A case report on unicentric form of Castleman's disease

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ABSTRACT

Castleman's disease, also known as angiofollicular lymph node hyperplasia, is a heterogeneous group of lymphoproliferative disorders. It is a rare disease and difficult to diagnose preoperatively. Localized Castleman's disease is almost always hyaline vascular variant type. We present the case of a 30-year-old male with swelling over the left side of the neck for 10 years which was diagnosed as Castleman's disease on histopathology.

Key words: *Angiofollicular hyperplasia, Castleman's disease, Rare*

Castleman's disease (CD) is a proliferative disorder of lymph nodes and related tissues. It was first described by Dr. Benjamin Castleman in 1956 [1]. It is also known as giant lymph node hyperplasia and angiofollicular lymph node hyperplasia [2]. It is not officially cancer, but the multicentric form of this disease acts very much like lymphoma. There are two main forms – localized and multicentric. The localized CD only affects a single group of lymph nodes and lymph nodes in the chest or abdomen are affected most often. The mediastinum is the most common site, accounting for 60% of cases and the neck is involved in only 14% [3]. Multicentric CD affects more than a single group of lymph nodes. It can also affect other organs containing lymphoid tissue. This form sometimes occurs in people infected with human immunodeficiency virus (HIV). The multicentric CD is more serious than the localized type, particularly in people with HIV infection.

CASE REPORT

A 30-year-old male presented with a complaint of painless swelling over the left side of the neck for 10 years. There were no constitutional symptoms. The swelling was initially small and was gradually increasing in size. On general examination, the vitals were stable. There was no generalized lymphadenopathy and organomegaly. On neck examination, the lesion was firm, non-tender, movable, and non-pulsatile and showed well-defined margins, measuring 7 cm×4 cm.

On routine blood investigation, complete blood count and other investigations were normal. The Erythrocyte sedimentation rate (ESR) was 30 mm. Liver and kidney function tests were found to be within the normal range. C-reactive protein (CRP), HIV, and hepatitis B surface antigenemia were non-reactive. On ultrasonography, the neck showed a large soft tissue mass measuring 6.8 cm×4.0 cm with echogenicity in the left side of the neck, separate from the thyroid gland. On contrast-enhanced computed tomography of the neck region, a soft density mass of size 7 cm×4 cm with enhancement seen on the left perivascular region along with a calcified focus was seen. A few subcentimetric size lymph nodes in the left jugular chain and the left cervical region were also present.

Based on these findings, the possibility of neurogenic tumor/glomus tumor was given. Fine needle aspiration was done using a 24-gauge needle. The slides were stained with giemsa stain. Smears revealed features of reactive lymphadenitis (Fig. 1). Excision biopsy was planned. Under all aseptic conditions, the patient was operated under general anesthesia and excised tumor tissue was sent for histopathological examination. On gross examination, it consists of a single gray-white tissue bit measuring 8 cm×3 cm. Cut surface is soft homogenous gray-white and rubbery in consistency (Fig. 2).

Sections showed the presence of lymphoid tissue surrounded by capsule and nerve tissue. The architecture of the node was mildly effaced. There was lysis of follicular architecture with the expansion of the paracortical and interfollicular region. Hyalinized follicle, interfollicular vascular proliferation, and infiltration by many eosinophils, few plasma cells and neutrophils were also evident. At the intersection of a capsule and nerve tissue, few foreign body giant cells were seen. CD 21 was found to be positive in the follicular dendritic cells (Fig. 3). On the basis of the above findings, the diagnosis of CD of hyaline vascular type was rendered.

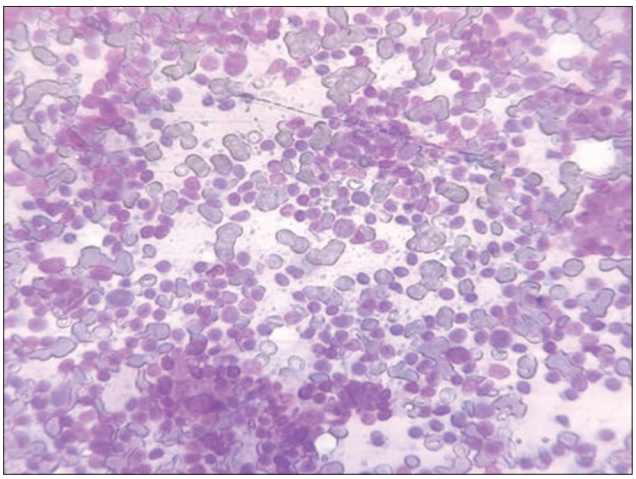

Figure 1: Giemsa stain, ×40, showing features of reactive lymphadenitis

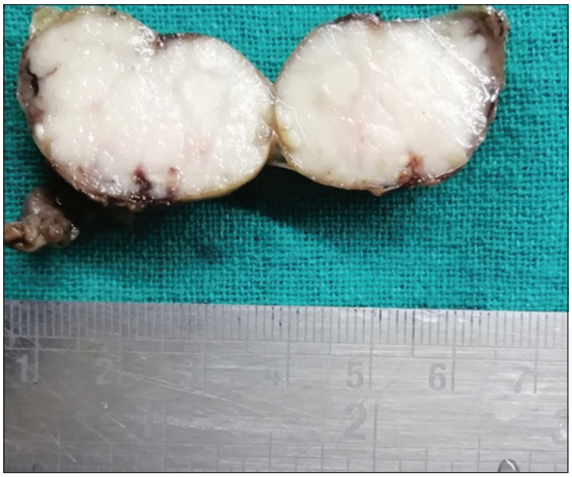

Figure 2: Gross examination of the lymph nodes

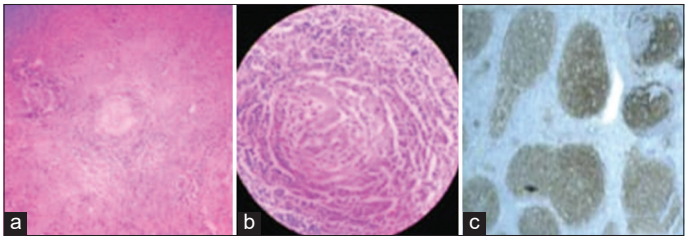

Figure 3: (a) H&E, ×20, showing lysis of follicles. (b) H&E, ×40, showing onion skin-like appearance. (c) IHC CD 21 positive in follicular dendritic cell

DISCUSSION

CD is a rare disease of lymph node of unknown etiology. In a few cases, infection with HIV and HHV 8 has been suggested [4]. Males and females are affected equally. Mostly, it affects the 4th and 5th decades of life. Younger people are more likely to have localized form and older to have a multicentric form. Unicentric is asymptomatic and affects the single lymph node and the gland slowly enlarges. Multicentric forms are symptomatic and affect more than one lymph node gland and other organs such as spleen, liver, and bone marrow.

Microscopically, it is of three types-hyaline vascular type, plasma cell type, and mixed type. The hyaline vascular type showed the proliferation of lymphoid follicles with hyalinized vessels in the center of the follicles along with the concentric layering of lymphocytes in the periphery (onion skin-like appearance). Marked vascular proliferation was seen in the interfollicular regions. Mitosis is absent.

Clinically, patients present with fever, anemia, fatigue, weight loss, pain at the site of swelling, increased ESR, and positive CRP. Interleukin 6 is found to be increased, maybe the cause of systemic symptoms [4]. In the present case, the patient did not have any symptoms except the neck swelling. Similar cases were reported by Melkundi *et al*. in an 18-year-old girl with neck swelling [5] and Reddy *et al*. in a 46-year-old lady [6]. Jiang *et al*. have described four cases of CD of the neck [7]. The two cases were unicentric hyaline vascular variant types, similar to the current case. A few cases have been described in the literature about the CD of cervical nodes.

It is very difficult to diagnose preoperatively. The cytology and radiology imaging findings are non-specific [8]. However, histopathological findings are characteristic. The differential diagnosis includes infection, lymphoma, autoimmune disorders, and metastasis. Proper evaluation of morphology, immunohistochemistry, and correlation with clinical findings helps in differentiation from other diseases. Atypical lymphoid cells are not seen in this. Lymphoma and metastasis can be ruled out due to the absence of atypical cells. In the case of infection, culture, sensitivity test, and special stains help in confirming the diagnosis.

The treatment depends on the type of CD. Complete surgical excision is the treatment of choice. It is the curative treatment in case of a unicentric or localized form. However, the patient should be followed regularly with radiological imaging to see the recurrence. The combination of surgery, steroid, and chemotherapy is used for multicentric form; however, there is no clear consensus on this Chronowski *et al*. [9].

CONCLUSION

CD is a rare disease and difficult to diagnose preoperatively. Localized CD has a good prognosis. Awareness of this entity helps in differentiating it from more aggressive lymphoma.

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