Janaan Health case report of the week 2 November 2016

Bad Amyloid

75 years old lady known to have chronic kidney disease secondary to hypertension followed up routinely in renal clinic in January 2015. She was found to have worsening renal function and subsequent urinary protein evaluation revealed nephritic range protein 4.0g/l and creatinine 178. Her baseline creatinine was 120.

She then underwent further evaluation and serum protein electrophoresis revealed a small band of IGG kappa paraprotein with no immunepaeresis.

Blood tests as below

HB 100g/l ,WCC 6.0 , Neutrophils 3.1, Lymphocytes2.5, PLT 170.

Na 132, K 3.6, Urea 10, Creatinine 180,

Total protein 67, globulin 30, bilirubin 7, ALT 17, ALP 76

B2 microglobulin 2.1, Calcium 2.35

Serum free light chain slightly raised kappa light chain 120 with kappa and lambda ration 2.15

Urine BJP positive with kappa

She is otherwise well a keen gardener.

Lives alone

Her only medication were

Amlodipine 10mg OD

Lisinopril 10mg OD, occasional paracetamol .

She was evaluated in haematology clinic by a specialist register who discuss the case with supervising consultant.

A decision was made to arrange a bone marrow test with trephine congo red to exclude amyloidosis and multiple myeloma.

Her bone marrow test and skeletal survey together with biochemistry results were presented in MDT.

A diagnosis of renal failure secondary to hypertension and MGUS were made.

She then came back to haematology clinic 3 months after the MDT as routine follow up.

During her consultation she mentioned that she is going to have her carpal tunnel release operation by a plastic surgeon in a different hospital in 2 weeks time.

The consultant haematologist immediately got interested on the carpal tunnel syndrome and found on the noted that her nerve electrophysiology report suggested bilateral carpal tunnel syndrome.

Her operation then was postponed.

Her case was discussed in a tertiary centre with national interest on amyloid.

She was reviewed reasonably early where she underwent following tests

1. Caridiac ECHO : no evidence of cardiac amyloidosis

(Of note: most sensitive test for cardiac amyloid is ECHO, NOT SAP Sacn)

1. SAP Scan
2. Bone marrow test with congo red staining Negative with 10% plsam cells.
3. Free light chains were unchanged Kappa lambda ration 2.1
4. Biochemistry and haematology unchanged

Umbilical fat biopsy confirmed AL amyloidosis

Her case was discussed again in MDT

She was commenced on MPV ( Non transplant intention)

After 4 courses of therapy

Her renal function became normal

Para protein disappeared

Serum free light chain ratio normalised

Her carpal tunnel symptoms resolved

Nephritic range protein resolved

Kidney function normalised

FBC normalised

She completed total 6 courses mpv.

She is well and active and enjoying a normal life.

She thanks the team and grateful to get her life back (November 2016) almost 12 months post treatment is continued to be in remission

Learning points

Bi lateral carpal tunnel syndrome with any monoclonal protein and or light chain, amyloidosis until proven otherwise

High degree of clinical suspicion

Low threshold to referring such case to tertiary referral centre with specialist interest