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Systemic Amyloidosis in Hodgkin's Disease

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ABSTRACT

Secondary amyloidosis as a complication of Hodgkin's disease has been described as being unusual to rare in occurence. We report a case in which the clinical picture was that of a renal failure, etiology of which could not be determined but which proved to be amyloidosis secondary to clinically unrecognised Hodgkin's disease.

Key words : Amyloidosis, Hodgkin's disease.

CASE REPORT

A 17 year old man was admitted in January 1991 with complaints of diarrhoea, breathlessness, cough with expectoration and loss of appetite since 10 days. In January 1990 he was hospitalized for generalized weakness and fatigue and was treated as a case of anaemia with 2 units of blood and oral hematinics. In November 1990 he suffered from fever with edema feet. Examination had revealed hepatosplenomegaly and enlarged cervical lymph nodes. He was diagnosed as a case of anaemia with hypoproteinemia. Lymph node biopsy at that time was reported by private consultant as 'atypical lymphadenitis'. This biopsy was not available to us for review. Physical Examination revealed that patient was emaciated and drowsy, weighed 32 Kg, there was pallor, bilateral pitting oedema of feet and enlarged, mobile, nontender cervical lymph nodes measuring 0.5 - 1 cm in diameter. Lymph nodes at other sites were not palpable. B.P. was 60 mm Hg (Systolic). Spleen was palpable 3-4 cm, soft and nontender, Liver was just palpable. Respiratory system revealed coarse crepitations at the base of both the lungs.

Laboratory investigations revealed a Haemoglobin of 4.1 gm%, a total WBC count of 13,000/cu mm and a differential count of Neutrophils 70%, Lymphocytes 28%, Eosinophils 1% & Monocytes 1%. ESR was 80mm at the end of 1 hour. Urine examination showed moderate proteninuria. BUN was 312mg%. S. Creatinine was 6.5 mg%. An arterial blood gas analysis revealed metabolic acidosis. Patient's subsequent clinical course was marked by two episodes of generalized tonic clonic convulsions and respiratory arrest which led to his death on 4th day of hospitalisation.

Autopsy findings

External Examination revealed pallor, bilateral cervical lymphadenopathy and pitting edema of feet.

Gross Pathology

Examination of individual organs revealed :

Lymph nodes : cervical, mediastinal and paraaortic group of lymph nodes were enlarged and measured 1-3 cm in diameter. Cut section of these lymph nodes showed homogenous grayish white appearance.

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Spleen : was enlarged and weighed 600 grams. Capsular surface and cut surface of spleen revealed grayish white nodular tumor deposits of 0.2-1 cm diameter (figure 1).

Liver : was enlarged and it's cut surface showed waxy appearance.

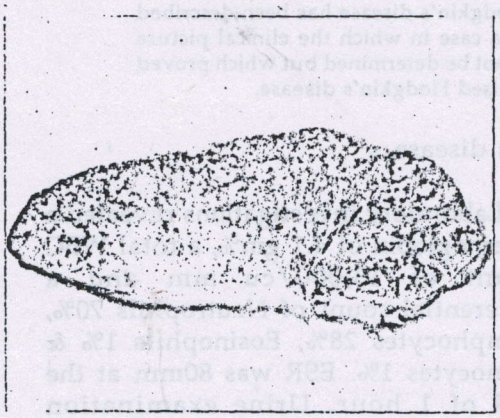

Fig. 1 : Gross photograph of cut surface of the spleen showing diffusely distributed grayish white tumor deposits.

Kidneys : Both kidneys were enlarged and pale. Cut surface of both kidneys revealed widening of cortex.

Examination of lungs did not reveal evidence of tuberculosis. Examination of heart, brain and gastrointestinal tract showed normal anatomical features.

Light Microscopy findings

Lymph Node : Architecture of lymph node was effaced and was replaced by diffuse cellular infiltrate of plemorphic variant of R-S giant cells, histiocytes plasma cells and lymphocytes along with presence of homogenous eosinophilic interstitial deposits which I showed greenish birefringence by polarising microscope on congo red stained slide. Spleen : Microscopic examination showed presence of R-S giant cells along with histiocytes, lymphocytes and plasma cells (figure 2). Nodular deposits of homogenous pink amyloid material was also demonstrated in histological sections of spleen (figure 3). Amyloid was also demonstrated in liver, kidneys & adrenals. There was no evidence of tuberculosis or other chronic inflammatory lesion in any of the organs. Microscopic examination of brain did not reveal any pathology.

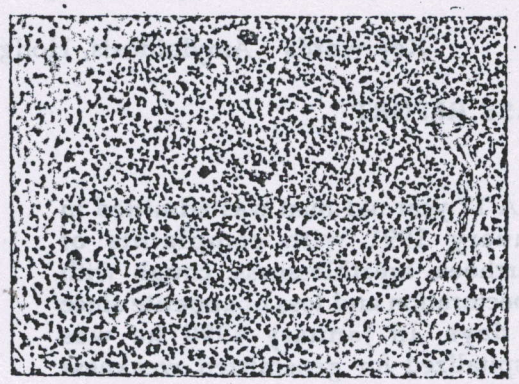

Fig. 2 : Photomicrograph of the spleen showing lymphocytes, histocytes and R-S giant cells having binucleate and multinucleate forms (H & E × 500).

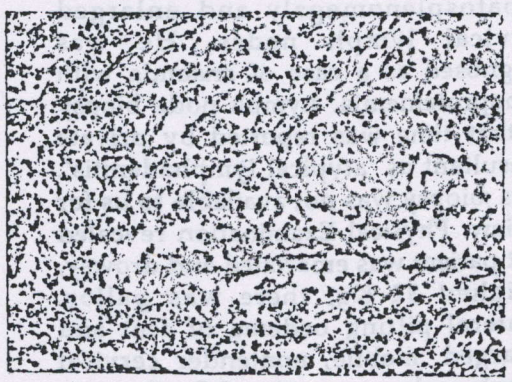

Fig. 3 : Photomicrograph of the spleen showing large, nodular massess of amyloid deposits (H & E × 500).

DISCUSSION

The significance of coexisting amyloidosis with Hodgkin's disease

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was for the first time reported by Wallace et al[1] and till today about 50 cases have been reported[2-+6]. All the reported cases, including ours, are typical of the secondary type of amyloidosis that is involving liver, spleen, kidneys and adrenals. There seems to be no association between amyloidosis and any specific form of Hodgkin's disease[1]. In majority of the cases amyloidosis developed after the diagnosis of Hodgkin's disease, however, occasionally amyloidosis and Hodgkin's disease were diagnosed simultaneously[4]. In our case, the picture presented was that of a progressive renal failure secondary to massive renal amyloidosis and the diagnosis of amyloidosis with Hodgkins disease was established after death. In a case of Hodgkin's disease, development of proteinuria and edema should make one suspect of secondary amyloidosis.

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ERRATA

Indian Journal of Pathology and Microbiology 40(3), 1997, Page 433 authors should be read as Seema Dhiraj Jain, P. Bhalla and SNA Rizvi.