

Clivus Chordoma: An interesting clinical presentation

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Citation

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Abstract

Clivus chordomas are rare central nervous system tumors which arise from base of the skull. We present a 10 year old female who presented with unique bulbar symptoms and and vascular compressive symptoms. We emphasize the fact that these tumors are rare in children with less than 25 cases reported in children, and pose a diagnostic and therapeutic challenge

CASE

A 10-year-old female presented with three-month history of moderate frontal headaches. Three days prior to her admission she developed double vision and drooping of her right eyelid. Past medical, family, birth history was insignificant. Comprehensive physical examination was normal except, presence of right third nerve palsy and left sixth nerve palsy with binocular diplopia. Anisocoria with right pupil 5mm and left 3mm both reacting to light, was present. MRI of the brain revealed mass arising from clival skull base. The tumor was very large 4.5x2.7x2.6 cm compressing the brain stem Figure (FIG 1-C) with deviation of right internal carotid and basilar artery flow voids. Day 4 she developed acute onset left sided weakness. MRI of the brain revealed right MCA infarct and patient was taken for two-staged subtotal chordoma resection. CT Angio revealed right occlusion of the cavernous part of the right internal carotid artery. Pathology confirmed clivus chordoma.

Figure 1

Figure 1a: Saggital T-1 weighted MRI imaging depicting compression of the brainstem by a heterogenous mass arising from the clival plate



Figure 2

Figure 1b: Axial MRI Diffusion Weighted imaging shows Right middle cerebral artery infarct. This finding was confirmed with corresponding hypo intensity on ADC mapping consistent with an ischemic infarct

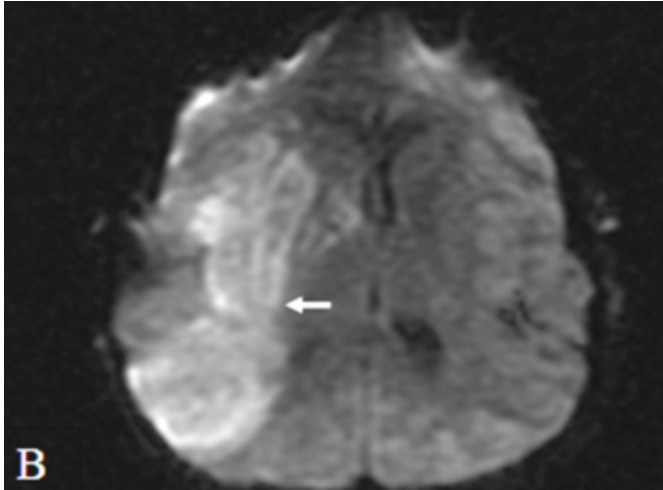
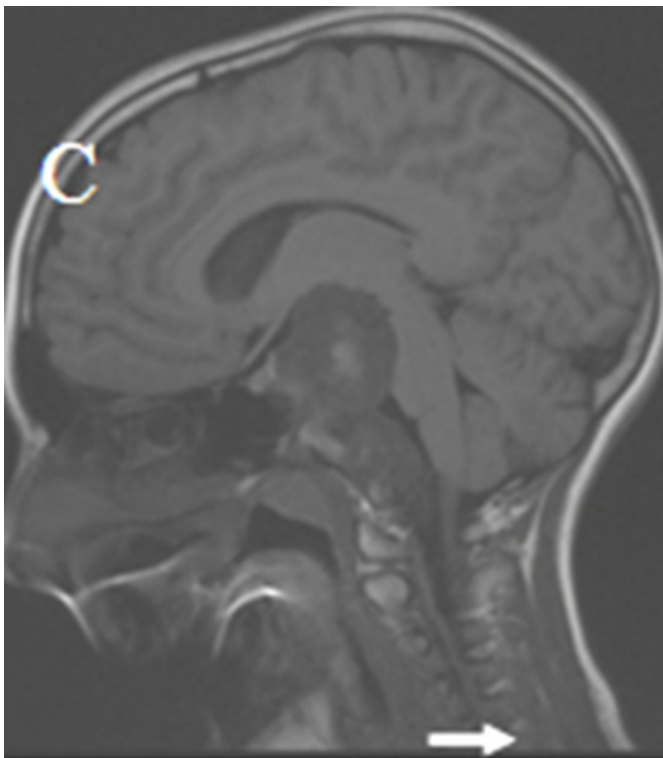


Figure 3

Figure 1c: Saggital CT Angiogram of the neck and brain shows right carotid occlusion.



DISCUSSION

Clival chorodomas are extremely rare skull based tumors arising from notochordal remnants, which have difficult surgical access, high local recurrence rate and metastatic potential. As per review by Yadav et al (1), less than 25 cases have been reported in young children. Our patient had compressive right carotid occlusion with clivus chorodoma, which was partially resected. Postoperatively, patient has left sided hemiplegia with resolved cranial nerve deficits. We emphasize importance this unique clinical presentation with vascular and bulbar compressive symptoms.

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References

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