

# A Rare Retroclival Intradural Lesion: Ecchordosis Physaliphora

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**Abstract:** *Ecchordosis Physaliphora is a rare benign lesion derived from the notochord remnants. It can be found anywhere from skull base to sacrum in midline. Most commonly it is located in the retroclival prepontine region. It could be confused with the differentials of chordoma, skull base metastases, neuroenteric cysts, epidermoid cyst and arachnoid cyst. Hence, knowledge about this rare entity is important to differentiate it from other differentials.*

**Keywords:** Ecchordosis Physaliphora, Hamartomatous lesion, Benign, Conservative management

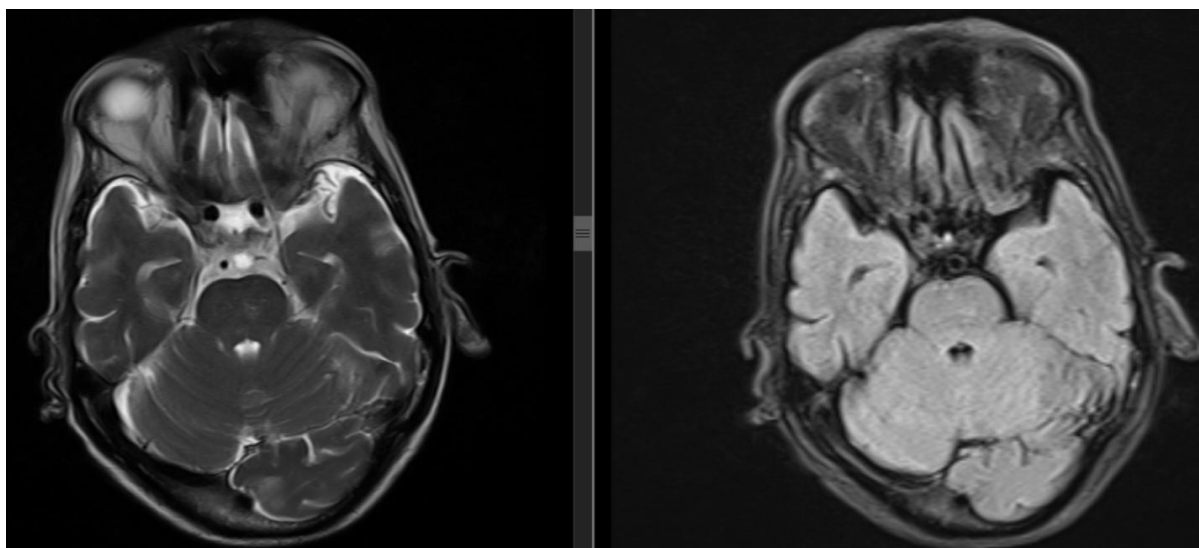
## 1. Introduction

Ecchordosis Physaliphora was first described by Hubert von Luschka in 1856 as a pathologic ectopic notochordal tissue at the posterior clivus. After the embryogenesis Ecchordosis Physaliphora arises from the remaining notochordal cells along the axis of the spine. It is a rare congenital hamartomatous lesion. It lies along the midline craniospinal axis from the clivus to the sacrococcygeal region. It is a small, gelatinous nodule, from a few millimeters to 2 cm, exhibiting a slow growth pattern. They are found intradurally in the prepontine cistern where it is attached to the dorsal wall of the clivus via a small pedicle, cartilaginous/osseous in origin. Usually asymptomatic and found incidentally in 0.4–2% of autopsies.

## 2. Case Report

66 year old male patient presented with history of headache since 6 months. He was a known case of carcinoma lung since one and half years and on treatment. No history of diplopia, altered behavior or loss of consciousness. MRI Brain was requested to look for any organic cause.

On MRI Brain, there was a well defined, thin walled, T2 hyperintense lesion in the midline in prepontine location which is suppressed on FLAIR images. No diffusion restriction on DWI. No post contrast enhancement seen. It measures approximately 1.8x2.1cm. This cystic lesion was appreciated on SPACE images.



**Figure 1:** Well defined midline retroclival intradural thin walled T2 hyperintense lesion in prepontine cistern which is suppressed on FLAIR images

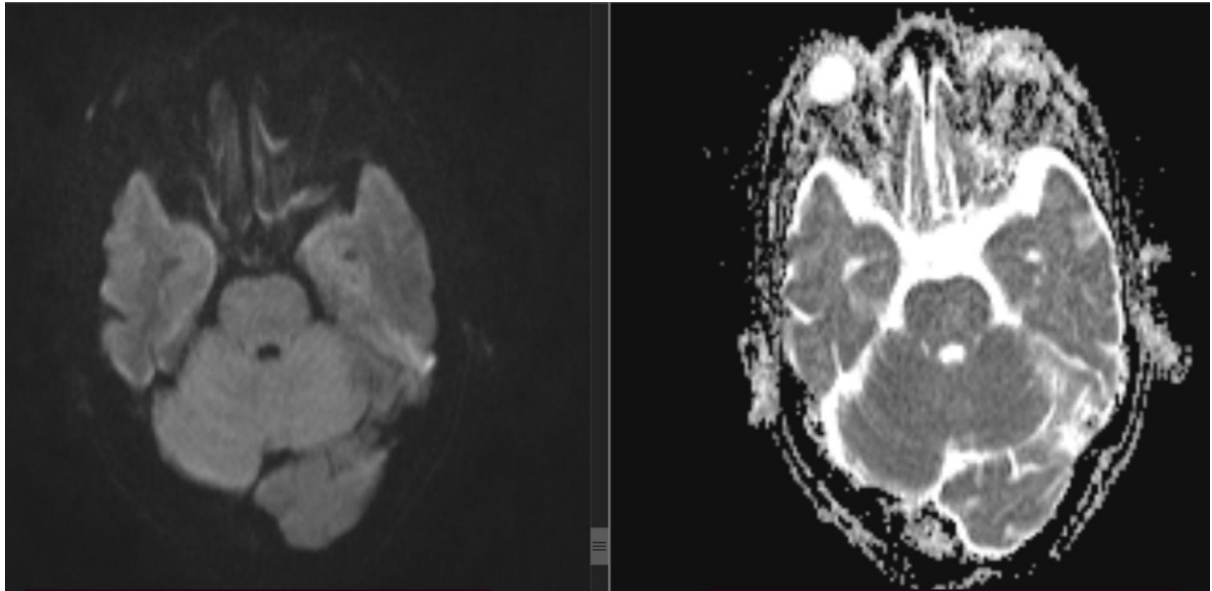


Figure 2: No diffusion restriction on DWI

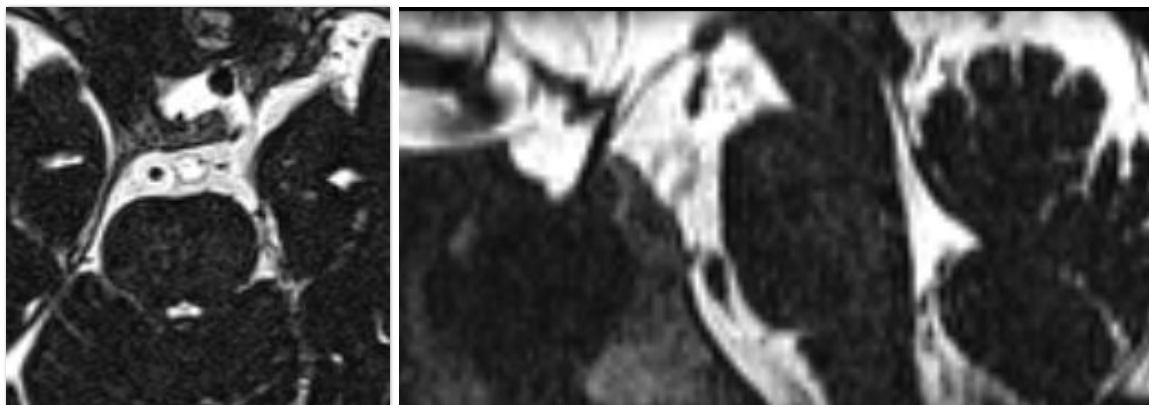


Figure 3: Well-defined thin-walled T2 hyperintense retroclival prepontine lesion with pedicle on SPACE images

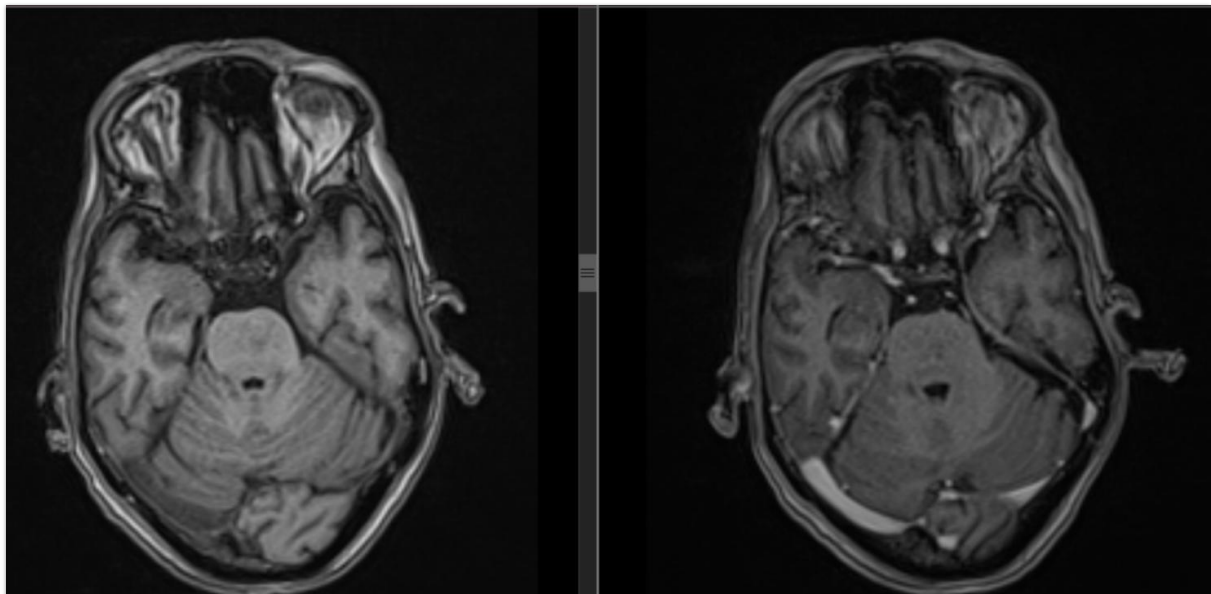


Figure 4: It is hypointense on T1 WI with no enhancement on T1 Post Contrast images

### 3. Discussion

In this patient a well defined intradural cystic lesion was seen in the retroclival location and it was attached to the clivus with a pedicle. It has following differential diagnosis:

Neuroenteric cyst: T1 hyperintense, No enhancement.

Epidermoid cyst: Diffusion restriction.

Volume 11 Issue 6, June 2022

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Arachnoid cyst: CSF intensity lesion, Imperceptible wall and no stalk.

Chordoma: Post contrast enhancement with infiltrative features.

Metastases; No post contrast enhancement, diffusion restriction or infiltrative feature.

Ecchordosis physaliphora: Cystic lesion in midline, No post contrast enhancement or infiltrative features.

Hence, Final diagnosis is Retroclival ecchordosis physaliphora.

It is typically situated intradurally and mostly attached to the dorsal clivus with a delicate pedicle. It is rarely located in the sacrococcygeal region. It is usually asymptomatic due to small size and slow growth. Rarely symptomatic, only if extra tumoral haemorrhage occurs. Histologic, immunohistochemistry and ultrastructural studies are often ineffective due to small samples. Therefore radiologists play an important role in diagnosing this rare entity.

#### 4. Conclusion

Ecchordosis physaliphora is a rare benign lesion which is managed conservatively. It has close differentials with above described lesions which are managed surgically or by radiotherapy. Therefore, before hand knowledge of this a rare entity is important for a radiologist and neurosurgeon to avoid unnecessary surgical or medical management.

#### References

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