

## CASE REPORT

# CASTLEMAN'S DISEASE IN THE LONG TERM FOLLOW-UP OF A PATIENT WITH NON HODGKIN'S LYMPHOMA: AN UNUSUAL PRESENTATION

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## ABSTRACT

Castleman's disease is a benign condition characterized by localized or generalized lymphadenopathy. It is an inherited disorder and usually seen concurrently with lymphomas. We present here a case of multi-centric hyaline vascular type of Castleman's disease detected in the long term follow-up of a patient who was being previously treated for non Hodgkin's lymphoma. In the long term follow-up of patient with lymphomas it can be a cause of lymph node enlargement bearing a clinical resemblance to recurrence of lymphoma. The diagnosis should be made by a combination of clinical, radiological examination and histopathological examination with immunohistochemistry study.

**Keywords:** Castleman's disease, non Hodgkin's lymphoma, long term follow-up.

## INTRODUCTION

Castleman's disease (CD) was first described by Dr. Benjamin Castleman in 1956,<sup>1</sup> and it is a benign condition. CD is characterized by localized or generalized lymphadenopathy. Clinically CD can be uni-centric or multi-centric depending upon the localization of the lymphadenopathy, and histologically it can be hyaline vascular (HV), or plasmacytic or mixed cellularity type. CD is also known as angiofollicular hyperplasia of the lymph nodes, where there is a follicular hyperplasia of lymph nodes with abnormally increased inter-follicular vascularity. <sup>2</sup>CD is an inherited disorder, but it also arises concurrently in lymphomas.<sup>3</sup> We present here a case of multi-centric CD detected in the long term follow-up of a patient who was being previously treated for non Hodgkin's lymphoma (NHL).

## CASE REPORT

A 54-year-old male who was a known and treated case of NHL 8 years back, presented at the head and neck surgery OPD with the complaints of recurrence of swelling on both the sides of the neck along with a swelling on the right side of the axilla of 3 months duration. The swellings were gradually growing in nature and were not associated with pain. There was no associated history of pharyngeal and/or laryngeal symptoms. On examination of the neck, there were multiple non tender, firm, mobile, and discrete swellings on both sides of the neck all measuring less than 3cm in size. The axillary swelling was firm, non tender, mobile and was of 4cm X 6cm in its maximum dimensions. Clinically

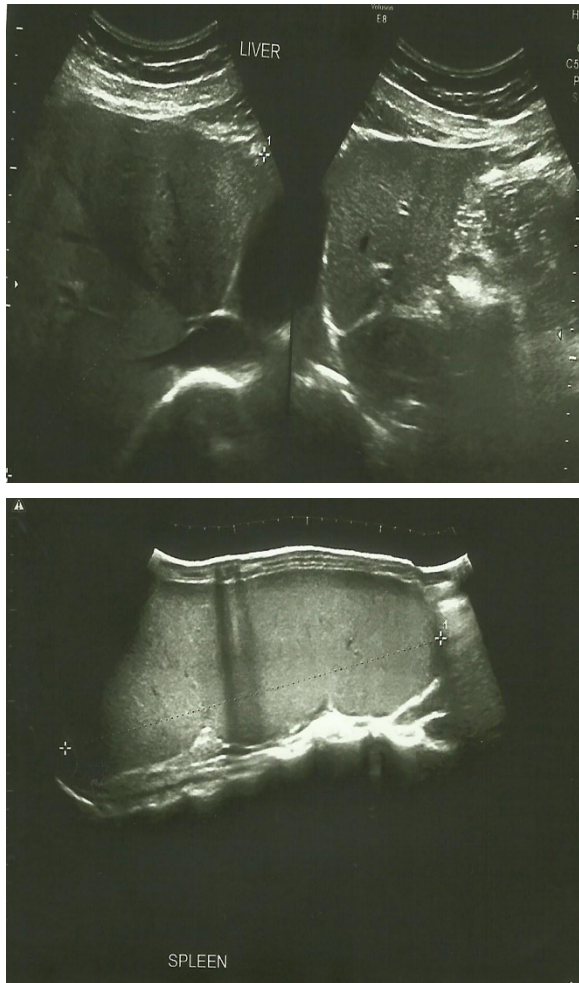
the swellings appeared to be multiple enlarged lymph nodes of the neck and axilla.

Ultrasonography (USG) of the abdomen revealed moderate hepatomegaly with splenomegaly (Figures 1a & b). Excision biopsy of a cervical lymph node showed morphological features suggestive of hyaline vascular type of CD (Figures 2 a & b). Immunohistochemistry (IHC) was done for Bcl2, CD45, CD30, CD21 and CD35. IHC was positive for the expression of CD21 (Figure 3). The diagnosis of Castleman's disease was confirmed after clinical, radiological, and histopathological examination supported by IHC. Serological test for human immunodeficiency virus was negative in this patient.

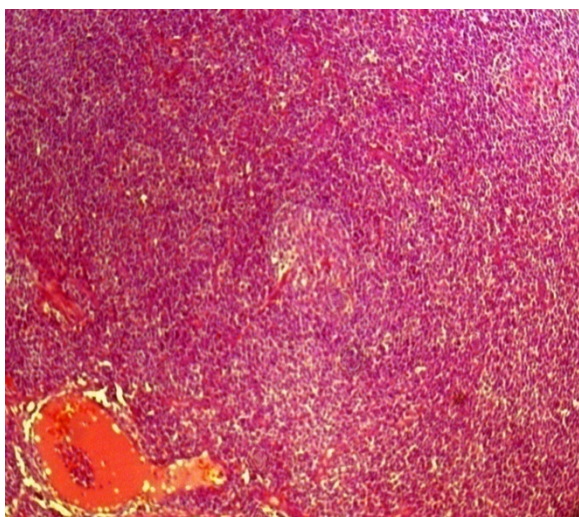
The patient was treated with systemic corticosteroids. There was near total remission of the cervical and axillary lymphadenopathy at the three month post treatment follow-up.

## DISCUSSION

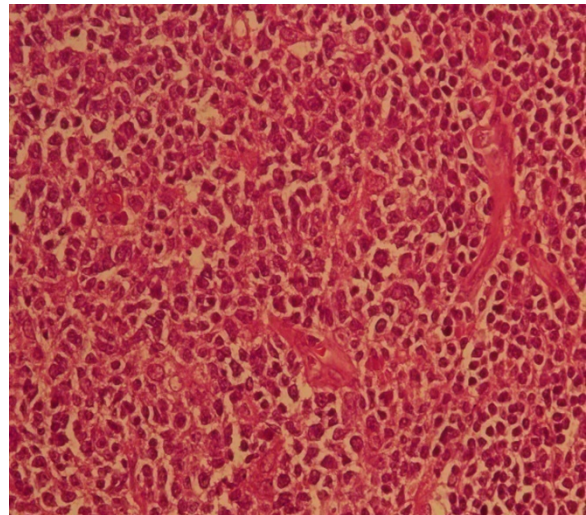
The prevalence of CD in lymphomas is not precisely known due to the rarity of this entity. Uni-centric CD is seen in young or in 3<sup>rd</sup> decades of life and multi-centric CD is usually seen in the elderly or in sixth decade of life with systemic manifestations.<sup>3</sup> Our patient was a case of multi-centric CD due to the involvement of cervical, axillary and there was associated hepato-splenomegaly, and this case of multi-centric CD was seen in 54 years old male patient without systemic symptoms. CD's are mostly seen concurrently with Hodgkin's lymphomas, non Hodgkin's lymphomas, and Kaposi's sarcoma.<sup>3-5</sup>



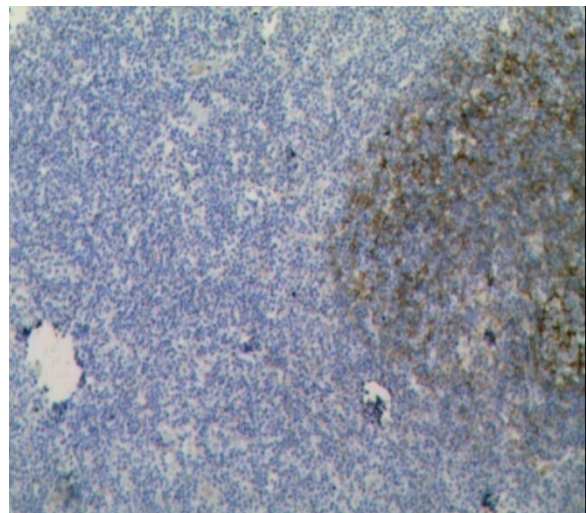
**Figure 1: Photograph of ultrasonography showing (a) hepatomegaly in both the lobes of liver (upper one), (b) splenomegaly (Lower one).**



**Figure 2:(a) Photomicrograph with hematoxylin and eosin stain (10X) showing small compressed follicles surrounded by mantle zone of lymphocytes and hyaline blood vessels, one of them penetrating the follicle with absent sinuses**



**Fig 2(b) Photomicrograph with hematoxylin and eosin stain (40X) polymorphic cellular infiltrate and prominent vessels with hyalinized wall**



**Figure 3: Photomicrograph showing the expression of CD21 on immunohistochemistry**

However, in our case the CD occurred following non Hodgkin's lymphoma after a long period of remission of 8 years, which was very unusual and not reported before this case. Our case was pathologically a hyaline vascular type (HV), and there was CD21 positivity which highlighted the follicular dendritic cells. The pathogenesis of HV changes in CD and the causes of the prominent HV-CD-like changes associated with lymphomas are not well understood.<sup>6</sup>The treatment for CD depends upon the localized (uni-centric) or generalized disease (multi-centric). It ranges from surgery for uni-centric disease to chemotherapy, immune-therapy and corticosteroids in multi-centric disease.<sup>7</sup> In the present case as it was a multi-centric HV-CD, the patient was treated with systemic corticosteroids only and the response to treatment was good.

## CONCLUSION

CD is a rare condition and in patients with lymphomas it presents concurrently. However, in the long term follow-up of patient with NHL it can be a cause of lymph node enlargement bearing a clinical resemblance to recurrence of lymphomas. The diagnosis of CD should be made by a combination of clinical, radiological examination and HPE with IHC study.

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