



## CASE REPORT: RARE CASE OF CLASSICAL HODGKIN'S LYMPHOMA, MIXED CELLULARITY IN A MALE BOY AGED 12 YEARS.

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**ABSTRACT...** Castleman disease was initially explained in 1956 as a syndrome that is lymphoproliferative, also recognized as angiofollicular lymph node hyperplasia. It is a rare disease and may proceed to malignant lymphoma that is difficult to diagnose while the treatment is complex. Hodgkin lymphoma (HL) is a malignancy that is related to lymphoid and can be characterized as Reed-Sternberg cells in the lymph nodes. Prognosis as well as treatment is complex and depends upon the Ann Arbor staging as well as age of the affected patient. HL is divided into 2 distinct subtypes, Nodular Lymphocyte Predominant HL (LPHL), and classical HL (cHL). The present case was a 12 year old male boy who presented with fever, cough and swelling on the body at multiple sites. Diagnosis of cervical lymph node biopsy was labeled as classical Hodgkin's Lymphoma, mixed cellularity. Early diagnosis, better understanding of the pathophysiology as well as the biology of HL can assure much better prognosis.

**Key words:** Castleman, Hodgkin Lymphoma (HL), Mixed Cellularity.

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## BACKGROUND

Castleman disease was initially explained in 1956 as a syndrome that is lymphoproliferative, also recognized as angiofollicular lymph node hyperplasia. It is a rare disease with an incidence of about 25 cases / million person annually.<sup>1</sup> Castleman disease can be classified in subtypes as unicentric or multicentric. Castleman disease may proceed or can be linked to malignant lymphoma that is difficult to diagnose while the treatment is complex.<sup>2</sup>

Hodgkin lymphoma (HL) is a malignancy that is related to lymphoid and can be characterized as Reed-Sternberg cells in the lymph nodes. Prognosis as well as treatment is complex and depends upon the Ann Arbor staging as well as age of the affected patient.<sup>3</sup> HL involves 12.1 / million individuals aged less than 20 years, encompassing around 9% of cases having cancer in this specific age group.<sup>4</sup> HL was 1<sup>st</sup> described in 1832 and since then it has been classified into various subgroups.<sup>5</sup> Several risk factors depending upon the pathology and prognostic

features are documented. The etiology is still not fully understood but factors like Epstein-Barr Virus (EBV), human herpes virus 6 (HHV6), cytomegalovirus (CMV), hepatitis C virus (HCV) and human immunodeficiency virus (HIV) are said to implicate in the genesis.<sup>6</sup>

WHO divided HL into 2 distinct subtypes, Nodular Lymphocyte Predominant HL (LPHL), 5% of all HL cases and classical HL (cHL) as 95% of all HL cases. cHL is further separated as nodular sclerosis (NSHL), 70% cases, mixed cellularity (MC), 20% of cases and lymphocyte depleted HL as 5% of all HL cases.<sup>7</sup>

## CASE PRESENTATION

The present case was a 12 years old male boy who presented at department of Oncology, Children's Hospital and The Institute of Child Health, Multan, with fever, cough and swelling on the body at multiple sites. The patient started noticing multiple swelling on the neck and profuse sweating about 19 days back. There was associated pallor for that packed cells were transfused. The patient received

multiple treatments but to no avail. Fine needle aspiration cytology (FNAC) of left cervical lymph node revealed scanty lymphoid cells, suboptimal for definitive diagnosis. Histological examination of the sections revealed effaced architecture of the lymph nodes. There was seen polymorphic lymphoid population, hyalinizing proliferating blood vessels and extensive fibrosis. Picture was suggestive of hyaline vascular castlemans disease however the pathologist advised immunochemistry for confirmative diagnosis to rule out lymphoma. Anti HCV and HBsAg were found negative. Typhidot IgG and typhidot IgM were found negative. Other lab reports regarding liver functions, urinary functions and electrolytes were found normal.

CT neck had evidence of enlarged lymph nodes in left submandibular region, level II, level III, level IV, level V, level VI and level VII. Few of them were calcified in supra-clavicular levels. CT Chest showed enlarged lymph nodes noted in pre-vascular region and along internal mammary artery on right side. Subcentimal lymph nodes were seen in bilateral axillary regions. Patch of consolidation with air bronchograms seen in apical segment of the left lung with adjacent bronchiectatic changes. Another patch of consolidation / collapse noted in posterior basal segment of right lower lobe with pleural thickening and pleural effusion on right side. CT abdomen and pelvis showed hepatosplenomegaly with lymphadenopathy, ascites and lymphomatous deposits in liver, spleen and vertebral bodies.

Cervical lymph node biopsy was ordered. Two friable grayish tissue fragments collectively measuring 1.5 cm x 1.0 cm was examined. Histological examination disclosed lymph node showing effacement of nodal architecture by a lymphoid neoplasm composed of scattered large mono and binucleated tumor cells having vesicular nuclei and prominent eosinophilic nucleoli. These large cells were present in a mix inflammatory background comprising of lymphocytes, plasma cells, histocytes and eosinophils. Immuno/Histochemical Stains revealed; CD30: positive in large atypical cells, CD15: positive in rare large atypical cells, PAX5:

weakly positive in large atypical cells, CD20: negative in large atypical cells, CD: negative in large atypical cells, LCA: negative in large atypical cells. Diagnosis of cervical lymph node biopsy was labeled as classical Hodgkin's Lymphoma, mixed cellularity.

The patient was then put on COPD (Cyclophosphamide, Oncovin, Prednisolone, Dacarbazine) therapy where he started recovery following 8 days of treatment.

## DISCUSSION

Classical Hodgkin's Lymphoma of mixed cellularity is a rare disease specially in children. HL usually presents with non-tender enlarged lymph nodes which are usually asymmetric, painless and non-adherent to the skin.<sup>8</sup> Rubbery consistency is associated whereas the neck and supraclavicular region is featured in about 75% cases as was seen in the present case. HL is found to spread through lymphatic system. Frequent symptoms found are fever, night sweats and weight loss. We also found fever and profuse sweating in our patient. Other symptoms include cough, pain in the chest area, shortness of breath, abdominal pain and ascites.<sup>9</sup> Biopsy is noted to be the best method for the confirmatory diagnosis. Fine needle aspiration is not usually indicative of diagnosis that is why excisional biopsy is the most authentic tool. Lymph node examination is usually evident of fibrosis of the tissue architecture.<sup>4</sup> Early diagnosis, better understanding of the pathophysiology as well as the biology of HL can assure much better prognosis.

## CONCLUSION

Classical Hodgkin's Lymphoma, mixed cellularity was diagnosed by cervical lymph node biopsy.

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