Light Chain Myeloma: A Diagnosis Likely To Be Missed

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ABSTRACT:

Multiple myeloma is diagnosed by 'M' band on serum protein electrophoresis in the appropriate clinical setting of bone pains, evidence of bone destruction on X ray, renal failure, anaemia, hypercalcemia and hyperuricemia. Light chain myeloma is a type of multiple myeloma which has the clinical features of multiple myeloma with absence of 'M' band on serum protein electrophoresis. The diagnosis depends on detection of Bench Jones proteins in urine & urine electrophoresis along with confirmation by bone marrow examination. This patient was initially misdiagnosed as spinal tuberculosis due normal serum protein electrophoresis. Demonstration of Bence Jones proteinuria, urine protein electrophoresis & bone marrow examination confirmed the diagnosis of light chain myeloma.

Introduction:

Multiple myeloma represents a malignant proliferation of plasma cells from a single clone which produce immunoglobulins consisting of heavy & light chains[1]. The most common screening test for multiple myeloma in appropriate clinical setting is serum protein electrophoresis which detects 'M' band consisting of heavy chains of immunoglobulin[2]. Light chain myeloma is a type of multiple myeloma in which plasma cell tumors produce only monoclonal light chain proteins which are filtered by kidneys & hence not detected by serum protein electrophoresis[3]. We present a case of a patient with light chain myeloma whose diagnosis was confounded due to normal serum protein electrophoresis

Case report:

A 62 year old retired executive presented with history of pain in lower cervical region of 4 months duration. He had history of root pains. The pain was excruciating, progressive & had incapacitated the patient. With this complaint he consulted an orthopedic surgeon who advised an X ray of cervical spine. The X ray of cervical spine showed collapse of C4 cervical vertebra; subsequent MRI confirmed the diagnosis of collapse of body of C4 cervical vertebra with cord compression. There was no involvement of other vertebrae or paradiscal areas. Suspecting multiple myeloma his serum protein electrophoresis was done which did not show presence of M band. He was started on AKT with presumptive diagnosis of cervical spine tuberculosis. After 2 months of treatment with AKT, he had no symptomatic relief and hence was referred for physician's opinion. On examination his vital parameters were stable, there was severe tenderness over cervical spine & systemic examination did not reveal any abnormality. On investigation his Hb was 7.8 gm/dl, TLC 3200 /cmm, platelet count was 86000/cmm with ESR 105 mm at end of 1 hour. Serum creatinine was 3.2 mg/dl, serum calcium mg 10.9/dl, serum uric acid 7.6mg/dl. In view of these findings and spinal tuberculosis being uncommon in cervical area his repeat serum protein electrophoresis was done which failed to show presence of M band. However, urine for Bence Jones protein done in view of strong suspicion of myeloma was positive. Urine electrophoresis done subsequently showed

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presence of M band. Diagnosis of myeloma was confirmed on bone marrow examination which showed 40% plasma cells. He was treated with liposomal doxorubicin, cyclophosphamide, vincristine, solumedrol for 2 cycles along with plasmapheresis. With treatment the marrow plasma cell count came down to 8%.Subsequently he was treated with Thalidomide & steroids in view of renal impairment.

Discussion:

Multiple myeloma is a monoclonal plasma cell disorder. The multiple myeloma cell clone produces an excess of monoclonal (M proteins) and free light chain proteins. The M proteins may be recognized as IgA, IgD, IgG, IgE or IgM, depending on their heavy chain class. This excess of M proteins is responsible for the hyperviscosity syndrome, which interferes with fibrin aggregation and platelet function. The serum M component is IgG in 53% of patients, IgA in 25%, IgD 1%.20% patients produce only light chains[4]. 1% of patients do not secrete any immunoglobulins (non secretory).Persons with light chain myeloma also develop lytic bone lesions, hypercalcemia, hyperuricemia, renal failure and have a poor prognosis [5].The light chain proteins may be designated as kappa or lambda. They may precipitate and deposit, producing organ damage. The organ most commonly affected is the kidney. When these monoclonal light chains appear in the urine, they are called Bence Jones proteins.

Multiple myeloma is suspected in elderly with bone pains, headache, anaemia, renal failure. Work up involves biochemical investigations (serum creatinine, serum calcium, serum uric acid), complete blood count, ESR, serum protein electrophoresis. Type IgG or IgA paraproteins associated with multiple myeloma may be found by serum protein electrophoresis testing; however, in case of light chain myeloma the tumor produces only Ig light chains that are removed from the blood by the kidneys due to their lower molecular weight. Hence they are not detected by serum protein electrophoresis [6]. This Ig light chain is detected by heat test for Bence Jones proteins or urine protein electrophoresis and is found nearly exclusively in patients with multiple myeloma. Moreover, urine for Bence Jones protein is not commonly employed as screening test because the heat test used for detecting Bence Jones protein is falsely negative in 50% of patients[6]. Confirmation of diagnosis is by demonstration of more than 10% plasma cells on bone marrow examination. Treatment of light chain myeloma is the same as that of multiple myeloma. The recent introduction of a commercial immunoassay for measurement of free light chains potentially offers an improvement in monitoring disease progression and response to treatment, particularly where the paraprotein is difficult to measure accurately by electrophoresis (for example in light chain myeloma, or where the paraprotein level is very low)[7].

Conclusion:

Few patients of multiple myeloma may have normal serum protein electrophoresis inspite of clinical signs & symptoms. Normal protein electrophoresis does not rule out the diagnosis of multiple myeloma. Such cases can be detected by urine test for Bence Jones protein & urine protein electrphoresis.

References:

1. Nikhil C.Munshi, Dan L. Longo, Kenneth C. Anderson; Harrisonn's Principles of Internal Medicine;17th ed;2007;vol.1:701.
2. Seema Gupta, Raymond L.Comenzo; NACB:Practice guidelines & recommendations for the use of tumor markers in the clinic ;Section 3
3. Mosby's medical dictionary 8th Ed; 1999.
4. Nikhil C. Munshi, Dan L.Longo'Kenneth, C. Anderson; Harrison's Principles of Internal Medicine;17th ed;2007;vol.1:702
5. Xiuli Cong, Xiujuan Sun; Chinese Journal of Clinical Oncology; June 2004, Vol 1 No.3:1672-1673.
6. Hoffbrand, Moss P, Pettit J; Essential Haematology; 5th Ed. 2006: p218
7. Bradwell et al; Cin Chem 2001; 47:4; p673-680.