The treatment and management of patients who have myeloma

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This article aims to provide an understanding of myeloma and its management. It highlights patients’ concerns, the various treatment options, and the implications for nurses caring for people with myeloma. The authors also look at the role of the cancer charity International Myeloma Foundation UK, with particular reference to Myeloma Awareness Week, 21–28 June.

Myeloma is a relatively rare cancer of the plasma cells. These cells are found in bone marrow and are responsible for protecting the body against viruses and infections. In myeloma a single defective plasma cell multiplies rapidly, disrupting the immune system and displacing healthy bone marrow. The abnormal plasma cells can crowd out normal haemopoietic cells in the bone marrow, leading to bone marrow failure, with associated anaemia and immunosuppression.

Myeloma cells have an affinity for bone, resulting in bone damage and pain, the most common and distressing factor of the disease. The disease often appears at several sites in the body; which is why it is often termed multiple myeloma. It is characterised by periods of treatment and varying periods of remission and can progress at different speeds in each patient. Although there is no cure for myeloma, the many treatments now available and in development make it increasingly treatable.

**Epidemiology, aetiology, and symptoms**

Myeloma makes up one per cent of all cancers and 15 per cent of haematological malignancies (International Myeloma Foundation UK, 2000). The annual UK incidence is 4.0 (for women) and 5.8 (for men) per 100,000 (Office for National Statistics, 2002).

The causes of myeloma are not certain but exposure to some chemicals, viruses, and a weakened immune system may be triggers. Exposure to radiation and benzene have traditionally been linked to myeloma but the evidence is inconclusive (Joshua and Gibson, 2002).

The clinical symptoms of myeloma are due both to the consequence of an uncontrolled growth of abnormal plasma cells and to the presence of paraprotein in the blood or urine. The most common presenting symptoms are bone pain or fractures (70 per cent), anaemia (59 per cent) and malaise (46 per cent) (Lokhorst, 2002).

Pain, loss of height, osteoporosis, pathological fractures, spinal cord compression, and hypercalcaemia are all complications of myeloma bone disease. Impaired bone marrow function and immune paresis can cause recurrent or persistent infections, anaemia and associated fatigue, neutropenia and/or thrombocytopenia. The high levels of paraprotein in the blood cause renal impairment and raised plasma viscosity.

**Staging and prognostic indicators**

Staging of the disease is necessary to determine likely prognosis and to assist in decisions regarding treatment. The Durie/Salmon staging system, which was developed in 1975, is still in use today (Durie and Salmon, 1975). Evaluation of haemoglobin, calcium, paraprotein, bone damage, renal impairment, and myeloma cell mass are used to divide patients into three stages – I, II and III, with A and B differentiating between relatively normal and abnormal renal function.

**Treatment**

Guidelines on the treatment of myeloma have been developed by the UK Myeloma Forum (2001) on behalf of the British Committee for Standards in Haematology. Treatment focuses on both control of the disease and improving symptoms. Median survival for patients undergoing treatment is currently 3–5 years (NICE, 2003a).

 Patients with no symptoms (stage IA or asymptomatic myeloma) may remain stable for a long period without treatment. Therapy is indicated only when there is evidence of progression or symptoms of bone disease.

**Standard chemotherapy**

Alkylating agents such as melphalan and cyclophosphamide are effective in treating myeloma. They can be used alone or in combination with steroids, and can be given both orally and intravenously. A variety of chemotherapy combinations, for example VAD (vincristine, doxorubicin (having replaced Adriamycin), and dexamethasone), are used as primary therapy for patients who will be offered high-dose therapy and stem-cell transplant (UKMF, 2001).

**High-dose therapy and stem-cell transplant**

This treatment provides a means of giving higher doses of chemotherapy to consolidate standard chemotherapy, without causing permanent damage to blood cell production. Giving back a patient’s previously collected healthy stem cells after high-dose chemotherapy effectively rescues the marrow and enables healthy blood-cell production to continue (IMF UK, 2004). It is often the
treatment of choice for patients under 60 years, but can be considered in those up to the age of 70. Allogeneic transplants, usually with a sibling donor, are a potential option for a small group of younger patients and should be performed within the context of a clinical study.

**Thalidomide**

Thalidomide was first used to treat relapsed disease but it is increasingly used as a primary treatment and maintenance treatment after autologous stem-cell transplant. Owing to its history of teratogenicity, it is prescribed in the context of strict risk management guidelines.

**Bortezomib**

The first new treatment for myeloma in over a decade, bortezomib is a revolutionary approach to treating myeloma. It is a new type of drug called a proteasome inhibitor that acts on a group of enzymes found in cells in the body. Proteasomes play a part in controlling cell function and growth and this therapy interferes with their function, causing cells to die and possibly stop the cancer from growing. This method of action is unique.

Bortezomib has recently been licensed for patients who have received at least two prior therapies but in whom disease progression has continued.

**Supportive measures**

Various treatments can be used to relieve the symptoms of myeloma. Bisphosphonate treatments such as sodium clodronate and zoledronic acid have been shown to reduce myeloma. Bisphosphonate treatments such as sodium clodronate and zoledronic acid have been shown to reduce the underlying disease. Chemotherapy can relieve pain by treating the underlying disease.

Radiotherapy is also an effective treatment, particularly at the site of painful lytic lesions, and myeloma cells are particularly sensitive to radiotherapy. Opiate analgesia is often required and can be used alone or in combination with adjuvant methods. The use of non-steroidal anti-inflammatory drugs should be avoided owing to their association with renal impairment.

Recently, percutaneous vertebroplasty has been used to treat back pain caused by vertebral bone disease, with some success. The procedure involves an injection of surgical cement into the vertebral space of collapsed vertebrae. NICE has produced guidelines on its use (NICE, 2003b). Spinal-cord compression is an oncological emergency and often requires immediate treatment with steroids and radiotherapy.

Symptomatic anaemia (caused by both the disease and chemotherapy treatment) is managed via blood transfusion or recombinant human erythropoietin. Antibiotic therapy, IV immunoglobulins, and growth factors are all used to treat or prevent complications associated with immunosuppression. Prophylactic vaccination against influenza may also be used (UKMF, 2001).

Adequate hydration (3L daily) is essential to prevent and treat renal impairment (UKMF, 2001). Other measures include treating the underlying myeloma and plasmapheresis to remove excess paraprotein. Whereas a degree of renal impairment occurs in up to 50 per cent of patients (UKMF, 2001), renal failure occurs in 3–12 per cent (Clarke et al, 1999) and is managed by dialysis.

New therapies for treating myeloma are under investigation, including Revlimid (a thalidomide analogue), skeletal targeted radiotherapy, arsenic trioxide, and vaccines. **Nursing considerations**

A diagnosis of myeloma is devastating and often worsened because many patients have not heard of the disease (Box 1). Nurses contribute to all aspects of care of a patient with myeloma - most importantly, in the prevention and management of complications of the disease and its treatments, and in providing information and support. Key goals for effective management include: pain assessment and control; educating patients on the need for adequate hydration; prompt recognition of the signs of spinal-cord compression and hypercalcemia; management of fatigue; psychosocial support of the patient and family; and provision of relevant, high-quality, up-to-date information at all stages of their disease.

**The role of IMF (UK)**

The International Myeloma Foundation (UK) informs and supports people affected by myeloma, and helps to improve treatment and standards of care through research and education. The MAGIC for Nurses programme (myeloma, advocacy, guidelines, and information community) aims to provide specialist myeloma information to haematology nurses, to enable them to offer patients information and support. It is partly supported by a grant from the Department of Health and is free to join.

**Awareness week**

Awareness of myeloma is relatively low among the public, the general medical community, and certain levels of government. The week of 21–28 June is UK Myeloma Awareness Week 2004, which aims to increase awareness of myeloma at all levels in the community and raise vital funds for research.