ORIGINAL RESEARCH PAPER
Volume - 9 | Issue - 8 | August - 2020 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

RIGHT SIDED PNEUMOTHORAX IN A KNOWN CASE OF CASTLEMAN'S DISEASE WITH THYMOMA WITH LICHEN PLANUS:- A RARE CASE REPORT

Pulmonary Medicine

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ABSTRACT

Castleman's Disease (CD, angiofollicular lymphnode hyperplasia) is a rare lymphoproliferative disorder, while it has been associated with several disorders,there are only rare reports of lung involvement in castleman's disease.Here in We report A 32 years old female patient,who presented with difficuly in breathing since 2 months and weight loss as initial symptoms.The diagnosis was confirmed by CT guided biopsy of lymph node. Her respiratory symptoms was improved by steroids.

KEYWORDS

Castleman's disease, pneumothorax, lichen planus, thymoma

INTRODUCTION

Castleman's disease(CD) is a rare lymphoproliferative disorder that can involve enlargement of one(unicentric) OR multiple(multicentric) lymphnode. The mediastinum is most common location of unicentric disease and lesion most frequently occur in anterior and middle compartment. Human herpes virus 8(HHV8) infection has been implicated in the pathogenesis of multicentric disease but not in the unicentric disease, notably patient with castleman's disease respond well to steroids.

CASE REPORT:

A-32-year-old hindu female patient was admitted to our hospital for gradual onset of breathlessness with weight loss, dry cough, fatigue and night sweat since 2months with no complaint of fever,chest pain and pedal edema.she had no past history of hypertension, diabetes, tuberculosis and she was not having any addiction. She didn't had similar illness in her family. She was a known case of lichen planus since 2018 and was taking tab. Wysolone and tab. Mycophenolate mofetil on regular basis.

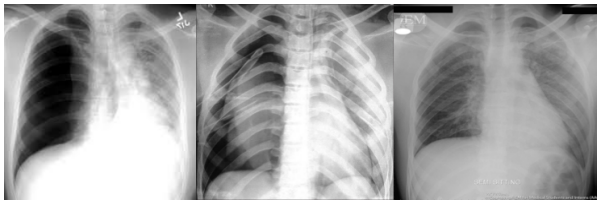
At the time of admission general condition was fair,afebrile,vitaly stable,temperature was 36.8°C, pulse was 98/min, respiratory rate was 22/min, blood pressure was 110/70mm Hg, Spo2 70 on room air. There was no cyanosis, clubbing or distension of jugular vein.

Trachea was pushed towards left side, there was increase in respiratory movements. On auscultation, there decrease air entry on right side.

ABGA showed PaO_2 56,PCO_2 36, pH 7.36,$Sp0_2$ 70%.

CBC,RFT LFT,PT/INR,RBS were within normal limits. sputum for AFB was negative.

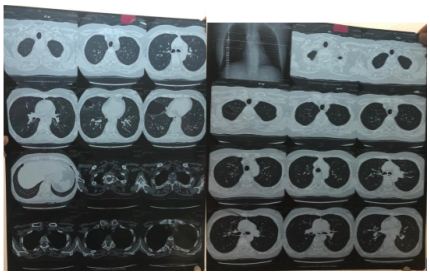
Chest Radiography PA view.



Right sided pneumothorax
Post icd insertion Xray
Post icd removal Xray

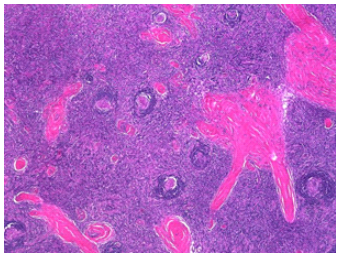
Pneumothorax was resolved after putting icd and kept for 3 days. Later on after stabilizing the patient CT scan was done which shows:-

Large homogenously enhancing hypodence lesion measuring 6.9×6.7×11.3cm seen in prevascular location extending from left supraclavicular region superiorly upto the main pulmonary artery inferiorly.Fibrocystic and bronchiectic changes in right middle lobe with elongated lobulated bulla in lateral segment of right middle lobe extending into the right middle fissure.P/O- Neoplastic etiology such as Lymphoma/Thymoma. Adv: CT guided biopsy.



Thyroid scan:- s/o thyroiditis

CT guided biopsy done which shows left sided invasive Thymoma, for that Open Thymectomy was done by surgical consultant.Tissue sample sended for HPE which shows " Castleman's disease of Hyaline vascular type" which was managed conservatively by steroids and discharged and was followed after one month.



Castleman's disease "hyaline vascular type".

DISCUSSION

Castleman's disease describes a group of 3 immunological disorder that occur in individual of all ages and share a similar microscopic lymph node appearance.

There is significant variability in clinical feature, treatment and survival across the 3 subtypes:

1)UCD. Involves a single region of enlarge lymph nodes
2)HHV-8 associated MCD. Involves multiple regions of enlarge lymph nodes

3)HHV-8 negative/idiopathic MCD. Involves multiple region of enlarge lymph nodes due to unknown cause.

Etiology: of UCD is unknown but some cases shows somatic mutations in monoclonal cell populations likely lymph node stromal cells. Active HHV-8 infection is associated with MCD.

Epidemiology: UCD more common in women and younger individual. HHV-8 associated MCD is more common in men with individual associated with HIV.

Dr. Benjamin Castleman first described the constellation of lymph node features observed in 1950's.

Clinical features: includes flu like symptoms(fever,night sweats,weight loss,weakness),shortness of breath,numbness,cough, leg edema,skin rashes.

The preferred diagnostic method of CD is "Lymph node biopsy".

Lymph node features of different histopatho subtypes of UCD are as follows:

1)Hyaline vascular 2)Plasma cells 3)Mixed

Differential Diagnosis:
1)UCD-Rheumatoid arthritis, Plasmacytoma, Toxoplasma lymphadenitis, NHL with nodular growth pattern
2)HHV-8 neg MCD- HIV lymphadenitis, Kaposi sarcoma

Treatment:-
UCD-Surgical removal of enlarged lymph node is the gold standerd. Patients with unresectable disease can be treated by immunomodulator & chemotherapy. Systemic steroids can provide symptomatic relief but don't reduce tumor size.

HHV ass MCD- Rituximab is highly effective
HHV neg MCD- Siltuximab

Prognosis:-
UCD- Excellent, cured after lymph node excision
HHV-8 ass MCD- Good prognosis, when treated with rituximab
HHV neg MCD- Bad prognosis

CONCLUSION

We described a patient with CD who presented with anterior mediastinal mass causing respiratory symptoms. Lung involvement is rare. We confirmed diagnosis by Open biopsy of mediastinal mass which shows highly vascular type of UCD of Castleman's disease.

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