CASTLEMAN’S DISEASE OF THE NECK — A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Castleman’s disease is a rare lympho proliferative disorder with unclear pathogenesis. It is found in two forms, localized (uni-centric) and multi-centric. Histologically there are two forms; hyaline vascular and plasma cell type.

The disease is definitely diagnosed by histological examination after biopsy. Computed Tomography (CT), Magnetic Resonance Imaging (MRI), Gallium 67 scintigraphy can prove to be useful adjuncts.

Following is a case report of 20 years old male patient reporting with mass in neck for the last 10 years. On FNAC the lesion was confused with lymphoma. Excisional biopsy was performed and the histopathology showed the lesion to be hyalinized vascular type of castleman’s disease.

Key Words: Lymph node, cervical lymphadenopathy, Hylization and Calcification, Castleman Disease.

INTRODUCTION

Castleman’s disease (CD) is a rare lympho-proliferative disorder. It is also known by many other names such as angiofollicular lymph node hyperplasia, giant lymph node hyperplasia, and angiofollicular lymphoid hamartoma. The pathogenesis of this rare condition remains unclear. Different theories have been suggested in past like hyperplastic response to chronic infection, immunologic abnormality of B cells and hamartamous proliferation of lymphoid tissue. Recently IL-6 has been linked in the pathogenesis of the systemic forms.

CD occurs in two clinical forms, localized (unicentric) and multicentric. Localized form is usually benign while the multicentric form is associated with systemic signs and symptoms. Also the multicentric form is more aggressive. Histologically the disease may occur in hyalinized vascular pattern (HV) or the plasma cell variant (PC); a small percentage also presents with a mixed histological appearance.

The diagnosis of CD on the basis of clinical signs symptoms is difficult owing to diverse presentation. The disease is definitely diagnosed by histologic examination after biopsy. Computed Tomography (CT), Magnetic Resonance Imaging (MRI), Gallium 67 scintigraphy can prove to be useful adjuncts. The imaging characteristics vary. Plain radiographs may show radiating pattern of calcification. On ultrasound the appearance is similar to a lymphoma. A prominent vascular pattern is seen angiographically and may cause profuse hemorrhage following biopsy. CT findings include homogeneous soft tissue mass, with intense early contrast enhancement seen in the hyaline-vascular Form. On MRI Low signal intensity is seen on T1W unenhanced images and high signal intensity seen on T2W images and T1W contrast enhanced images. The disease is often treated by surgical excision. Recurrence in the unicentric variety is low and prognosis is favorable. Radiotherapy is also considered as an alternate option for patients who are poor candidates for surgery.

CD of the neck is a rare occurrence. Due to the similarity with lymphoma, it is important to timely diagnose the condition in order to avoid over and aggressive treatment. A case of adult cervical CD presenting as a solitary neck mass is reported here.

CASE HISTORY

A 20 years old male patient reported with a history of swelling in the left sub-mandibular area for the last ten years (Fig 1), according to the patient, there is a recent increase in size and along with pain the pain is dull in nature and very well localized. The patient was extremely concerned and was anxious, as he was
from a poor socio-economic background and was under treatment of so many local dentists/physicians had a big envelope in the hand full of prescription papers. On extra oral examination there was slight facial asymmetry because of the swelling in the sub-mandibular area. On palpation there was cervical lymphadenopathy. On intra oral examination the patient had complete set of teeth present with no the dental/medical history. No bony or dental abnormality was found on radiological examination i.e., Orthopantomograph (OPG). A CT scan was advised which revealed the involvement of a single group of submandibular lymph nodes as a homogenous mass on the ipsilateral side. A decision was made to perform Fine Needle Aspiration cytology (FNAC). The FNAC revealed reactive lymphadenitis with a possibility of non-hodgkin's lymphoma, which further increased the suspicion, a scalpel biopsy was decided to confirm the diagnosis. On subsequent Scalpel Biopsy, lesion was confirmed as lymph node with extensive hyalinization and calcification i.e., Castleman’s Disease. The patient was counseled and was perfectly well at follow up visits at one month and 6 months period (Fig 2).

DISCUSSION

CD was first reported by Castleman and Towne in 1954. They described it as mediastinal lymph node hyperplasia similar to thymoma. In 1969 a case series of a group of patients with CD was published. Keller et al further specified two distinct groups of CD based on histopathological examination. After that the multicentric form of this disease was reported by Gaba et al in 1978.11

CD affects patients of all races and gender however it has a varying age distribution. The prevalence of CD is expected to be 1:100,000.12 The onset may be at any age from adolescence to seventh decade of life.13 Cervical CD in children is rare. Only a few cases have been reported. The most common age of presentation is between 10 and 18 years.14 Age of the patient in this case report was 20 years but the swelling was present since 10 years, meaning the onset was in childhood. Boguila et al reported a similar case of cervical CD in a child patient of age 13 years.15 CD is reported to be a rare occurrence in pediatric neck, only 18 reports have been found with neck involvement.16

The patient in current case report was HV type of localized form of CD. Among the two forms of CD, localized unicentric type is more common. Histologically HV type occurs in about 85% of patients and PC type in 15%. Moreover a small percentage of people also show a mixed form. The HV type is often asymptomatic while PC type shows more aggression along with constitutional signs symptoms and laboratory abnormalities.17 Clinically the most common presentation is that of a slowly growing painless mass. Regarding site distribution 86% of cases involve the mediastinum. Among the head and neck region, neck is the most common extra thoracic site involved (6%). It either presents under the sternomastoid or as an extension from the mediastinum. Less common sites include parotid (either intra-glândular or para-glândular lymphoid tissue), palate and tongue.18 According to Rhee et al the most common site for CD was the lymph nodes in the mediastinum and least common sites were the neck, abdomen, and extra-nodal tissues such as the buccal cavity, vulva, skin, and muscle.19 The most common presentation of cervical CD is level II lymph node involvement.20 In a 1980 review of 315 cases of localized CD, 65% were in the mediastinum, 16% in the neck, 12% in the abdomen, and 3% in the axilla.21 In a 2005 review of 195 cases of abdominal and retroperitoneal CD localizations, 47 cases were in the abdomen, 27 in the pelvis, and 97 in the retro-peritoneum.22
It is important to include CD in differential diagnosis of cervical swellings. It is a dilemma because the differential diagnosis includes head and neck tumors. This disease has been termed as “lymphoma imposter” by Denenberg and Levine. It can be confused with unknown primaries as well. Other differentials include infectious and inflammatory lesions such as lymphadenitis, tuberculosis, sarcoidosis, toxoplasmosis, cytomegalovirus, mononucleosis, HIV, and cat scratch disease. Likewise, some tumors must be included, such as neurofibroma, cervical lipoma, Hodgkin disease, thymoma, non-Hodgkin disease, and lymph node metastasis. As in this case report the disease was confused with lymphoma on FNAC, other cases have been reported in which CD was confused with Warthin’s tumor on FNAC.

As the patient had no other signs symptoms, an FNAC was advised. FNA is considered helpful by some authors in the diagnosis of CD. Hyalinized small blood vessels penetrating follicular germinal centers are characteristic of HV-CD. Ultrasound guided FNAC is more accurate and less traumatic than lymph node biopsy. However according to other reports there is limited role of FNA in the diagnosis of this condition although physicians are still encouraged to advise FNA in cases of persistent neck swellings to rule out other conditions.

The indicators for diagnosis of HV-CD on histopathology include lymphoid proliferation, where the follicles are regressed or depleted of germinal center cells and have expanded mantle zones with small lymphocytes arranged concentrically in an “onion-skin” fashion, an interfollicular region variably expanded with prominent hyalinized blood vessels; expanded, often dysplastic, follicular dendritic cell networks; myoid cells; dendritic reticulum cells; and small, mostly T lymphocytes.

Surgical excision of the lymph node was performed after which the patient was followed up for six months. Similar results were obtained after surgical excision of localized cases of CD by others as well.

CONCLUSION AND RECOMMENDATIONS

Castleman’s disease should be considered in the differential diagnosis of the sub-mandibular swelling. A detailed counseling with these patients and proper execution and planning is important to avoid unnecessary psychological disturbances.

REFERENCES
Castleman's disease of the neck


CONTRIBUTIONS BY AUTHORS

1 Muslim Khan: Principle author of the case report/ operating surgeon of the case.

2 Bushra & Saima Mahboob: Helped in literature review search and discussion writing.