Angiofollicular lymph node hyperplasia (castleman’s disease) – A Case report

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Case Report

A 55 years old female presented with a painless swelling on the left side of the neck of 20 years duration. She had history of trauma over that region following which she noticed the swelling. It increased in size gradually. It was pyriform in shape and firm in consistency. It occupied the entire left anterior triangle and part of the posterior triangle of the neck. She had no dysphagia, voice changes, weight loss or fever. No significant lymphadenopathy in other areas of the body could be detected. Liver and spleen were not palpable. Previous haematological tests, X-rays neck and chests, thyroid scan, serum T3, T4 and TSH were within normal limits. Rheumatoid factor was negative. Her bleeding time and clotting time were 1 min. 15 seconds and 4 min. 20 seconds respectively. Fine needle Aspiration Cytology (FNAC) revealed non specific lymphadenitis. Excision biopsy revealed an oval firm encapsulated mass, externally covered with greyish white membrane, with prominent blood vessels and areas of shaggy looking fibrous adhesions. The specimen was 7.0 cm x 4.5 cm x 1.0 cm in size. Cut surface was irregular, lobulated and soft yellowish in colour. On microscopic examination, multiple sections revealed structure of a lymph node with cellular infiltrates. There were irregular bands of connective tissues, containing a large number of hyalinised blood vessels running from the capsules to the interior, dividing the lymph node into lobules. Through out the specimen there were lymphoid follicles of varying sizes having both prominent and inconspicuous germinal centres, with hyalinised blood vessels in many of them and having a thick mantle zone of lymphocytes. The interfollicular zone contained plenty of hyalinised blood vessels of post capillary venule type, many capillaries, plasma cells, eosinophils, immunoblasts, sinus, endothelial cells and a fair number of histiocytes. Considering the multiple hyalinised blood vessels in the germinal centre and the polymorphic cytology, a diagnosis of hyaline vascular type of Angiofollicular lymph node hyperplasia (Castleman’s disease) of lymphocyte subtype was made (Fig. 1a and 1b). Follow-up till June 2003 revealed no evidence of regrowth.

Discussion

Castleman’s disease has been classified into two histologic types: the “Hyaline vascular” and the “Plasma cell” type lesions. Hyaline vascular lesions are commoner and characterized by small hyaline vascular follicles and interfollicular capillary proliferation. The Plasma cell type comprises 10 to 20 percent of reported lesions. It is characterized by large follicles with intervening sheets of plasma cells. Systemic manifestations such as fever, hyperglobulinemia and anaemia are frequently associated with the plasma cell type1. Solitary form of this disease has been reported in neck, lung, axilla, mesentry, broad ligament, retroperitoneum, soft tissue of extremities2, para adrenals3 and adrenals4, parotids5, and leptomeninges6.

The differential diagnosis of giant lymph node hyperplasia includes other lymphoid lesions that contain follicles and can attain large size1. Hodgkin’s
disease of mixed cellularity type may involve lymph nodes partially so that a number of follicles remain within the infiltrate. In such cases prominent mononuclear cells increase the resemblance to giant lymph node hyperplasia. However, capillary proliferation and vascularised hyalineic follicles are not features of Hodgkin’s diseases. Nodular lymphoma and thymoma may pose diagnostic challenges, especially in case of mediastinal lymphadenopathy. Rheumatoid lymph nodes and plasma cell type of Castleman’s disease both contain numerous plasma cells and prominent follicles throughout the lymph node7,8. Abnormalities found in association with rheumatoid arthritis such as hypergammaglobulinemia and anemia, resemble manifestations of giant lymph hyperplasia. However, rheumatoid lymph nodes are usually smaller, occur through out the body without a predominant mass and are usually associated with arthritis and rheumatoid factor in the serum2. Viral lymphadenitis3 and atypical lymph node hyperplasia may share with giant lymph node hyperperplasia, the features of prominent follicles, inflammatory cells, basophilic stem cells, the large size and even distribution of follicles throughout the lesion. The presence of hyaline vascular follicles or sheets of plasma cells serves to exclude the diagnosis. Lastly, the follicular hyperplasia present in AIDS related complex10 of this disease. These include hyperplastic, reactive11, hamartomatous2,12,13 and mixed14,15.

Recent works have focussed on possible immunologic pathogenesis for this disease. Some disease with immunologic background have regressed following curative resection. After surgery for hyaline vascular type of Castleman’s disease, remission of pemphigus vulgaria15, an autoimmune disease, could be achieved with reduced doses of steroids in all cases and in at least two cases, steroid treatment could be discontinued.

Castleman’s disease generally follows a benign course. An anterior mediastinal Castleman’s disease of 30 years duration has been reported16. Our case had a history of 20 years duration Multicentric Castleman’s disease with involvement of spleen, bone marrow and bone with, paradoxically, a very benign course has been documented17. However, malignant transformation is also a distinct possibility. 7 cases of vascular neoplasia with Castleman’s disease has been described by Gerald W et al18. This remarkable association may be viewed as a manifesatation of the intimate functional relationship that exists between the immune and the vascular system.

The treatment of choice of Castleman’s disease is surgical resection1. If complete excision is not possible partial resection may be useful as regrowth is not expected. Radiotherapy has produced little shrinkage. The anemia and hypergammaglobulinemia have resisted all forms of therapy except surgical resection of the mass. The treatment of multicentric Castleman’s disease is difficult. Surgical resection is not indicated in multicentric variety19. Long term high dose steroids (80-120 mg of prednisolone per day) has been tried with variable success. Some patients may respond to melphalan and or interferons.

References