International Journal for Multidisciplinary Research (IJFMR)
E-ISSN: 2582-2160 ● Website: www.ijfmr.com ● Email: editor@ijfmr.com

Misdiagnosis of Clival Lesion As Macroadenoma and Subsequent Identification As Clival Chordoma: Emphasizing the Role of MRI

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Abstract

Clival chordomas are rare, malignant tumors arising from remnants of the notochord, comprising less than 1% of all bone tumors. Due to their insidious growth and location at the skull base, they are frequently misdiagnosed as pituitary macroadenomas, which present with similar clinical symptoms, including headaches and visual disturbances.

This case study highlights the diagnostic challenges encountered in a 65-year-old male, initially diagnosed with pituitary macroadenoma based on clinical presentation and early imaging findings. Despite the proximity of the lesion to the sellar region, initial CT imaging failed to differentiate the clival chordoma from a pituitary macroadenoma due to overlapping features such as bone destruction and soft-tissue mass appearance.

However, subsequent magnetic resonance imaging (MRI) provided crucial details that led to the correct diagnosis. T2-weighted MRI revealed heterogeneous hyperintensity, necrotic and cystic components, and low-signal-intensity septations—features not typically seen in macroadenomas. Post-contrast enhancement patterns and evidence of local invasion into surrounding structures further supported the diagnosis of clival chordoma. This case underscores the importance of MRI in the differential diagnosis of skull base tumors, especially when clinical and initial imaging findings are inconclusive. Early and precise use of MRI can prevent misdiagnosis and guide appropriate treatment planning, particularly in rare pathologies like clival chordomas.

Introduction

Clival chordomas are rare, malignant tumors originating from embryonic remnants of the notochord, typically arising from the clivus at the skull base. Accounting for less than 1% of all bone tumors, clival chordomas are slow-growing yet locally aggressive, posing diagnostic challenges due to their location and symptom overlap with other skull base tumors. The most common initial clinical presentation includes headaches, visual disturbances, and cranial nerve deficits.

This case highlights the diagnostic challenges of differentiating clival chordoma from pituitary macroadenoma. We present the case of a 65-year-old male initially diagnosed with macroadenoma on a computed tomography (CT) scan, with a subsequent magnetic resonance imaging (MRI) revealing the correct diagnosis of clival chordoma.

A 65-year-old male with no history of addiction or significant past medical history presented with progressive visual disturbances and headaches over several months. His symptoms gradually worsened, leading to suspicion of an intracranial lesion. Initial imaging via CT suggested a pituitary macroadenoma, but further investigation with MRI revealed a clival chordoma.

Clinical Presentation and Examination

The patient reported a gradual decline in vision and chronic, progressively worsening headaches. Visual acuity testing revealed reduced visual fields in both eyes. Despite these symptoms, ophthalmological examination showed no abnormalities in the retina or optic nerve, prompting further investigation through neuroimaging.

Approach to Diagnosis and Management

A non-contrast CT scan of the head was performed as the first-line imaging modality. The scan revealed a centrally located, well-circumscribed, expansile mass in the sellar region, suggestive of a pituitary macroadenoma. The lesion appeared to originate near the clivus, raising concern for invasion into adjacent structures. While macroadenoma was initially suspected due to the location and mass effect, there was subtle bony destruction in the clivus, prompting further evaluation.

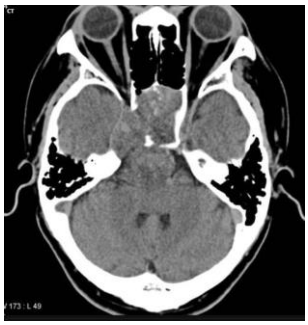
The initial CT findings led to a working diagnosis of pituitary macroadenoma, and the patient was referred for MRI for more detailed assessment of the mass and surrounding structures.

A contrast-enhanced MRI scan was performed to clarify the nature of the sellar mass and guide treatment planning. MRI revealed several key differences that helped differentiate clival chordoma from macroadenoma:

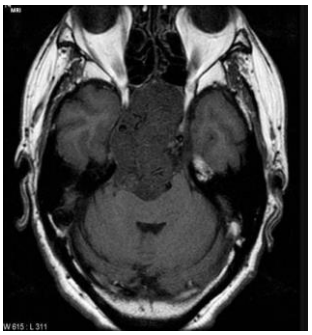
- **T1-Weighted Imaging (T1-WI)**: The tumor appeared isointense to slightly hypointense compared to surrounding brain tissue. Small foci of hyperintensity within the lesion suggested intratumoral hemorrhage or mucous accumulation, features not commonly associated with pituitary macroadenomas.
- **T2-Weighted Imaging (T2-WI)**: The mass was predominantly hyperintense on T2-WI, consistent with the fluid-rich nature of clival chordomas. In addition to cystic areas and necrosis, low-signal-intensity septations were observed, which further indicated the chordoma's complex internal structure. Pituitary macroadenomas, by contrast, typically show more homogeneous signal intensities.
- **Post-Contrast Enhancement**: The lesion demonstrated heterogeneous enhancement, with peripheral areas enhancing more prominently than the center. This enhancement pattern, combined with the extensive bony erosion of the clivus, pointed towards clival chordoma rather than macroadenoma.
- **Local Invasion**: MRI also revealed local extension of the tumor into the cavernous sinus and encroachment toward the brainstem, features more consistent with clival chordoma than pituitary macroadenoma.

Based on the MRI findings, the initial diagnosis of pituitary macroadenoma was revised to clival chordoma, a rarer and more aggressive tumor.

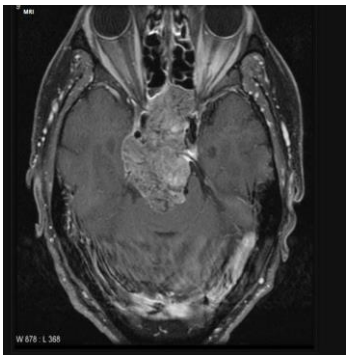
Imaging



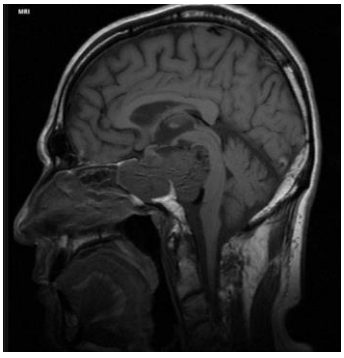
CT showing centrally located, well-circumscribed, expansile mass in the sellar region



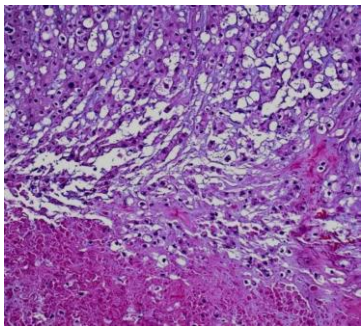
T1W axial images showing slightly hypointense lesion compared to surrounding brain tissue



T1 C^+ fat sat images demonstrated heterogeneous enhancement, with peripheral areas enhancing more prominently than the center.



T1W saggital images showing hypointense lesion



HP confirming chordoma : characteristic physaliphorous cells, a diagnostic feature of chordomas, embedded in a myxoid stroma.

Management

Surgical Approach: Transsphenoidal Surgery (TSS)

Given the revised diagnosis of clival chordoma, surgical resection was planned. The preferred approach for accessing skull base tumors such as chordomas is **endoscopic transsphenoidal surgery (TSS)**, which allows for minimally invasive removal of the tumor. The patient underwent TSS for resection of the clival mass.

During surgery, the lesion was grossly identified as a soft, blush-grey mass with significant infiltration into the clivus. The tumor was carefully resected, and the surgical site was packed with fat and fascia to promote healing and prevent cerebrospinal fluid (CSF) leaks.

The endoscopic transsphenoidal approach is particularly useful for chordomas due to its ability to access the tumor directly through the sphenoid sinus, minimizing damage to surrounding structures while allowing for maximum tumor resection.

Histopathological Findings

Histopathological examination confirmed the diagnosis of clival chordoma.

Microscopy revealed characteristic physaliphorous cells, a diagnostic feature of chordomas, embedded in a myxoid stroma.

The presence of notochordal remnants and myxoid degeneration confirmed the diagnosis of clival chordoma, validating the revised diagnosis made after MRI.

Post-Operative Management and Radiation Therapy

Due to the high risk of recurrence, particularly in cases where total resection is challenging, the patient was advised to undergo adjuvant **proton beam therapy**.

Proton therapy is preferred for chordomas due to its precision in delivering radiation to the tumor while sparing surrounding critical structures, such as the brainstem and optic nerves.

Long-term follow-up was recommended to monitor for signs of recurrence, as clival chordomas have a high propensity for local recurrence, even after aggressive surgical resection and radiation therapy.

Differential Diagnosis

Clival chordomas can be challenging to differentiate from other skull base tumors due to their similar clinical presentations. In this case, the tumor was initially misdiagnosed as a pituitary macroadenoma based on CT findings. The following differential diagnoses were considered:

1. **Pituitary Macroadenoma**: These are the most common tumors in the sellar region and often present with visual disturbances due to compression of the optic chiasm. On MRI, macroadenomas typically show homogeneous enhancement and do not exhibit the same level of bony destruction as clival chordomas.
2. **Meningioma**: Meningiomas are slow-growing, benign tumors that can occur at the skull base. They typically show a dural tail on MRI and do not invade bone to the same extent as chordomas.
3. **Craniopharyngioma**: This tumor commonly occurs in the sellar and suprasellar regions and can mimic the imaging characteristics of chordomas due to its cystic and solid components. However, craniopharyngiomas often present with calcifications, which are less common in chordomas.
4. **Chondrosarcoma**: Another rare skull base tumor, chondrosarcomas can arise in the clival region and share imaging characteristics with chordomas. However, they are derived from cartilage rather than notochordal tissue and show different histopathological features.

Discussion

Clival chordomas are rare, locally aggressive tumors that often present with non-specific symptoms such as headaches and visual disturbances, making early diagnosis challenging. As in this case, the initial imaging findings may mimic more common sellar lesions such as pituitary macroadenomas. While CT scans can detect bone destruction, MRI is essential for detailed characterization of soft tissue and tumor composition, as well as for identifying local invasion.

Accurate differentiation between chordomas and other skull base tumors is crucial for guiding treatment. In this case, the correct diagnosis was only established after detailed MRI imaging, which revealed the tumor's heterogeneous nature, high fluid content, and extensive bony destruction—features more consistent with clival chordoma.

The surgical approach to clival chordomas has evolved with the advent of minimally invasive techniques, particularly endoscopic transsphenoidal surgery. This approach allows for better access to the clivus and surrounding structures, minimizing morbidity while maximizing tumor resection. Despite aggressive surgical treatment, chordomas have a high recurrence rate, necessitating adjuvant therapies such as proton beam therapy.

Conclusion

This case underscores the importance of comprehensive imaging in the diagnosis and management of clival chordomas. While initial CT imaging suggested a pituitary macroadenoma, MRI was crucial in identifying the correct diagnosis of clival chordoma. Endoscopic transsphenoidal surgery, combined with post-operative proton therapy, offers the best chance of long-term control, although close monitoring for recurrence is essential.

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