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Clival Chordoma in Children a Rare Condition: Case Report

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Abstract A three year child had presented to our OPD with complaints of nasal obstruction, change of voice and difficulty in ingestion. Patient was investigated and was seen having a mass in the nasopharynx. Further investigations revealed on CT and MRI brain a large tumour destroying the clival region with nasopharyngeal and intracranial extension with pressure effect on brain stem and lower cranial nerves. Patient underwent two stage surgery retromastoid craniectomy with excision of intracranial part and later a tracheostomy and transoral excision of the nasopharyngeal extension and part anterior to brain stem. Patient made a remarkable recovery and was discharged home after tracheostomy removal with advice to follow with pediatric oncology for chemotherapy as HPE revealed it to be clival chordoma a rare entity in children.

Keywords: *clival chordoma in children*

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1. Introduction

Clival chordoma a skull base condition is encountered commonly in adults. But in pediatric age group it is a rare condition. We however in our institution had this 3 year old child presented with nasal obstruction and later found to a clival chordoma and managed successfully with good outcome.

2. Materials and Methods

A 3 year old child had presented to our OPD with complaints of nasal obstruction, recurrent attacks of upper respiratory infections and voice change with difficulty in swallowing leading to episodes of coughing. Patient was seen initially by ENT surgeons and on post rhinoscopy found a mass in nasopharyngeal area. Patient underwent CT scan followed by MRI brain which revealed large mass destroying the clivus occupying the nasopharyngeal area with intracranial extension compressing the brain stem and infiltrating the lower cranial nerves as Figure 1.

3. Results

Patient after a detailed discussion with parents about all possible risks and benefits of surgery, underwent two stage surgery. Patient initially underwent a retromastoid approach with decompression of intracranial part of the tumour which was compressing the brain stem and engulfing the lower cranial nerves as Figure 2. Patient made a remarkable recovery after surgery. Wounds healed well weaning was started but extubation was difficult in view of lower cranial nerve palsy. After a gap of two weeks once patient was still on ventilator a tracheostomy was done and transoral excision of the tumour was done from the nasopharyngeal area and part anterior to brain stem after removing the invaded clival bony tissues as Figure 3. Postoperative period was uneventful. Patient after of ventilator was shifted to the ward and after downsizing the tracheostomy tube and seeing the tolerance of patient to tracheostomy closure, tracheostomy was removed. Patient was sent to pediatric oncology as histopathology report (H1708145) revealed It to be clival chordoma a rare entity.

4. Discussion

In review of literature Yadav YR in 1992 described the cranial chordomas in 1st decade a rare condition and its management [2]. Pediatric chordomas were further illustrated by Beccaria K in 2015 [3]. Vinke RS in 2015 described a prepontine chordoma in an 11 yr old boy [4]. Chang SW in 2008 also gave a detailed account of management protocols adopted for juvenile chordomas [5]. Warnick RE gave in 1991 gave a detailed account of chordomas in tentorium cerebella [6]. Hazarika D in 1995 gave details of diagnosis of oropharyngeal lesion diagnosed to be chordoma after fine needle aspiration biopsy [1].

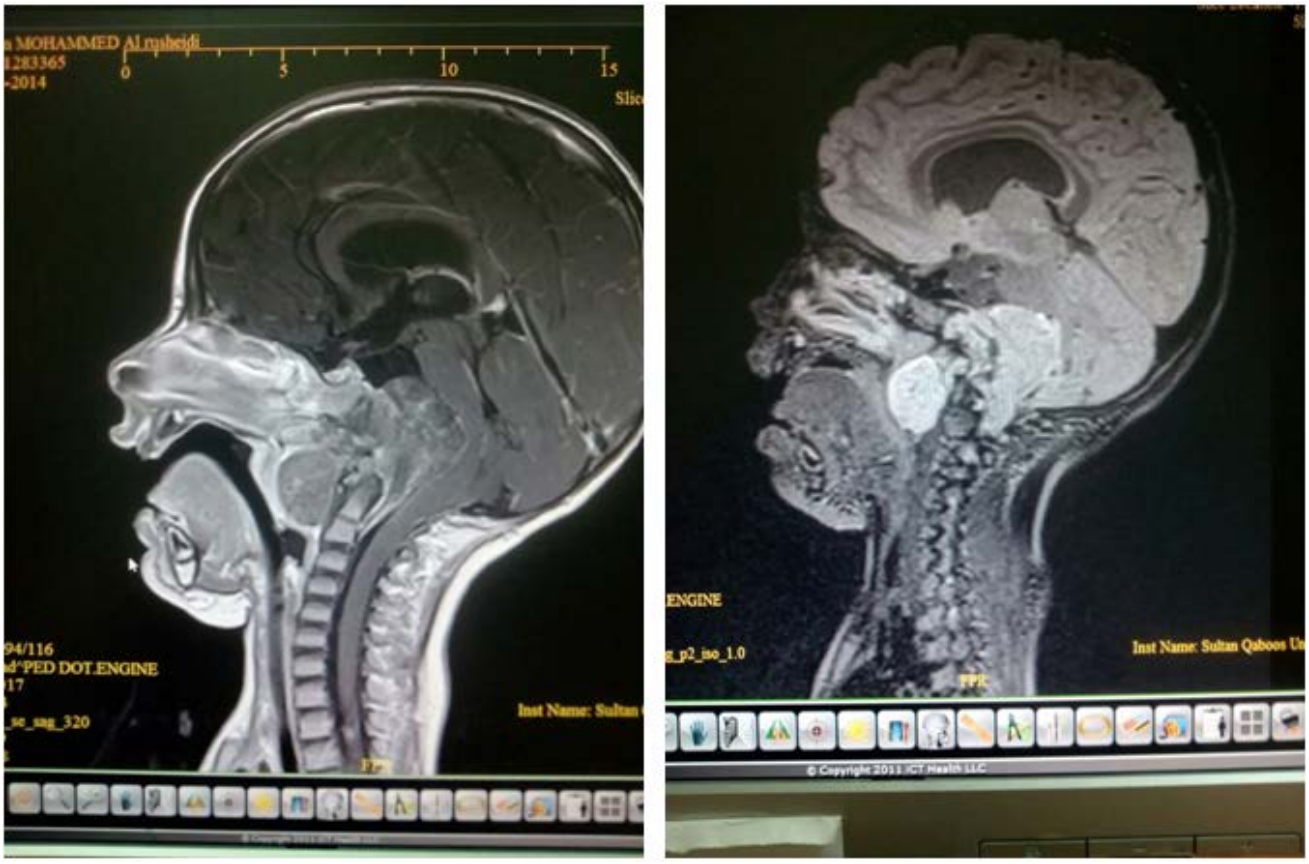

Figure 1. Pre op MRI pictures of the lesion

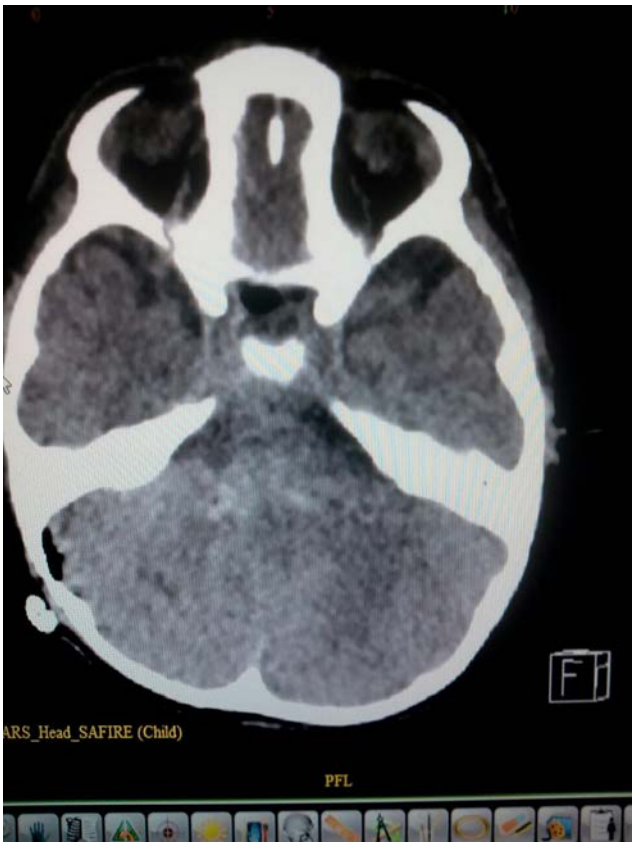

Figure 2. Post op picture after 1st stage retromastoid excision of the tumour

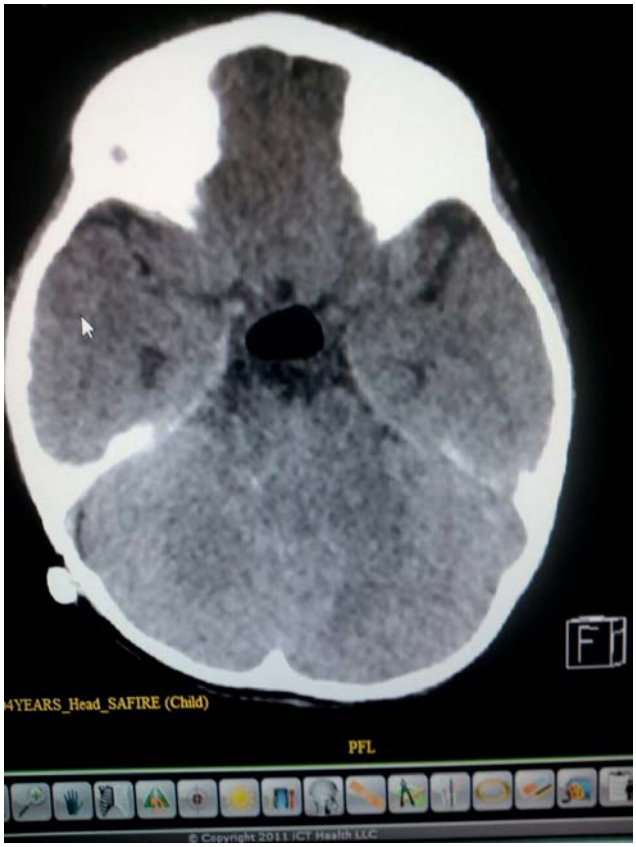

Figure 3. CT picture after 2nd stage transoral excision of the tumour

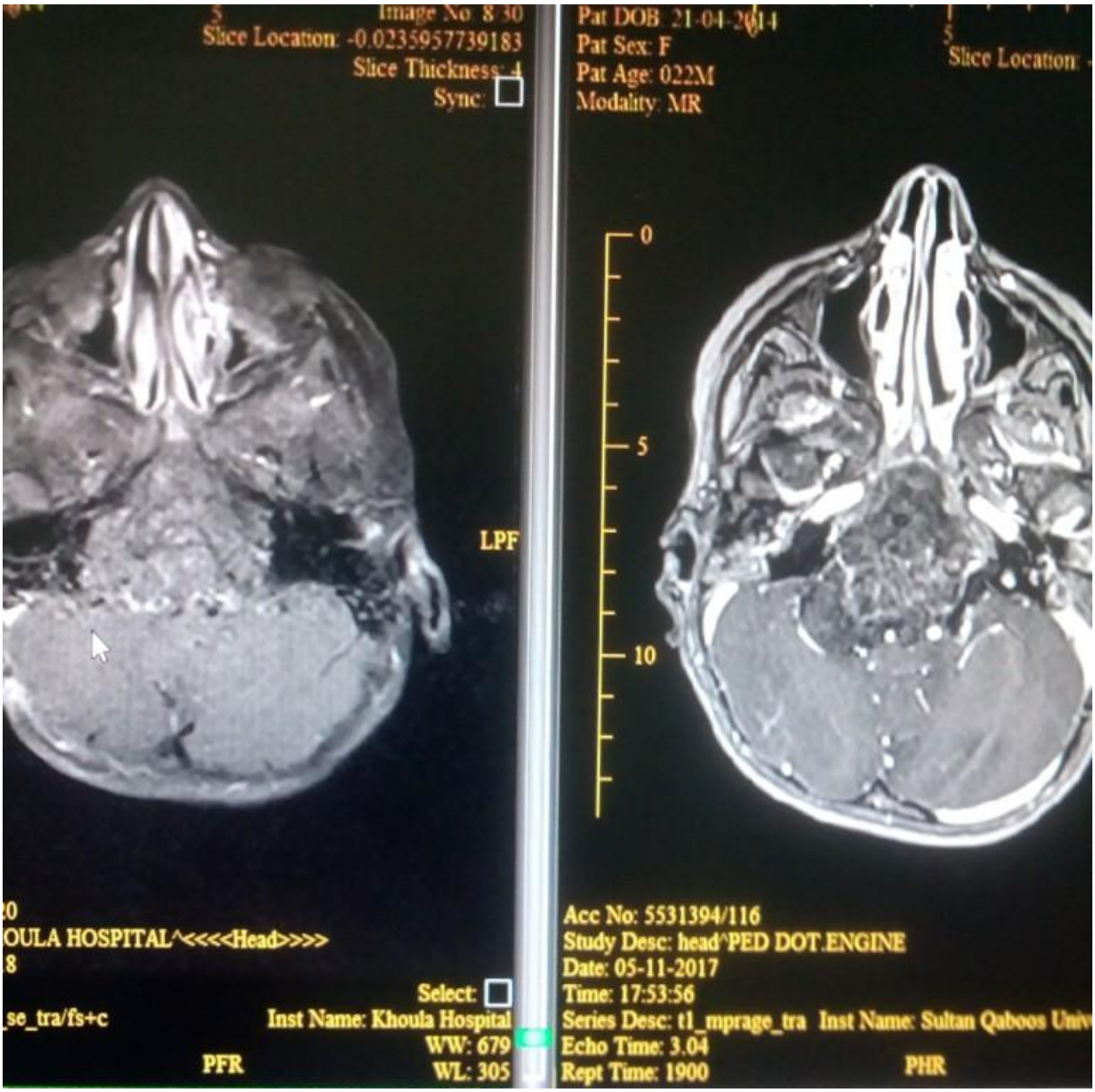

Figure 4. Pre and post op MRI of the patient

5. Conclusion

To conclude clival chordomas in children are a rare entity. Presentation can be due to local symptoms due to invasion of pharynx or intracranial due to lower cranial nerve involvement or brain stem compression. Diagnosis remains by imaging viz CT/MRI and surgical excision or decompression. Chemotherapy is required to delay the recurrence and for a fair outcome.

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