Multiple myeloma presenting as mandibular pain

Précis

Abstract
Introduction: Multiple myeloma (MM) is a systemic malignancy of plasma cells defined by monoclonal production of circulating immunoglobulins. Bone pain is a presenting feature in the majority of cases. Treatment may involve intravenous use of bisphosphonates, chemotherapy or haematopoietic stem cell transplantation. Here, we illustrate a first presentation of MM in a patient with mandibular pain and discuss radiographic, diagnostic and treatment challenges of orofacial issues in patients with MM.

Case report: A 69-year-old lady presented to an emergency oral surgery clinic with a month-long history of unilateral left-sided pain in her jaw. Examination revealed a buccolingual swelling of 2cm diameter in the lower left premolar region. Radiographic images demonstrated a 2cm lytic lesion in her mandible corresponding with the symptomatic region. Aspiration of the lesion was performed and histological analysis indicated an abundance of atypical plasma cells. Subsequent biopsy revealed sheets of plasmacytoid cells suggesting evidence of a plasmacytoma. Skeletal survey, bone marrow biopsy and serum analysis confirmed the presence of MM.

Discussion: MM, although a systemic malignancy, can present via a variety of orofacial manifestations. The presence of a lytic lesion on plain radiographs should alert the dental practitioner to the possibility of the diagnosis. Treatment of orofacial bone pain may respond to intravenous bisphosphonates but care must be taken to avoid osteonecrosis of the jaw.

Conclusion: This case is an important reminder of the potential oral presentations of MM and underlines the importance of radiographic evaluation in patients with atypical symptoms and presentations.
Introduction

Multiple myeloma (MM) is a systemic malignancy characterised by neoplastic proliferation of plasma cells in the bone marrow resulting in monoclonal immunoglobulin production. Primary clinical presentations vary from bone pain and pathological fractures to incidental abnormalities on routine blood testing such as anaemia, hypercalcaemia and impaired renal function. Bone pain is a feature of initial presentation in 58% of cases. The presence of a solitary extramedullary lesion (plasmacytoma) is seen in 7% of cases, and is associated with a more aggressive natural history of MM. Plasmacytoma may present as a subcutaneous palpable mass. It is estimated that MM accounts for 1% of all malignancies and 10% of haematological cancers. Here, we illustrate a case of MM, initially presenting as an orofacial abnormality. We take the opportunity to discuss potential orofacial and dental presentations, challenges and complications that may occur in patients with MM.

Case presentation

A 69-year-old lady attended an emergency oral surgery clinic with a one-month history of pain and swelling in the left side of her mandible, resulting in an inability to wear her lower denture. Initially described as “burning” in nature, and considered to be neuralgia, the pain did not respond to analgesia prescribed by her general practitioner. The pain began to affect her quality of life as the patient felt lethargic and was unintentionally losing weight due to difficulty eating with the pain in the mandible. She also described numbness in the distribution of the left mental nerve. Examination of the edentulous patient revealed a swelling expanding in a buccolingual direction occupying the lower left premolar region approximately 2cm in size. Extra-orally no abnormality was seen, although there was tenderness in the area of the swelling. On radiographic examination (Figures 1 and 2) a lytic lesion, approximately 2cm in size with a poorly-defined, non-corticated border was evident in the left premolar region, indicative of aggressive underlying pathology and necessitating further investigation.

Aspiration of this lesion was performed and histological examination demonstrated abundance and clustering of atypical plasma cells, highly suspicious for a plasma cell neoplasm. Subsequent biopsy of the same lesion revealed fragments almost completely composed of sheets of plasmacytoid cells. At presentation, full blood count and routine biochemistry for this patient were normal apart from a raised total protein (86g/L) in the presence of a normal albumin level (39g/L). Serum
immunoglobulin measurement found elevated levels of IgA with decreased levels of IgG and IgM. A paraprotein screen detected the presence of a monoclonal IgA Kappa band, raising the suspicion of active MM. In addition, radiological skeletal survey revealed multiple lytic lesions in the skull (classical appearance of ‘raindrop’ or ‘pepper-pot’ skull) and axial skeleton suggestive of myelomatous deposits (Figures 3 and 4). Referral to the haematology service was made and a diagnosis of MM was later confirmed following bone marrow aspirate and trephine, which demonstrated a clonal plasma cell infiltrate, with plasma cells comprising up to 40% of the nucleated cell count. This patient is currently receiving ongoing treatment under the haematology service. On reviewing the patient in our clinic there has been little, if any, change radiographically. Intra- orally the swelling in the left premolar region has reduced greatly. Since commencing bisphosphonate therapy the mandibular pain has subsided.

Discussion

The exact incidence of MM presenting initially with orofacial abnormalities is currently unknown. While rare, oral presentations do occur and include:

- jaw pain;
- painless or painful swelling;
- painless gingival mass;
- mandibular numbness;
- bilateral mandibular mass;
- mobility of teeth;
- migration/drifting of teeth; and,
- macroglossia (often secondary to amyloidosis).6

These symptoms may mimic common dental pathologies, which in turn can lead to delays in diagnosis and treatment. This highlights the importance of a thorough knowledge of the potential oral manifestations, both clinical and radiographic, to arrive at the appropriate diagnosis and referral pathway.

While excisional or incisional biopsy remains the gold standard for diagnosis of myelomatous lesions, there is evidence to suggest that fine needle aspiration (FNA) biopsy is a viable option in the early investigation of a suspicious mass or lesion.7,8 The advantages of FNA biopsy are ease of execution, low complication rate, and rapid diagnosis. In particular, FNA can provide a specimen for analysis in situations where an excisional/incisional biopsy could pose significant morbidity. If FNA cytology is inconclusive, progression to incisional or excisional biopsy should be considered. If the risk of morbidity is great and alternative lytic lesions are present elsewhere, alternative means of diagnosis should be discussed with the appropriate specialties.

Systemic treatment of MM depends on stratification of the severity of disease as determined by florescent in situ hybridisation (FISH) of the bone marrow biopsy at diagnosis. Treatment options may include haematopoietic cell transplantation and chemotherapy. For the treatment of bone pain and to reduce the risk of pathological fractures, use of intravenous bisphosphonates is effective but can result in medication-related osteonecrosis of the jaw (MRONJ).6,9 Prevention of MRONJ should be a clinical priority in patients with MM receiving bisphosphonate therapy. Careful consideration of oral hygiene with a review by a dentist and hygienist should occur before commencement of bisphosphonate therapy, followed by regular self-surveillance and dental follow-up.11 If dental extraction or oral surgery is required, it should be completed and time allowed for healing before commencement of bisphosphonate therapy.11 Implementation of preventive dental measures appears to reduce the risk of developing MRONJ.11

Conclusion

While orofacial presentations of MM are rare, they are not unheard of. The atypical and varied nature of their presentation represents a diagnostic challenge for dental professionals. As illustrated, practitioners should be aware that MM may manifest with abnormalities at a multitude of orofacial sites, with a variety of symptoms including both painful and painless mandibular swellings.

MM should be included in the differential diagnosis when such a poorly defined radioopacity is seen. A high index of suspicion should be held in patients with atypical symptoms, with a background history of monoclonal gamopathy of unknown significance (MGUS) or with abnormal radiological findings (e.g., lytic lesion).

References