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Fig. 1. A Panorex view of the mandible shows generalized medullary sclerosis and loss of the normal bony trabeculae. The closed arrow identifies a well-defined, sclerotic, bony protuberance at the right mandibular angle. Note the multiple unerupted teeth (open arrow)

History

This 18-year-old black male presented with an approximately one-year history of a large mass in the right mandibular angle noticed after a tooth extraction. The family history included an older sister with multiple soft tissue masses of the face and chest wall. Physical examination at the time of admission showed an 8 x 9 cm smooth, hard, fixed, non-tender mass in the region of the right mandibular angle. A similar lesion was observed in the mid-forehead region. The laboratory data were normal.

A complete skeletal survey, including studies of the skull and mandible, was obtained (Figs. 1, 2, and 3) as well as barium studies of the gastrointestinal tract (Fig. 4).
Fig. 2. A film of the chest shows multiple areas of cortical thickening in the ribs as well as several discrete, sclerotic, bony projections.

Fig. 3. Films of the humerus and femur demonstrate areas of cortical thickening and bony excrescences arising from these major long bones.

Fig. 4. A film obtained during an air contrast barium enema illustrates the presence of multiple polyps of varying size throughout the visualized segments of the colon (arrows).