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Castleman's Disease in a 35 Year-Old Woman with a Giant Mediastinal Mass

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ABSTRACT

The patient was a 35 year-old woman with a 10-month history of breathing difficulty, cough and chest pain. An anterior-posterior chest radiograph revealed opacification of the right hemithorax with reduced right lung volumes. As part of diagnostic evaluation, a computed tomography (CT) scan was performed which showed a huge mass in the right anterior mediastinum with extension to the right side of the pleural space. It was resected through postero-lateral thoracotomy .The patient was discharged in a good condition. The pathology report showed hyaline vascular variant of Castleman,s disease.

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Key words: *Castleman's Disease, Mediastinum, Mediastinal mass*

INTRODUCTION

Castleman's disease(CD) was first described in 1956 by Benjamin Castleman, who identified a series of patients with solitary hyperplastic mediastinal lymph nodes (1). These lymph nodes showed a prominent proliferation of small and hyalinized follicles [hyaline vascular variant or unicentric form of Castleman's disease (UCD)]. The same investigators later identified a similar lesion in lymph nodes with more hyperplastic germinal centers and sheets of plasma cells in the interfollicular regions [plasma cell variant or multicentric form (MCD)] (2-4). Approximately 10 to 20 percent of all cases were of the plasma cell variant(MCD) (3). MCD is usually the plasma cell variant and is a systemic disease with generalized peripheral lymphadenopathy, hepatosplenomegaly, recurrent fever, and night sweats (5). We report a 35 year-old woman with a 10-month history of dyspnea, cough and chest pain and a huge right anterior mediastinal mass which in the pathology report was described as the hyaline vascular variant of Castleman's disease. (UCD).

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CASE SUMMARIES

Our patient was a 35-year-old woman with a 10-month history of breathing difficulty, cough and chest pain. Her dyspnea worsened during the previous week and she admitted to the hospital. She was non-smoker and had unknown occupational exposures. An anterior-posterior chest radiograph

revealed an almost complete opacification of the right hemithorax with reduced right lung volumes (Figure 1).

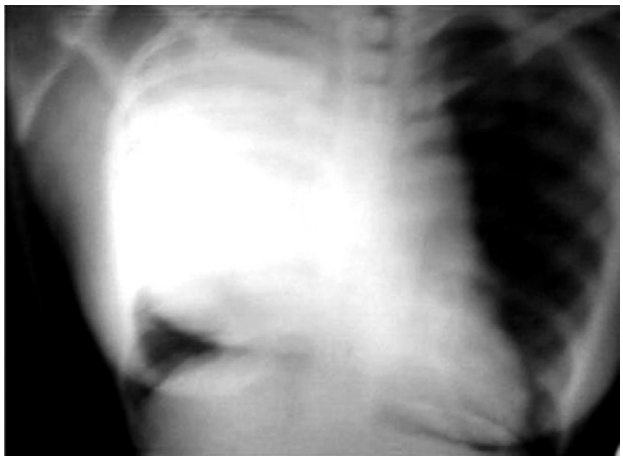

Figure 1. CXR: An almost complete opacification of the right hemithorax.

Pulmonary function tests showed a decrease in forced vital capacity (57% predicted) and the forced expiratory volume (52% predicted) with a total lung capacity of 72%. These findings were interpreted as being consistent with mixed obstructive and restrictive lung disease. As part of the diagnostic evaluation, a CT-scan was performed which revealed a huge mass in the right anterior mediastinum overlying the heart with extension into the right hemithorax (Figures 2).

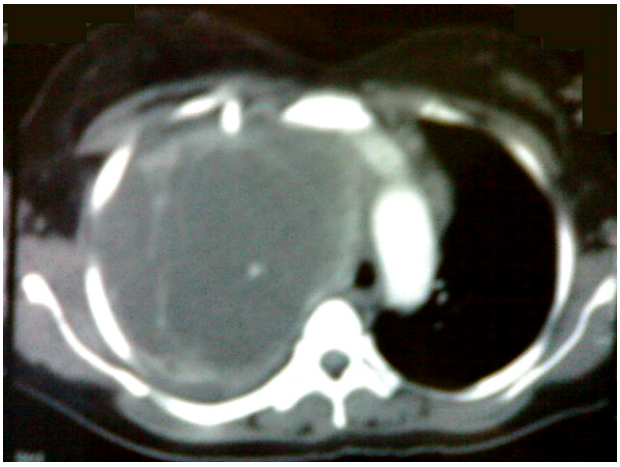

Figure 2. CT- scan: A huge right anterior mediastinal mass extending into the right hemithorax.

During the right side postero-lateral thoracotomy, we found a large encapsulated, vaguely lobulated mass in the right anterior mediastinum entering the right pleural cavity and occupying approximately 80% of the right pleural space. The mass was resected completely weighing 1,200g. (Figures 3 and 4).

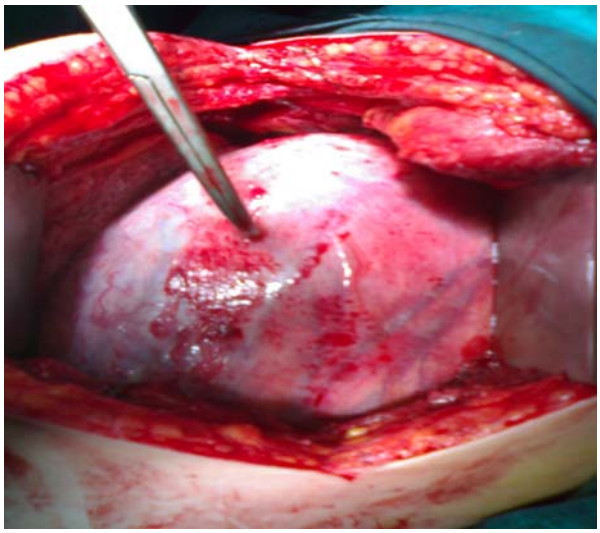

Figure 3. Huge mass found during postero-lateral thoracotomy

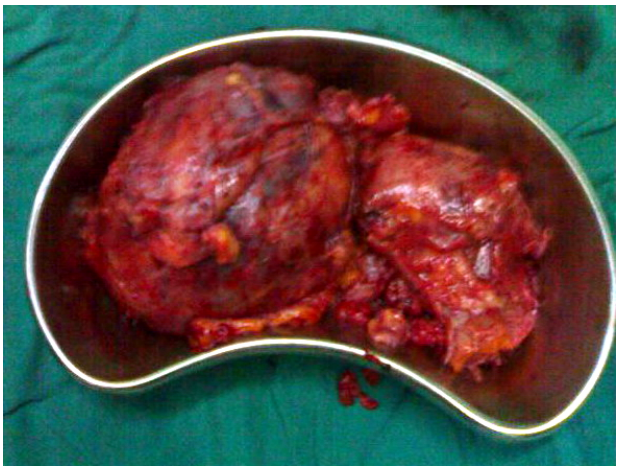

Figure 4. Huge mass after complete resection.

Histopathological examination revealed the hyaline vascular variant characterized by a marked increase of abnormal follicles with atrophic or "regressed" germinal centers and broad mantle zones

of small lymphocytes, often with an "onion-skin" arrangement around the germinal centers. Often multiple regressed germinal centers are present within the same follicle. The patient was discharged in a good condition on day 6. The patient is well 8 months after the surgical excision.

DISCUSSION

Castleman's disease or angiofollicular lymph node hyperplasia is a lymphoproliferative disorder that has attracted attention because of its association with the human immunodeficiency virus (HIV) and human herpes virus 8 (HHV-8). Castleman's disease comprises at least two distinct diseases with very different prognoses. It is associated with a number of malignancies, including Kaposi's sarcoma (KS), non-Hodgkin's lymphoma, Hodgkin's lymphoma, and POEMS syndrome (2-4).

Castleman's disease was described in 1956 by Benjamin Castleman, who identified a series of patients with solitary hyperplastic mediastinal lymph nodes with small germinal centers resembling Hassall's corpuscles of the thymus (1).

All cases described in these studies had localized form of disease, which is now termed unicentric Castleman's disease (UCD); although a subset of patients with the plasma cell variant did have systemic symptoms. The multicentric form of Castleman's disease (MCD) was recognized in 1978 (4-7). Our knowledge regarding MCD has greatly increased since the identification of its association with HIV and HHV-8 infection (8-10).

Unicentric Castleman's disease (UCD) is most often an isolated benign lymphoproliferative disorder of young adults, and is generally curable with surgical resection. The largest published series included 81 cases; approximately 90% of which were of the hyaline vascular variant (3). The vast majority of patients were asymptomatic and their disease was identified incidentally on imaging studies. The mean age was 35, with equal male/female ratio. The mean size of the lesion was 5 to 9 cm; 70% of lesions were located in the mediastinum or hilum of lung, with the abdomen being the next most frequent site. UCD should be differentiated from other huge mediastinal masses such as non-Hodgkin's lymphoma, Hodgkin's lymphoma, neuroblastoma, ganglioneuroblastomas, thymomas, germ cell tumors and Askin's tumor (2,15). Complete resection of the involved node in either type of unicentric or multicentric CD is curative, with no reported recurrences in any series. Systemic symptoms, if present, will also resolve (3,11-14). Our case was symptomatic (breathing difficulty, cough and chest pain). The size of mass was 30x18 cm. It was the unicentric form (UCD) and totally resectable. All symptoms resolved after resection of the mass.

REFERENCES

1. Castelman B, Iverson L, Menendez VP. Localized mediastinal lymphnode hyperplasia resembling thymoma. ***Cancer***.1956; 9 (4): 822- 30.
2. Flendrig JA, Schillings PHM. Benign giant lymphoma: the clinical signs and symptoms. ***Folia Med Neerl*** 1969; 12: 119.
3. Keller AR, Hochholzer L, Castleman B. Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and other locations. ***Cancer*** 1972; 29 (3): 670- 83.
4. Gaba AR, Stein RS, Sweet DL, Variakojis D. Multicentric giant lymph node hyperplasia. ***Am J Clin Pathol*** 1978; 69 (1): 86- 90.
5. Ocio EM, Sanchez-Guijo FM, Diez-Campelo M, Castilla C, Blanco OJ, Caballero D, San Miguel JF. Efficacy of rituximab in an aggressive form of multicentric Castleman disease associated with immune phenomena. ***Am J Hematol*** 2005; 78 (4): 302- 5.
6. Ide M, Kawachi Y, Izumi Y, Kasagi K, Ogino T. Long-term remission in HIV-negative patients with multicentric

Castleman's disease using rituximab. ***Eur J Haematol*** 2006; 76 (2): 119- 23.

7. Casquero A, Barroso A, Fernández Guerrero ML, Górgolas M. Use of rituximab as a salvage therapy for HIV-associated multicentric Castleman disease. ***Ann Hematol*** 2006; 85 (3): 185- 7.
8. Peterson BA, Frizzera G. Multicentric Castleman's disease. ***Semin Oncol*** 1993; 20 (6): 636- 47.
9. Soulier J, Grollet L, Oksenhendler E, Cacoub P, Cazals-Hatem D, Babinet P, d'Agay MF, Clauvel JP, Raphael M, Degos L, et al. Kaposi's sarcoma-associated herpesvirus-like DNA sequences in multicentric Castleman's disease. ***Blood*** 1995; 86 (4): 1276- 80.
10. Yamasaki S, Iino T, Nakamura M, Henzan H, Ohshima K, Kikuchi M, Otsuka T, Harada M. Detection of human herpesvirus-8 in peripheral blood mononuclear cells from adult Japanese patients with multicentric Castleman's disease. ***Br J Haematol*** 2003; 120 (3): 471- 7.
11. Dupin N, Diss TL, Kellam P, Tulliez M, Du MQ, Sicard D, Weiss RA, Isaacson PG, Boshoff C. HHV-8 is associated with a plasmablastic variant of Castleman disease that is linked to HHV-8-positive plasmablastic lymphoma. ***Blood*** 2000; 95 (4): 1406- 12.
12. Parravicini C, Corbellino M, Paulli M, Magrini U, Lazzarino M, Moore PS, Chang Y. Expression of a virus-derived cytokine, KSHV vIL-6, in HIV-seronegative Castleman's disease. ***Am J Pathol*** 1997; 151 (6): 1517-22.
13. Bowne WB, Lewis JJ, Filippa DA, Niesvizky R, Brooks AD, Burt ME, Brennan MF. The management of unicentric and multicentric Castleman's disease: a report of 16 cases and a review of the literature. ***Cancer*** 1999; 85 (3): 706- 17.
14. Herrada J, Cabanillas F, Rice L, Manning J, Pugh W. The clinical behavior of localized and multicentric Castleman disease. ***Ann Intern Med*** 1998; 128 (8): 657- 62.
15. Maslovsky I, Uriev L, Lugassy G. The heterogeneity of Castleman disease: report of five cases and review of the literature. ***Am J Med Sci*** 2000; 320 (4): 292- 5.