Bulbar palsy in multiple myeloma

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Case report

A 51-year-old presented with stage IIIB light chain multiple myeloma with marrow plasmacytosis and proteinuria. She achieved a partial response with three courses of VAD (vincristine, doxorubicin and dexamethasone), and underwent non-myeloablative allogeneic bone marrow transplantation (BMT) with marrow from an HLA-identical sibling. She developed grade II mucocutaneous chronic graft versus host disease six months after BMT, and was put on oral cyclosporine A and methylprednisolone therapy. Bone marrow biopsy then showed 8% plasma cells. Serum immunoglobulin showed persistent immune paresis of serum IgG, A and M, and twenty-four hour urine collection revealed proteinuria of 2.5/day. She was admitted nine months after BMT because of a two-week history of difficulty of swallowing and recurrent choking. Physical examination showed absent gag reflex, and the patient could not protrude her tongue, which was thus consistent with bilateral bulbar palsy. (Figure 1) Magnetic resonance imaging (MRI) scan of the skull showed a mass lesion arising from the clivus extending superiorly into the dorsum sella. (Figure 2) Abnormal signal change was also noted in the basi-occiput and in both hypoglossal canal suggestive of infiltration. The abnormal tissue was isointense on T1-weighted and mildly hyperintense on T2-weighted scan, with enhancement after contrast administration. (Figure 2) Repeat Bone marrow showed 60% plasma cells. Twenty-four hour urine collection revealed proteinuria of 11g/day. Cerebrospinal fluid and MRI of spine was not obtained. She was kept nil by mouth, and started on total parenteral nutrition. One dose of bortezomib (1.3 mg/m^2) was commenced but she developed aspiration pneumonia and died one week afterwards. Her family refused a post-mortem examination.

Discussion

The mass lesion at the base of skull in our patient was likely a plasmacytoma that had arisen as a result of progressive myeloma disease, which was evidenced by concurrent 60% marrow plasmacytosis. A skull base abscess was unlikely in the absence of fever and headache. In addition, the MR signal characteristics were one of a solid lesion, compatible with a plasmacytoma[1] and not of a fluid containing peripheral enhancing abscess or haematoma.

As the patient had light-chain disease, serum immunoglobulin levels would not have been useful in monitoring disease activity. This lack of serum immunoglobulin surveillance had also precluded detection of disease progression, which might have occurred sometime before presentation with bulbar palsy. However, the development of serum free light chain assay in the future may be useful in disease monitoring and early detection of progression.[2] In this case early detection would have effected intervention with augmentation of graft versus myeloma effect,[3] which might have halted the development of the skull base plasmacytoma.

While plasmacytoma is a known complication of multiple myeloma, its occurrence at the base of skull is extremely rare. The plasmacytoma had infiltrated the hypoglossal canal, resulting in bulbar palsy. It is also highly probably that the absent gag reflex was the result of tumour infiltration of the invasion of IX, X and XI nerves that traverse both jugular foramina. The resultant bulbar palsy had reduced the patient to a poor nutrition status, thus predisposing her to the fatal aspiration pneumonia.

Figure 1. The patient could not protrude when asked to.

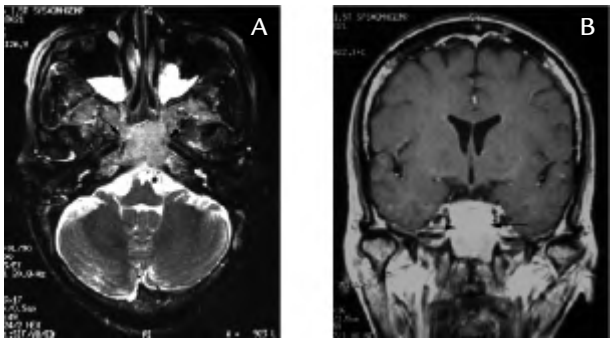

Figure 2. A. Post contrast T1-weighted axial scan through the brain showing enlargement of the clivus (arrows) with enhancing tissue. Note polyp in the left maxillary sinus and fluid in both maxillary sinuses **B.** Axial T2-weighted scan through the skull base showing abnormal signal in the basi-occiput (long arrow) and both hypoglossal canal (small arrows).

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