

Castleman's disease. Report of a case and review of literature

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ABSTRACT

We report here a case of solitary form of Castleman's disease (hyaline-vascular type) with its unusual location in the neck. The case is being reported for its rarity.

Keywords: Castleman's disease; lymphadenopathy.

INTRODUCTION

Castleman's disease (giant lymph node hyperplasia) represents a morphologically distinct form of lymph node-hyperplasia rather than a neoplasm or a hamartoma. It is a rare cause of lymphadenopathy. Since its initial description by Castleman and coworkers in 1956, many papers have been published on this subject reporting new cases and redefining the syndrome. They in their original report described thirteen cases of large, benign, localised, asymptomatic mass of mediastinal lymph nodes which resembled thymomas both clinically and histologically.¹ In subsequent observations, the entity also

included extramediastinal lymph nodes and spleen.^{2,3} This lesion was further expanded by the description of two histologic variants² and by inclusion of multicentric cases.³⁻⁵ Now it has been regarded as a benign systemic lymphoproliferative disorder.⁴⁻⁶ The two histologic variants of Castleman's disease are hyaline-vascular type and plasma-cell type. Clinically it may present as a solitary or multicentric forms.

CASE REPORT

A 20-year-old male patient presented with the right side neck swelling for 4 years. The

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patient noticed a small, solitary, painless swelling on the right cervical region, gradually increasing in size. He had been treated with various antibiotics and a full course of antituberculous therapy at Biratnagar Zonal hospital but with no improvement. He was referred to B.P. Koirala Institute of Health Sciences, Dharan with a provisional diagnosis of lymphoma. He gave history of cough on and off with minimal expectoration. There was no history of dyspnoea, haemoptysis, fever, weight loss or anorexia.

General physical examination was unremarkable. There was a solitary painless right cervical swelling measuring 7x4x3 cm, firm in consistency, freely mobile and not fixed to underlying soft tissue. Other systems were normal.

Laboratory investigations revealed the following: haemoglobin 13.7 g/dl, total leucocyte count 8,900/mm³ with a differential count of 40% neutrophils, 38% eosinophils and 22% lymphocytes; ESR (Westergren) 12 mm/1st hour; platelet count 1,80,000/mm³. Stool examination showed eggs of *Ascaris lumbricoides*. Total serum protein and serum albumin were 7.2 g/dl and 4.3 g/dl respectively. Chest X-ray was normal. Ultrasound examination of the abdomen and pelvis was normal. Fine-needle aspiration (FNA) of the right cervical swelling revealed polymorphous population of mature lymphocytes, immunoblasts and occasional plasma cells and multiple fragments of vessels in the background.

The mass was excised under general anaesthesia and sent for histopathological

examination. On gross examination, the mass was well encapsulated measuring 7.0x3.5x3.0 cm in size with the outer surface being smooth. Cut surface was greyish white and homogeneous with no areas of haemorrhage or necrosis. Histopathology revealed altered nodal architecture with increased number of lymphoid follicles present throughout the parenchyma. The follicles showed marked vascular proliferation and hyalinization of germinal centres (Fig. 1).

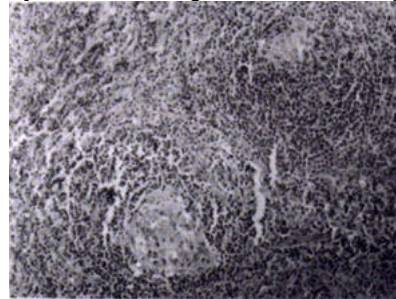


Fig. 1: Follicular hyperplasia with small, involuted, and partly collagenized germinal centres and marked vascular proliferation in the interfollicular region (H & E stain; 250 X).

There were multiple concentric layering of lymphocytes in onion-skin fashion surrounding the germinal centres (Fig. 2).

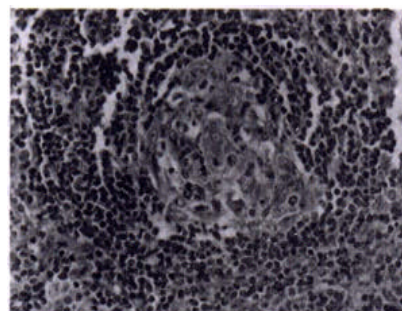
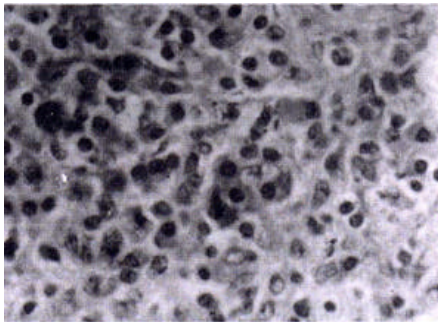


Fig. 2: Lymphoid follicle with collagenized germinal centre and concentric layers of small uniform lymphocytes (H & E stain; 500 X).

The interfollicular stroma was also prominent, with numerous hyperplastic vessels of the post-capillary-venule type with admixture of plasma cells, immunoblasts and eosinophils (Fig. 3). The diagnosis of hyaline-vascular type of angiofollicular lymph node hyperplasia (Castleman's disease) was made. For the parasitic infestation the patient was given Tab. Albendazole 400 mg orally on the first day and repeated on the 7th day. The peripheral eosinophilia came down to normal



level.

Fig. 3: Interfollicular region showing infiltration by plasma cells and immunoblasts (H & E stain: 1000 X).

DISCUSSION

Among the two histologic variants of Castleman's disease, hyaline-vascular type is more common. Most lesions are mediastinal and asymptomatic; often discovered on routine chest radiographic examination. In few cases, symptoms of tracheobronchial compression maybe seen. A wide spectrum

of ages have been reported, varying from childhood to the seventh decade and no predilection for either sex. On histopathology, the lesion maybe mistaken for follicular hyperplasia of reactive lymphadenopathy or a follicular lymphoma. However, paucity of true germinal centres with tingible-body macrophages excludes the former and absence of evenly distributed nodules of atypical lymphocytes excludes the latter. The presence of small, involuted and collagenized germinal centres with marked vascular proliferation in the interfollicular region ensures a correct diagnosis of the disease.

A less commonly observed histologic pattern is the plasma cell type, which is not restricted to mediastinum and most often is accompanied by systemic manifestations. It is seen in older individuals and involves mesenteric or retroperitoneal lymph nodes. The symptoms include fever, sweating, weight loss and fatigue. The most frequent laboratory abnormalities are anaemia, elevated erythrocyte sedimentation rate, hypergammaglobulinemia (polyclonal), and hypoalbuminemia. Histologically, the lesion is characterized by a diffuse plasma cell proliferation in the interfollicular region and hyaline-vascular changes in the follicles are inconspicuous.⁷

The solitary form of Castleman's disease presents most commonly as mediastinal mass but has also been described in the neck, lung, axilla, mesentery, broad ligament, retroperitoneum, soft tissues of extremities, and in nasopharynx.⁸ Microscopically, over

90% of the cases are of hyaline-vascular type, and the remainder are of the plasma-cell type. Treatment of the disease is surgical excision following which there is rapid regression of both clinical and laboratory abnormalities.

The multicentric or systemic form is nearly always the plasma-cell type.⁹ It presents with generalized lymphadenopathy and may also involve the spleen.^{4,5} Clinical and laboratory features are similar to those of angioimmunoblastic lymphadenopathy, suggesting a related pathogenesis resulting from an abnormal immune response.¹⁰ The disease has been reported in association with POEMS syndrome (peripheral neuropathy, organomegaly, endocrinopathy, monoclonal [M] protein, and skin lesions).¹¹ The long-term prognosis of systemic Castleman's disease is poor as it tends to persist for months or even years and sometimes gives rise to renal or pulmonary complications.¹² An association of the disease either the plasma-cell or hyaline-vascular type with Kaposi's sarcoma, lymphoma¹³, plasmacytoma¹⁴, and Hodgkin's disease¹⁵ has also been reported.

Although mixed or transitional forms have been described, the progression from one variant to another has not been confirmed, and two variants usually do not coexist or occur consecutively. With so much dissimilarity in clinical, morphologic, and therapeutic features of its variant types, Castleman's disease, is presently defined by characteristic histologic patterns that are probably related to conditions of immune deregulation.

Our case was that of solitary form and hyaline-vascular type of Castleman's disease. The patient was asymptomatic except for the swelling in the neck. The patient is on follow-up for the last 2 years with uneventful course. The case is being reported to highlight the unusual presentation as a solitary cervical lymphadenopathy in Castleman's disease. Clinically, the disease maybe misdiagnosed as lymphoma and histologically as follicular hyperplasia or follicular lymphoma. Recognition of the characteristic histopathological features lead to the definitive diagnosis.

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