Atypical Presentation of a Chordoma Mimicking a Pituitary Adenoma

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Abstract

69-year-old male presented with a history of hypo-cortisolism and mild vision loss. MR imaging of the sella demonstrated a sellar based mass with heterogenous avid enhancement and T2 hyperintense signal. CT angiographic pre-opereative imaging done for surgical planning purposes demonstrated intralesional calcifications within the lesion. Surgical resection of the mass was performed utilizing endoscopic trans-nasal, trans-sphenoidal, trans-sellar, approach of the mass that was noted intra-operatively to involve the clivus. Successful gross total resection was performed. Final pathology demonstrated findings compatible with a Chordoma.

Introduction

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Chordomas are rare, locally aggressive malignant bone tumors arising from notochordal remnants, with an annual incidence of approximately 0.1 per 100,000 individuals.[1] The skull base accounts for approximately 30-32% of cases, with the clivus representing the most common site of skull base origin.[1]

Neuroimaging Findings
On MRI, clival chordomas characteristically demonstrate low to intermediate T1 signal and very high T2 signal intensity, with variable contrast enhancement.[2] CT imaging typically reveals bony expansion and intralesional calcifications.[2]

Differential Diagnosis
Chordomas presenting primarily within the sella are exceedingly rare and can mimic pituitary adenomas both clinically and radiologically.[3] Other differential considerations for sellar masses include craniopharyngiomas, meningiomas, and Rathke's cleft cysts.[3]

Pathology
Histologically, chordomas are characterized by physaliphorous cells, vacuolated cells with bubbly cytoplasm, arranged in lobules within a myxoid matrix.[2] Immunohistochemically, chordomas express brachyury (a highly specific nuclear marker), cytokeratin, and S100 protein.[1]

Image 1:

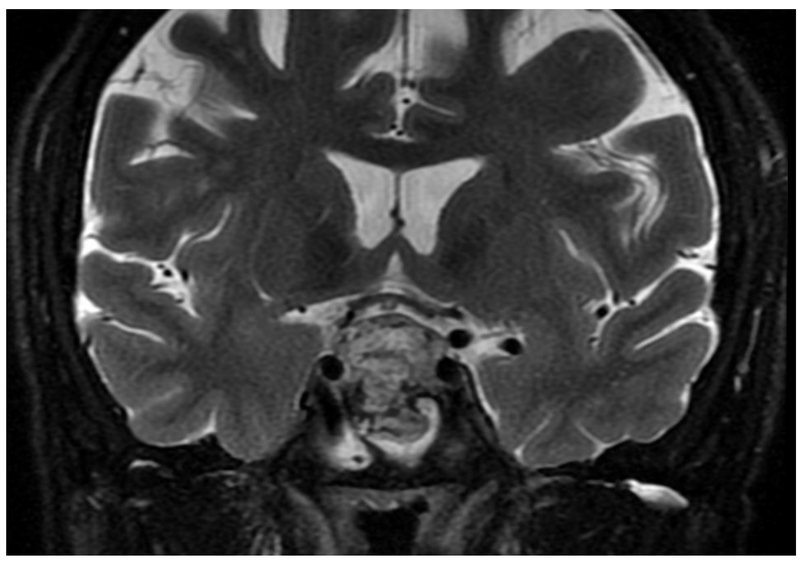

Image 2:

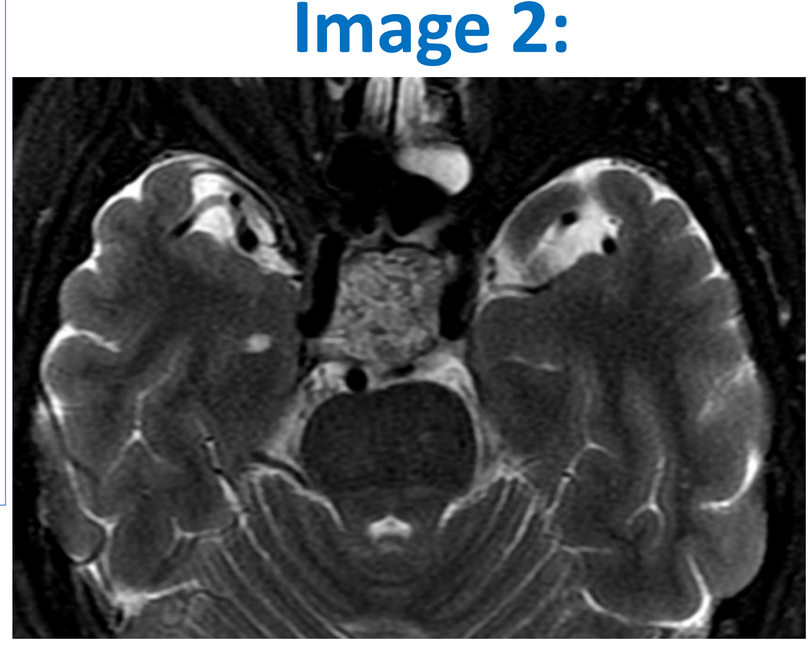

Image 1 and 2: Coronal T2 and Axial T2 weighted images demonstrate a heterogeneously hyperintense midline sellar based mass with lack of visualization of the normal pituitary gland and extension into the adjacent bony clivus. A key imaging feature is the degree intratumoral “bubbly” T2 hyperintense signal.

Image 3:

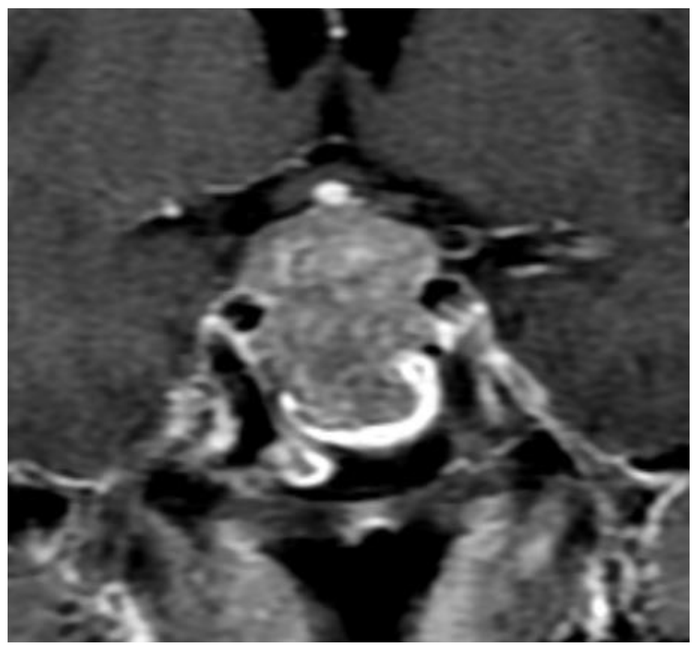

Image 3: Coronal post-contrast T1- weighted image demonstrates a well-defined mass with heterogeneous enhancement that is relatively hypointense compared to the pituitary gland that appears flattened along the inferior margin the mass.

Image 4:

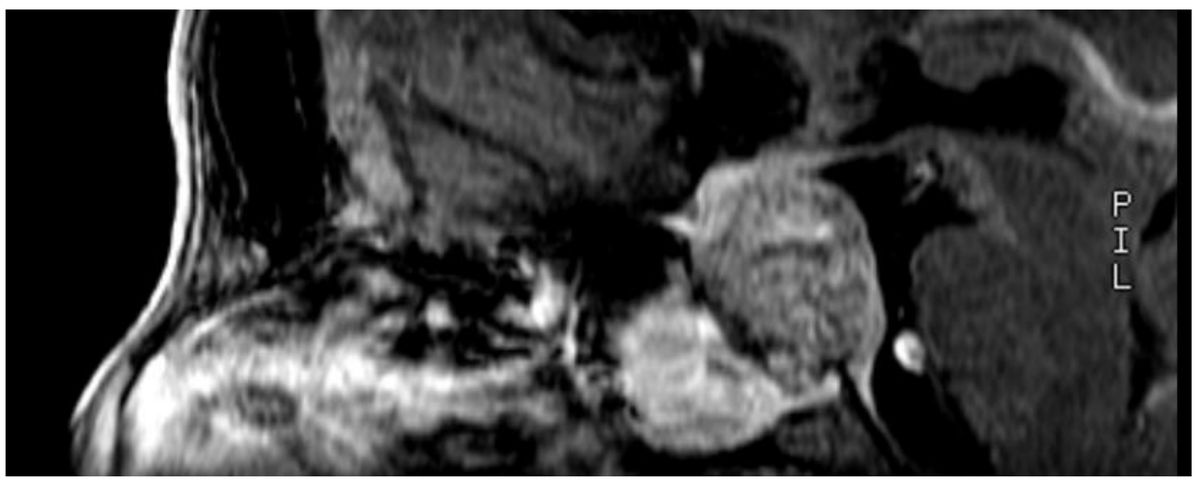

Image 4: Sagittal T1-weighted image redemonstrates the heterogeneously enhancing sellar-based mass with at least partial extension/involvement of the adjacent bony clivus and protrusion into the prepontine cistern.

Image 5:

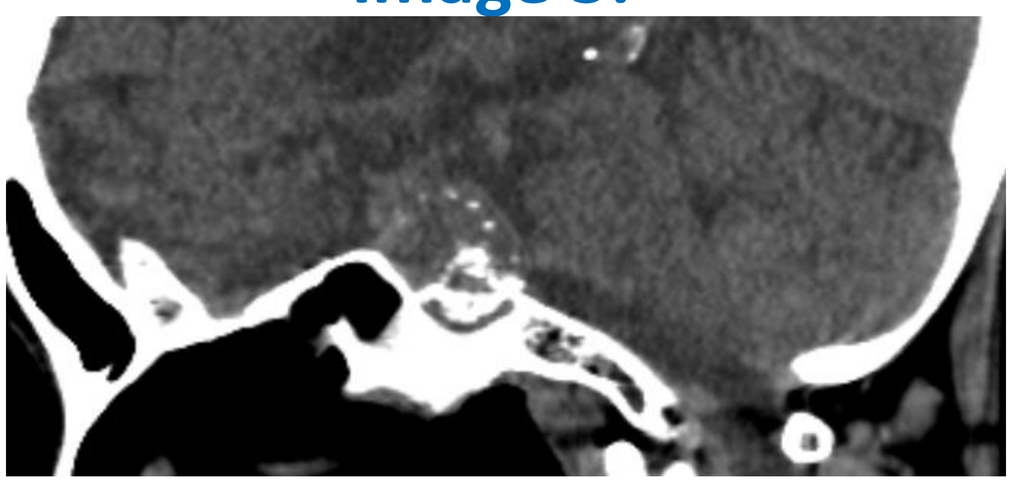

Image 5: Sagittal noncontrast CT demonstrates key findings of intralesional coarse, punctate, and curvilinear calcifications which represent sequestra from bone destruction rather than dystrophic calcifications of the tumor itself.

Conclusions

Clival Chordomas centered near the sella may have imaging features suggestive of pituitary adenomas, however, key findings such as intralesional calcifications and intense internal T2 hyperintense signal can server as distinct neuroimaging features in the preoperative evaluation.

References

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