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Clival Chondroma with Brainstem Invasion Findings in MRI: A Case Report

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Abstract **Case Report**

Chordoma is a rare neoplasm derived from remnants of the fetal notochord; the most commun locations are the lumbosacral spineand the clivus. Clival chordomas represent 35 to 40% of cases and are very difficult to treat because of the localization and high recurrence rate. We present the case of 60 years old male patient with no previous history consulting for diplopia and headach that worsened in the last week before the diagnosis, with a walking ability disorder. Neurological examination found a raised intracranial pressure syndrome with static cerebellar syndrome. However, he presented left hemifacial numbness due to left fifth cranial nerve palsy and diplopia at left gaze due to left sixth cranial nerve palsy. The diagnosis was made by MRI using the different sequences especially T1 SE, T1 GE, T2 FLAIR, DWI and spectroscopy.

Key words: Chordoma; Brain stem; Brain tumor; cancer; MRI, Spectroscopy.

INTRODUCTION

Chordomas are rare, usually benign, neoplasms that are thought to arise from remnants of the embryonic notochord along the spinal axis with incidence of approximately 1/1,000,000 [1]. Classic locations for chordoma include the lumbosacral spine (30-50%) and clivus (30-35%) [1]. Intracranial chordomas usually arise from the region around the clivus and account for about a third of all chordomas in most series. Clivus chordomas seldom metastasize and usually become symptomatic by slow growth and local invasion to affect nearby cranial nerves and brainstem structures. These tumors are difficult to manage because of their critical location and propensity to recur, and they are treated with various combinations of surgery and radiotherapy.

MR imaging is the single best imaging modality for both pre- and post-treatment evaluation of intracranial chordoma. On T1-weighted MR images, intracranial chordomas demonstrate intermediate to low signal intensity and are easily recognized within the high-signal-intensity fat of the clivus. On T2-weighted MR images, they characteristically demonstrate very high signal intensity, a finding that likely reflects the high fluid content of vacuolated cellular components. Moderate to marked enhancement is common and often heterogeneous on contrast material–enhanced images [1]. Combination treatment with radical surgical resection and proton beam radiation therapy achieves the best results. Conventional radiotherapy adds no benefit to the treatment of chordomas [2].

PATIENT AND OBSERVATION

We present the case of a 60 years old male with no previous history that consulted for a slowly progressive headach over a 3 months period, vomiting and diplopia with a walking ability disorder. Neurological examination found a raised intracranial pressure syndrome with static cerebellar syndrome; an ophthalmologist conducted a fundus exam noting a stage 3 bilateral optical disc swelling. On the base of these clinical findings, a head MRI was conducted and found a well-defined mass of the clival region invading the pons and mesencephalon. The lesion was hyperintense on T2W and predominantly hypointense on T1W with small scattered areas of hyperintensity possibly representing foci of hemorrhage and on Fluid attenuation inversion recovery (FLAIR) we found a hypointense mass with peritumoral hyperintensity (Figure 1).

Diffusion weighted imaging showed no areas of restriction suggestive of infarction, the mean ADC

value was 1376 x 10^6 mm2 /s (Figure 2). On T1 Spin Echo and Gradient Echo weighted sequences with injection of Gadolinium we found an important heterogeneous enhancement of the mass that measured in theses sequences 35.9 x 21.9 x 39.1 mm (Figure 3).

Mono and Multi VOXEL spectroscopy was performed on the tumor and showed an increased Choline level with low N-acetyl aspartate and lipid levels (Figure 4).

The patient benefited from surgical biopsy using the retro sigmoid approach which confirmed the diagnostic of chondroma.

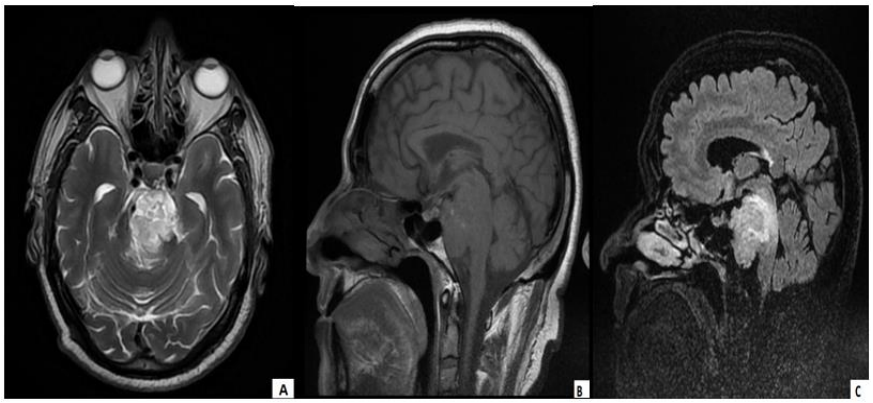

Fig-1: MRI sequences without Gadolinium injection T2W axial sequence showing a hight T2 intensity of the clival region with brain stem invasion, (B) T1W sagittal sequence showing an hypointense lesion of the clival region with Small scattered areas of hyperintensity representing foci of haemorrhage, (C) Sagittal CUBE FLAIR sequence showing a high signal tumor of the clival region represented by chordoma.

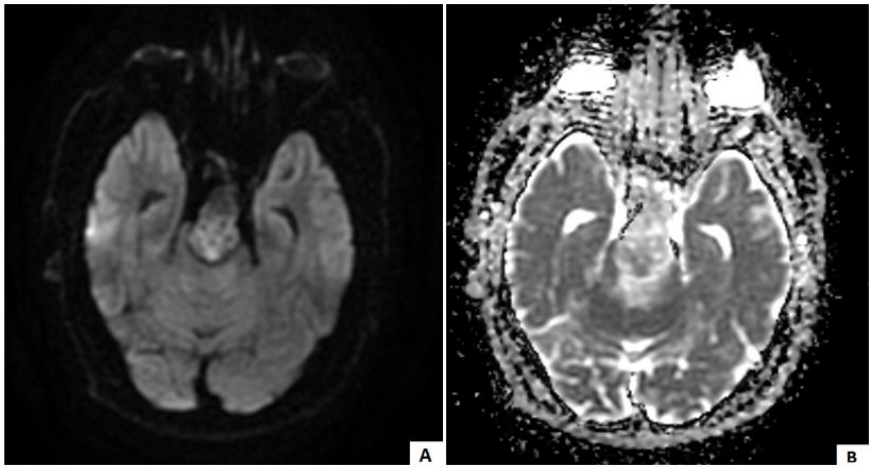

Fig-2: Axial diffusion weighted imaging and ADC images showing non areas of restricted diffusion within the brainstem Axial DWI sequence, (B) Apparent diffusion coefficien (ADC)

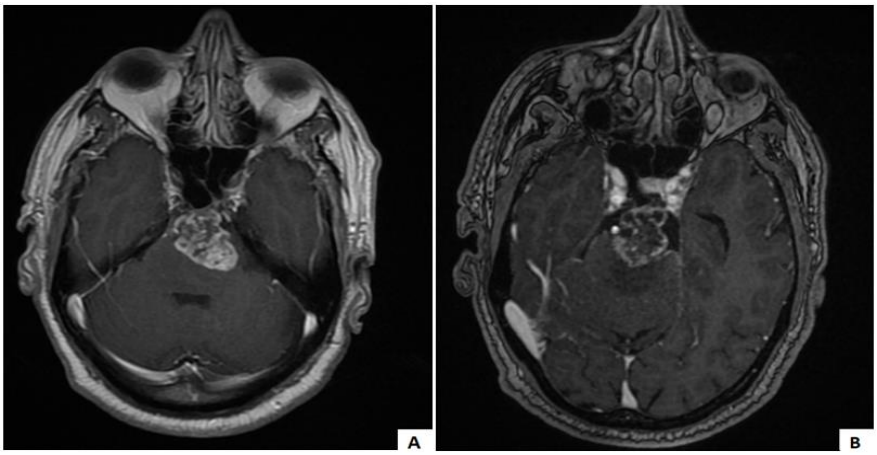

Fig-3: MRI T1 sequence after Gadolinium injection showing an important heterogeneous enhancement of the mass. Axial T1 Spin Echo Gado + (B) Axial T1 Gradient Echo Gado +

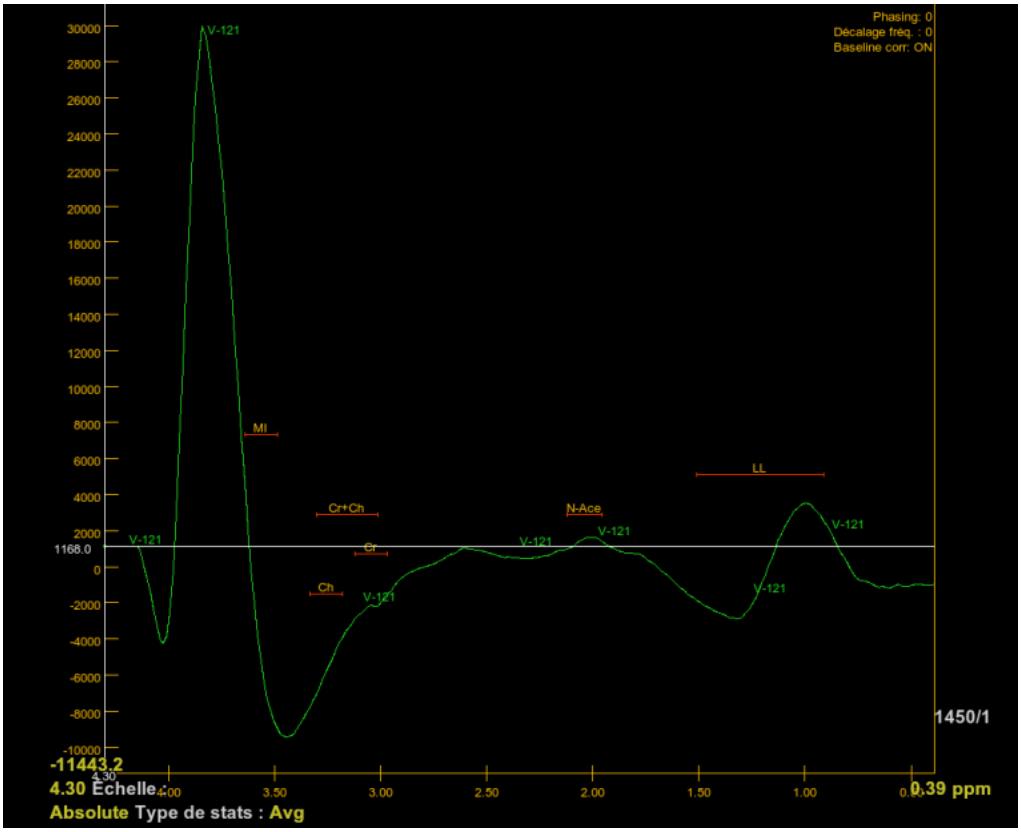

Fig-4: MRI Spectroscopy MONO VOXEL TE 144 showing an increased Choline level with low N-acetyl aspartate and lipid levels

DISCUSSION

Chordoma is a rare primary bone tumor that accounts for 2–5% of all primary bone tumors. It arises from embryonic remnants of the primitive notochord; they have an incidence rate of 01/1,000,000, with a median age at diagnosis of 58.5 years and prevalence in males [1]. The most common presenting symptom is headache and diplopia related to cranial nerve palsy with the abducent nerve being the most commonly affected.

Chordomas may occur at any site along the course of the embryonic notochord. McMaster *et al*. [3] found in a review of 400 patients, that the chordomas were relatively evenly distributed along the cranial (32%), spinal (32.8%), and sacral (29.2%) portions of the axial skeleton. Intracranial chordomas most often originate from the spheno-occipital synchondrosis of the clivus.

The classic midline clival chordoma can spread anteriorly, laterally, posteriorly, inferiorly and superiorly, thereby affecting the sellar area, petrous apex–middle cranial fossa, prepontine cistern, foramen magnum–nasopharynx, and chiasm– third ventricle, respectively. Usually more than one of these areas is involved [4].

Three histological subtypes are described: classical, chondroid and dedifferentiated. The chondroid subtype has a better prognosis [4].

Imaging and especially MRI is useful not only for diagnostic purposes, but also for selection of operative approaches. However, CT scan can be superior to MRI in the diagnosis of bone invasion of the clival region. Therefore the two imaging modalities are complementary in the diagnosis of chordoma.

On conventional spin-echo T1-weighted MR images, intracranial chordoma has intermediate to low signal intensity. Small foci of hyperintensity can sometimes be visualized in the tumor on T1- weighted images, a finding that represents intra tumoral hemorrhage or a mucus pool. The presence of hemorrhagic foci can be confirmed with gradient-echo imaging that is susceptible to blood, at which the foci appear as dark areas. Classic intracranial chordoma has high signal intensity on T2-weighted images, a finding that likely reflects the high fluid content of vacuolated cellular components. The intratumoral areas of calcification, hemorrhage, and a highly proteinaous mucus pool usually demonstrate heterogeneous hypointensity at T2-weighted imaging. Low-signal-intensity septations that separate high-signal-intensity lobules are commonly seen, corresponding to the multilobulated gross morphologic features of the tumor. Also, T2-weighted imaging is excellent for

differentiating tumor from adjacent neural structures especially the FLAIR sequence [5].

The enhancement pattern of the tumor sometimes has a "honeycomb" appearance created by intratumoral areas of low signal intensity. Sze *et al.* [6] reported that because a watery, gelatinous matrix is replaced by cartilaginous foci, chondroid chordomas have shorter T1 and T2 values than do typical chordomas. Therefore, chondroid chordomas may not be as bright as typical chordomas on T2-weighted MR images. This finding is an important prognostic factor due to the significantly better survival rate of patients with chondroid chordoma.

Proton magnetic resonance spectroscopy provides non-invasively a wide spectrum of the biochemical information, which can be used for the estimation of the tumor type, grade and proliferative activity, prediction of the response to therapy and prognosis, and monitoring of the therapeutic response. Chordoma usually shows predominance of Choline peak, absence of Creatine and N-acetylaspartate (NAA) peaks, and prominent Lactate and Lipid peaks [7].

CONCLUSION

Clival chordomas are a slow-growing intracranial neoplasm with many presentations. Posterior fossa involvement and especially brain stem extension are associated with several catastrophic complications; MRI plays a major role in the diagnosis and the quantification of the extension using different sequences with or without injection of contrast including proton magnetic resonance spectroscopy [8]. However, MRI can be inferior to CT scan in bone involvement of the tumor. The treatment should be based on a multidisciplinary team approach. Whenever possible, complete surgical removal of the tumor mass should be performed to ensure better local control and long-term survival.

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