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Outpatient

Glioependymal Cyst of Filum Terminale-A Rare Case Report

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Case Report

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Abstract: Filum terminale l cysts are rare, nonneoplastic intracranial cysts. Most of these are asymptomatic and present in the frontal and the parietal lobe. Conus medullaris and filum terminale are the rare sites for Glioependymal cysts. Here we present a rare case of glioependyma cyst of filum terminale in an 11year old female.
Keywords: Glioependymal cyst, nonneoplastic, glioependyma, female.

INTRODUCTION

Glioependymal cysts are rare, epithelial lined, nonneoplastic cysts of the central nervous system [1].Glio-ependymal cysts, also known as neuroepithelial cysts, are thought to arise from ectopic rests of primitive neuroglial tissue and hence, can arise anywhere in the neuraxis [2]. They are most commonly situated in the paraventricular white matter of frontal and parietal lobe but may also lie within the cerebellum, brain stem and spinal cord. Filum terminale is very rare site for Glio-ependymal cyst [3].

Generally, these cysts present in the second or third decade, but have been reported at all ages and even in intrauterine period [4]. Glioependymal cysts mostly are asymptomatic. They may clinically present due to mass effect and compression over surrounding neuroparenchyma. The cysts may even affect the CSF circulation system and may even cause hydrocephalus if located within the ventricular system [5].

The cyst wall has heterogenous histological appearance and is comprised of an ependymal lining with or without cilia, situated on either the basement membrane or glial tissue. Pathological diagnosis is based on lack of continuity with normal ependymal lined cavities of brain and spinal cord along with characteristic histological features [6]. Glioependymal cysts lining cells express GFAP and S100 [3].

CASE REPORT

An 11 year old female patient presented to surgery OPD with mass in the lumbo sacral region since 5-6 months. The mass was progressively increasing in size and associates with pain in the lower back. On physical examination a mass of 3x2 cm was palpable in lumbar region, which was soft to firm in consistency. On MRI there are seen two thecal sacs containing nerve fibres with a osseous spur in the sacral canal. Also seen a small T1/T2 hypointense tract in posterior left paraspinal soft tissue. Rest of the lumbar vertebrae and intervertebral disc appears normal. Lumbar curvature is maintained. Surgery was planned and filum terminale was excised which was send to pathology department GGSMC, Faridkot for histopathological examination.

We receive a grey white soft tissue piece measuring 1x1x0.5cm. Whole of the tissue has been processed. On microscopic examination a cyst was composed of inner glial layer with a luminal ependymal lining and outer fibrous layer was seen suggesting the possibility of Glioependymal cyst. (Figure 1,2). IHC is done, which shows S100 positivity in the lining epithelial cells. (Figure 3,4)

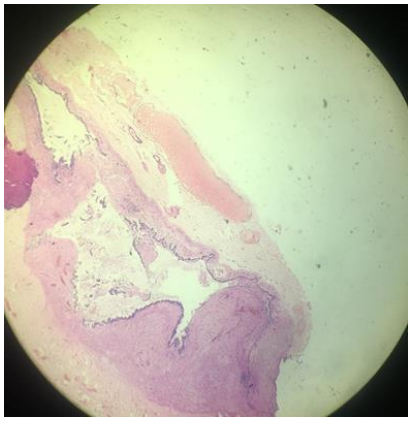

Fig-1: Microscopic Examination

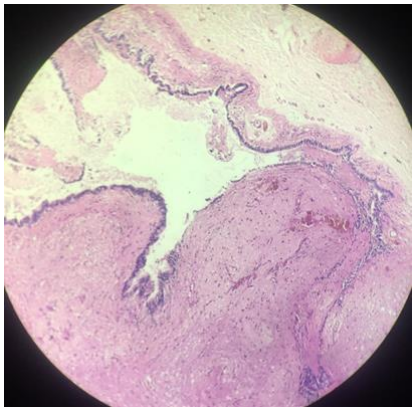

Fig-2: Microscopic Examination

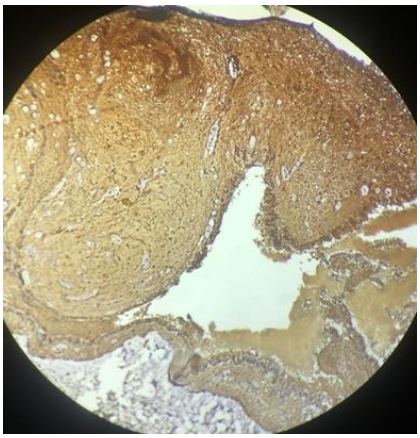

Fig-3: IHC (S100 positive)

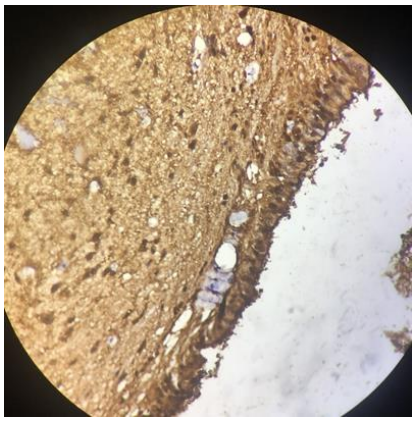

Fig-4: IHC (S100 positive)

DISCUSSION

Glioependymal cysts are rare with literature limited to case reports and small series of patients. They can be noted anywhere along the neural axis, however, when present, they are almost always seen in the frontal lobe .Filum terminale is the rare site. Various names have been used to describe ependymal lined cysts of brain including ependymal cyst, neuroepithelial cyst, epithelial cyst and choroidal epithelial cyst [7]. Friede and yasargil unify these entities under the description of glioependymal cyst [8].Glioependymal cysts accounts for less than 1% of all intracranial cysts [1].

Glioependymal cysts mostly are asymptomatic. They may clinically present due to mass effect and compression over surrounding neuroparenchyma. The cysts may even affect the CSF circulation system and may even cause hydrocephalus if located within the ventricular system. Depending on the site of cyst the patient presents clinically. If located within the intramedullary cavity, the presentation is of increased micturition [2]. If located near cerebellopontine angle, the presentation maybe in the form of compression over cranial nerves [9].

On imaging, glio-ependymal cysts appear as nonenhancing CSF containing unilocular thin-walled cysts, found both in intra-axial and extra-axial locations. They are difficult to be differentiated from arachnoid cyst, enterogenous cyst, ependymal cyst, dermoid cyst, encephalocele and congenital dysplasia of sphenoid wing in neurofibromatosis 1[4]. MR proton spectroscopy remains an inconclusive diagnostic tool till the present time; however, resonance of N-acetyl group of the compound may be a subtle indicator of glial tissue within ependymal lining [1].

The ideal treatment for these cysts is complete excision. Fenestration may sometimes result into recurrence. Cystoperitoneal shunt may also be performed. However, it requires monitoring [11].

CONCLUSION

Glioependymal cysts are rare with literature limited to case reports and small series of patients. They can be noted anywhere along the neural axis, however, when present, they are almost always seen in the frontal lobe. Filum terminale is the very rare site. We highlighted here the rare case of glioependymal cyst of filum terminale.

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