerthana Muppidi
CMR Institute of Medical Sciences, Kandlakoya, Hyderabad, Telangana, India

Dr. Nabila Afsar
CMR Institute of Medical Sciences, Kandlakoya, Hyderabad, Telangana, India

Corresponding Author:
Dr. Keerthana Muppidi
CMR Institute of Medical Sciences, Kandlakoya, Hyderabad, Telangana, India

Castleman disease in parotid gland - a rare disease in a rare location

Keerthana Muppidi and Nabila Afsar

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Abstract

Background: Castleman's disease is a rare lymphoproliferative disorder that predominantly affects mediastinal lymph nodes but can involve any nodal region, including the head and neck. Involvement of the parotid gland is extremely uncommon and often mimics primary salivary gland tumors, making accurate diagnosis challenging.

Case Presentation: We report a case of a 14-year-old male who presented with a gradually progressive, painless swelling in the right parotid region for one year. There was no history of trauma or systemic symptoms. Clinical examination revealed a solitary, firm, mobile mass without overlying skin changes. Routine laboratory investigations were unremarkable. Ultrasonography demonstrated a hypoechoic lesion within the right parotid gland, suggestive of pleomorphic adenoma. Initial and repeat fine needle aspiration cytology yielded reactive lymphadenitis. Due to persistence of the swelling, surgical excision was performed. Histopathological examination revealed partial effacement of nodal architecture, atretic germinal centres traversed by hyalinised vessels ("lollipop appearance"), concentric layering of mantle zone lymphocytes ("onion-skinning"), perivascular hyalinisation, and extensive interfollicular vascular proliferation, confirming the diagnosis of hyaline vascular type unicentric Castleman's disease.

Conclusion: Castleman's disease involving the parotid gland is a rare clinical entity that should be considered in the differential diagnosis of persistent parotid swellings in children and adolescents, particularly when imaging suggests a benign neoplasm and cytology remains inconclusive. Histopathological examination is the definitive diagnostic modality. Complete surgical excision is both diagnostic and curative, and awareness of this rare presentation is essential to prevent misdiagnosis and ensure appropriate management.

Keywords: Castleman disease, parotid gland, Unicentric, hyaline vascular

Introduction

Castleman disease also known as giant lymph node hyperplasia, angiomatous lymphnode hamartoma, is a rare lymphoproliferative disorder that primarily involves lymph nodes [1].

It can affect any lymph node in the entire body, including the head and neck region. But, the parotid gland is very rarely involved [2].

Castleman disease can be unicentric or multicentric. There are two morphologic variants of unicentric CD: hyaline vascular and plasma cell. HV variant is most common type.

Patients with hyaline vascular type are often completely asymptomatic and the lesion is detected accidentally. Laboratory abnormalities are rare in these patients; serum LDH levels can be elevated in a subset of patients.

Case presentation

Patient Presentation

A 14-year-old male presented with a gradually progressive swelling in the right parotid region for one year. There was no history of trauma or fever.

Clinical Examination

Local examination revealed a solitary, firm, mobile lump in the right parotid area. The overlying skin was normal, with no signs of inflammation, discharge, or sinus formation. No other palpable swellings were identified.

Investigations

Routine laboratory investigations, including complete blood counts and urine examination, were within normal limits. Ultrasonography demonstrated a hypoechoic lesion measuring 22 × 12 × 19 mm within the right parotid gland, initially suggestive of a pleomorphic adenoma. A few subcentimetric cervical lymph nodes at levels 2b and 3 were also noted.

An initial fine needle aspiration cytology (FNAC) was reported as reactive lymphadenitis. Due to persistence of the swelling over the subsequent year, a repeat FNAC was performed, yielding identical findings.

Management and Histopathology

The patient underwent surgical excision of the lesion.

Gross examination revealed a grey-brown nodular mass measuring 2.5 × 2 × 1 cm. The cut surface appeared diffusely grey-white to grey-brown.

Microscopic findings included

Partial effacement of lymph node architecture with obliteration of sub capsular and medullary sinuses is seen. Atretic germinal centres traversed by hyalinised penetrating vessels, creating a characteristic "lollipop appearance", follicles displaying "onion-skin" layering of mantle zone lymphocytes are noted as shown in figures 2 and 1. Focal twinning of germinal centers as shown in figure 3, perivascular hyalinisation, along with broad fibrotic and sclerotic bands, prominent vascular proliferation in interfollicular areas are also seen. Adjacent normal salivary gland acini are also included in the biopsy.

Based on the distinctive histopathological features, a diagnosis of Hyaline Vascular Type Castleman's Disease was established. It is noteworthy that immunohistochemistry has limited diagnostic utility in such cases.

Immunohistochemistry, though sometimes used to exclude lymphoma, generally has limited additional diagnostic value in typical cases, and was therefore not pursued here.

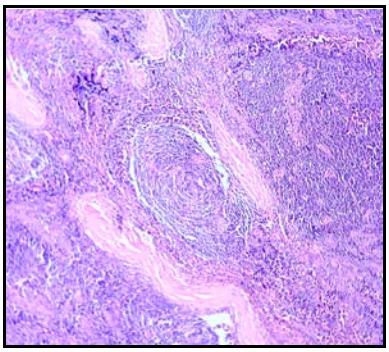

Fig 1: Haematoxylin and eosin (10 X) stained section shows an onion peel appearance

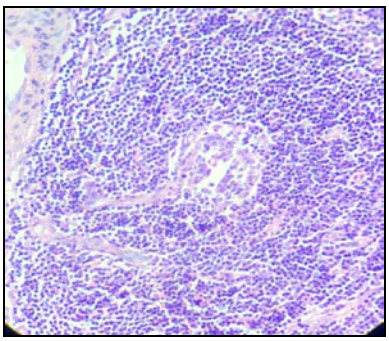

Fig 2: Haematoxylin and eosin (40 X) stained section shows a lollipop appearance.

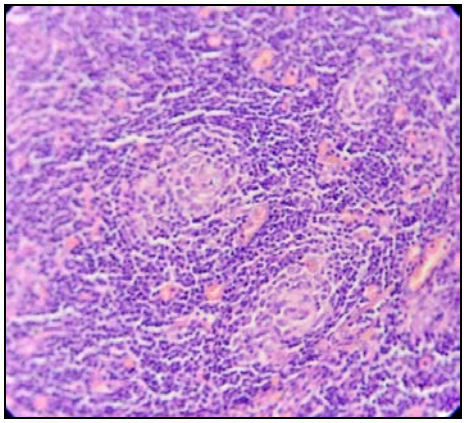

Fig 3: Haematoxylin and eosin (40 X) stained section shows twinning of germinal centres

Discussion

Castleman's disease is a rare lymphoproliferative disorder. The second most common site of occurrence is head and neck region. But, the salivary glands are rarely affected.

Unicentric castleman's disease poses a diagnostic challenge, due to its relative absence of symptoms and lack of specific markers and specific radiographic characteristic.

Histologically, it is of 2 subtypes: (1) hyaline vascular type and (2) plasma cell rich.

The cause and mechanism underlying Castleman's disease are not yet fully understood. It is proposed that the condition may arise due to persistent antigenic stimulation, potentially of viral origin. Excessive secretion of interleukin-6 (IL-6) by the affected lymph nodes is thought to contribute significantly to disease development, owing to its roles in promoting blood vessel formation (angiogenesis) and stimulating hematopoietic activity [3].

The parotid region involvement, though rare, has been documented in the literature and can mimic salivary gland neoplasms, particularly pleomorphic adenoma, as seen in the ultrasonographic findings in our patient [4]. Differentiation from other causes of lymphadenopathy, including reactive hyperplasia, lymphoma, and metastatic disease, is essential, particularly when cytology yields non-specific findings [5, 6].

In a case report by Fawaz Abo-Alhassan *et al* [7], 29-year-old Asian lady who presented with a 2-year history of an enlarging left parotid mass. Histopathology of the excisional biopsy confirmed the diagnosis of Castleman disease.

Iaconetta G *et al* [3] presented a case 35-year-old woman with a swelling in the right submandibular lodge. The histopathological examination was in favour of a hyaline vascular type of giant lymph node hyperplasia.

In a case report by Sean W.Delaney *et al* [8], an 11-year-old female presented with a 3-year history of a slowly enlarging asymptomatic 3.5cm right facial mass and a 7-year-old male presented with a painless right facial mass for one year.Both cases were diagnosed as castleman diseases (hyanline vascular type). Similar to the present case report in which castleman disease is diagnosed in a 14 year old child with painless right facial swelling.

This case highlights the importance of considering Castleman's disease in the differential diagnosis of persistent parotid region swellings in children and adolescents, especially when imaging suggests a benign neoplasm and FNAC remains inconclusive.

Conclusion

Castleman's disease involving the parotid region is an uncommon clinical entity that can closely mimic primary salivary gland tumors, leading to diagnostic challenges. This case highlights the importance of maintaining a high index of suspicion in patients with persistent parotid swellings unresponsive to conservative management and showing inconclusive cytological findings.

Histopathological examination remains the definitive diagnostic modality, with characteristic features such as atretic germinal centres with hyalinised vessels and concentric mantle zone layering confirming the diagnosis.

Complete surgical excision is both diagnostic and curative in unicentric hyaline vascular Castleman's disease, and long-term prognosis is excellent. Awareness of this rare presentation is essential to avoid misdiagnosis and ensure appropriate management.

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