Abstract: Castleman's disease (CD) is regarded as a polyclonal lymphoid proliferation of unknown etiology with synonyms of giant node hyperplasia, angiomatous lymphoid hamartoma, angiofollicular lymph node hyperplasia and follicular lymphoreticuloma. CD may occur anywhere along the lymphatic system with the majority of the cases seen in the mediastinum (70). Extrathoracic sites have been reported in the axilla, neck, pelvis, retroperitoneum, mesentery, central nervous system, and orbit. Here we report a case of unicentric castleman's disease mimicking intrapulmonary malignancy.

Keyword: castleman's disease, mediastinum, HIV, HHV-8

Case report: A 23 year old female came with history of cough and hemoptysis for one month. There was no history of fever/weight loss/easy fatigue/night sweats. On general examination there was no anemia/generalized lymphadenopathy/jaundice/clubbing. Patient had pulse rate of 80/mt, BP-120/80 mm/hg, TC-7100 cells/cumm DC- P59% L 39% E 2%, ESR-18mm/hr, Hb-11.8gm%, Blood sugar- 96mg %, urea-28mg% sr creatinine-0.8.

CT chest revealed well defined intensely enhancing mass lesion of size 10x6x7 cm involving the anterior mediastinum with no mediastinal lymphadenopathy, pleural and pericardial effusion. USG abd and pelvis was Normal. Sternotomy and excision of the mass with anterolateral thoracotomy extension was done. Per operatively an encapsulated mass of size 10x6x7 cm involving the anterior mediastinum with involvement of Rt phrenic nerve and brachiocephalic trunk was seen. Histopathology came as Hyaline vascular type of unicentric castleman's disease. Post op period was uneventful. Pathology:

On gross examination, a sharply demarcated mass lesion embedded in hemorrhagic soft-tissue with a yellow-brown cut surface and areas of calcification was seen. Sections showed a tumor consisting of lymphoid tissue with abnormal germinal center. The follicle is surrounded by a broad mantle zone consisting of a concentric layering of lymphocytes resulting in an onion-skin appearance. The follicles are frequently penetrated radially by a sclerotic blood vessel resembling a "lollipop". So the diagnosis of CD of hyaline vascular type was established. The patient had an uneventful postoperative course and is doing well after 1 year of follow-up.
GROSS SPECIMEN

HISTOPATHOLOGY
Discussion:
Castleman in 1954 first described this disease in a series of 13 patients and later on in 1956 eloquently bettered the definition as a "localized mediastinal lymph node hyperplasia resembling thymoma." The disease is more frequent in women with a median age at diagnosis in the 3rd or 4th decade. It can be associated with Kaposi's sarcoma, non-Hodgkin's lymphoma, Hodgkin's lymphoma and POEMS syndrome. Multicentric form usually affects older individuals. 70% of Castleman's disease cases are located in the thorax 10–15% in the abdomen, retroperitoneum, and pelvis and 10–15% in the neck. Castleman's disease has also been described in the extralymphatic tissues, including the lung, larynx, parotid gland, pancreas, and muscle. Flendrig in 1970 distinguished two basic pathologic types and one mixed variant. Based on these features, Keller et al. in 1972 sub-classified the disease as hyaline-vascular (HV), plasma cell (PC) and HVPC types or mixed type. CD can be clinically classified into unicentric and a multicentric disease with the former occurring in about 90% of cases, majority being of the Hyaline vascular type. The localized form usually has a benign course and clinical abnormalities frequently resolve after excision of the affected lymph nodes. Multicentric type is usually of the Plasma cell variant and patients present with a systemic illness with a potential for developing into malignancy of the non-Hodgkin's lymphoma in the absence of human immunodeficiency virus. In the localized form of plasma-cell type Hodgkin's lymphoma may occur. Prevalence is estimated to be less than 1/100,000. It has been postulated that the disease represents a reaction to chronic viral antigenic stimulation most probably to interleukin-6 in the disease associated with systemic manifestations. Human herpes virus 8 HHV-8 has been implicated as the probable virus capable of causing disease.

Conclusion:
Unicentric castleman's disease most commonly occurs in thorax (70%) . This case presenting with hemoptysis mimicked an intrapulmonary malignancy. Complete excision of the tumor is curative and the patient is under follow up for past 1 year.

References: