Pictorial CME

AL Amyloidosis

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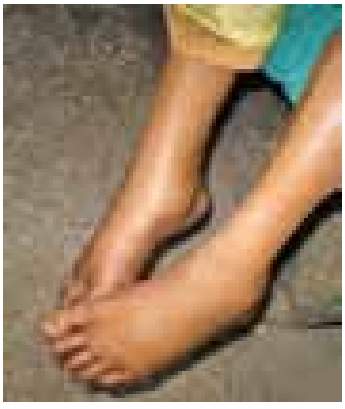

Fig. 1 : Bipedal swelling

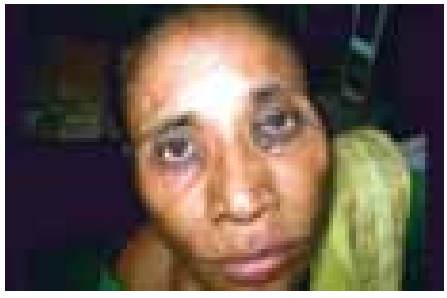

Fig. 2 : Raccon's eye

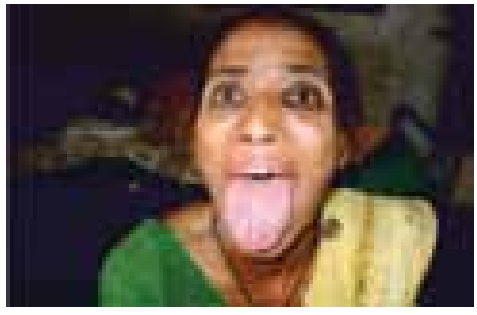

Fig. 3 : Macroglossia

A 53 year old female not known to be diabetic or hypertensive presented with insidious onset and gradually progressive exertional dyspnea, palpitation with history of PND along with weakness of both hand with tingling, numbness and burning dysesthesia for last 2 years, blackening discoloration around periocular area for last I year, swelling of both leg with multiple tongue bite for last 2 months (Figures 1-3). Her past history, family history and drug history nothing contributory.

On examination, mild pallor present, severe macroglossia with fissured tongue, blackening discoloration around periocular area suggestive of Raccon's eye, jugular venous pressure(JVP) elevated, bipedal swelling present, severe wasting of thenar muscles of both hand with broadening of both wrist with tinnel's and phalen's sign positive suggestive of carpal tunnel syndrome, Cardiovascular system-a MR murmur of grade 3/6, Respiratory system- bibasilar cracle, Gastrointestinal system reveals hepatosplenomegaly, Nervous system examination reveals bilateral CTS. Based on the above mentioned clinical findings, it was diagnosed as a case of Amyloidosis.

On investigation, moderate anemia (9.75gm%), L.F.T normal except ALP is 918u/L, routine urine shows 3+albumin with granular cast, 24 hrs urine protein shows nephritic range proteinuria, urine Bence Jone's protein negative, bone marrow study shows hypercellular marrow with increased plasma cells and background shows pink fibrillary amyloid like material, Echocardiography reveals restrictive cardiomyopathy, serum protein electrophoresis ravels monoclonal gammopathy (M Spike) in gamma globulin region (48.54%), immunofixation electrophoresis identifies the 'M' Spike as IgG Lambda, abdominal fat pad aspiration shows Congo red negative fibrillary amyloid material.

From above history, physical examination and investigation report, AL Amyloidosis was diagnosed.

AL Amyloidosis is most commonly caused by a clonal expansion of plasma cells in bone marrow that secrete a clonal IgG light chain(LC) that deposit as amyloid fibrils in tissue.[1,2] AL Amyloidosis can occur in multiple myeloma and other 'B' cell lymphoproliferative disorder. The kidney are the most frequently affected organ followed by heart. Bone marrow, subcutaneous fat pad or rectal biopsy is the preferred mode of tissue diagnosis.

Congo red staining itself is not without limitation.[3] Because Amyloid deposit may have a patchy distribution in the initial stage, Congo red staining may be negative in sampling error, in Light Chain Deposition Disease in particular KAPPA. Typical extra cellular fibrillar deposit can be detected by an electron microscopic examination even when Congo red is negative.

References

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2. Rajkumar SV, Gertz MA. Advances in the treatment of amyloidosis. *N Engl J Med* 2007;356:2413.
3. Falk RH et al: Diagnosis and management of the cardiac Amyloidosis. *Circulation* 2005;112:2047-60.

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