Skull Base Involvement in Waldenstrom's Macroglobulinemia: MR Findings

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Purpose
To report a unique case of Waldenström's macroglobulinemia (WM) with involvement of the central skull base, masticator space/buccal fat, cavernous sinus, and perineural disease. This pattern of disease involvement has never been described in WM.

Materials & Methods
A 70-year-old man with long history of Waldenström's macroglobulinemia who was asymptomatic for several years presented with altered mental status and decreased visual acuity. The patient underwent CT and MR scans of the head. A lumbar puncture was performed. An orbital mass was biopsied and fine needle aspiration of right pterygopalatine fossa was done. He was treated for WM, and his symptoms gradually dissipated, and his CSF cell count and protein content improved.

Results
The initial CT scan showed communicating hydrocephalus. The lumbar puncture demonstrated lymphoplasmacytic involvement of the meninges, by CSF cell count and electrophoresis, which was thought to have caused the hydrocephalus. MR imaging demonstrated infiltrative changes of the sphenoid bone and clivus, infiltration by soft tissue of masticator space including the buccal fat bilaterally, large bilateral orbital mass lesions, enhancement along the left V2, pterygopalatine fossa as well as left cavernous sinus (see figures). Infiltration of masticator space and buccal fat was ill-defined without focal mass lesions. The orbital mass lesions extended along the optic nerves. The masses were characterized as having low signal on T1- and T2-weighted images with intense enhancement following administration of Gd-DTPA. The imaging characteristics of orbital lesions are similar to those described by Ettl et al. CT findings were obliteration of normal fat in left inferior orbital fissure, pterygopalatine fossa, and in masticator/buccal space corresponding with sites of infiltration seen on MR imaging. The orbital masses were hyperdense on nonenhanced CT. To our knowledge, involvement of the skull base, infiltration of the soft tissues near the skull base, or perineural disease have never been described in WM. Orbital involvement in WM have been described in the past (1).
Conclusion
This case confirms the potential for extensive involvement of the skull base, cavernous sinus, and perineural disease in patients with lymphoplasmacytic infiltration of Waldenström's macroglobulinemia.

References
1. Ettl AR, Birbamer GG, Philipp W. Orbital involvement in Waldenstrom's macroglobulinemia: Ultrasound, computed tomography and magnetic resonance findings. Ophthalmologica 1992;205:40-45