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AvailableOnlineAt:<http://valleyinternational.net/index.php/our-jou/ijmsci>**Myoepithelioma: A tale of uncommon benign spinal cord tumor presenting with spastic paraplegia**

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**Abstract:** *The current study reports the case of 3year old female child who presented in our institute with the history of weakness of both lower limbs ,bowel and bladder incontinence since 2 years. Patient was evaluated further and found to have spinal cord lesion extending from C5 to D3 vertebral levels . The patient was taken for surgery and debulking of the tumor was done .Biopsy of the lesion was proven to be myoepithelioma .The patient was followed up for the past 2 years which showed significant improvement with near normal power on both lower limbs ,increased sensation to pain below T5 dermatomes and no evidence of bowel /bladder incontinence .*

**Key words :-** *Myoepithelioma , Pleomorphic adenoma , Warthins tumour ,Dorsal myelopathy, Transverse myelitis ,MIB1 - Mind bomb E3 Ubiquitin Protein Ligase 1*

**Clinical History :-**

3 Year old female child presented with the history of weakness of both lower limbs ,bowel and bladder incontinence since 2 years .On general examination ,the child appears unremarkable .Both the lower limbs appears spastic with grade 0 power and reduced sensations to pain below T5 dermatome including the perianal region .Deep tendon reflexes appears exaggerated in both the lower limbs. Based on the clinical diagnosis, remote dorsal myelopathy and transverse myelitis was done .



**Figure -1 Plain frontal view radiograph of the cervico dorsal spine shows widening of the spinal canal.**

Imaging Features on the plain X-ray of upper cervico-dorsal spine showed widening of the lower cervical and upper dorsal spinal canal with ill-defined soft tissue attenuated opacity (1,2).

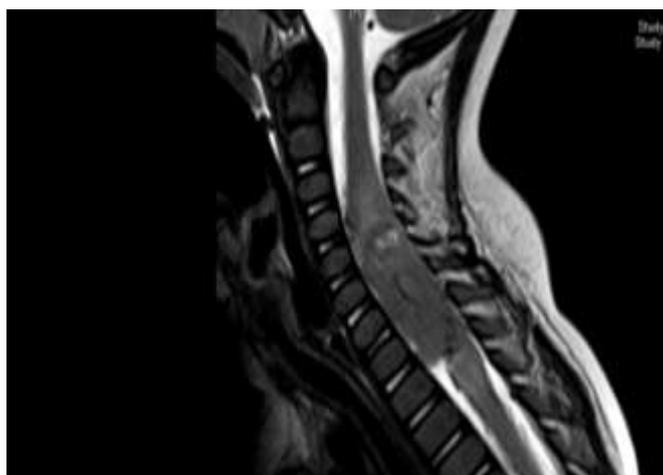
CT scan revealed ill-defined soft tissue attenuated lesion within the spinal canal from C5 to D3 vertebral levels .No bony abnormality of the spine detected.

MRI scan showed well-defined heterogenous intradural intra and extramedullary mass lesion extending from the level of C5-C6 till the level of T3 mid vertebral level.The lesion appears isointense on T1 W ,iso -hypointense on T2W with prominent vessels in and along the surface of cord both proximal and distally . No evidence of diffusion restriction noted.

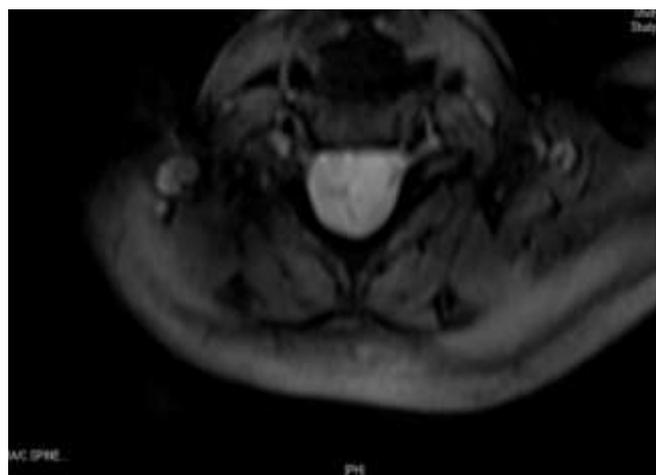
On post contrast imaging ,there is significant enhancement noted within this lesion . Based on the imaging features the differentials included are paraganglioma and ependymoma.

Patient was taken for surgery and following which debulking of the tumour was done and followed up regularly .The biopsy of the lesion revealed cervical intradural ,extra and intramedullary neoplasm with epitheloid and rhabdoid factor of uncertain malignant potential .MIB-1 labelling index of 10-15%.Finally the diagnosis of Myoepithelioma was made (3,4).

Follow up scan was done after 2 years which showed Grade 4 power on both lower limbs ,increased sensation to pain below T5 dermatomes.Deep tendon reflexes normalised in both lower limbs .No history bowel /bladder incontinence.



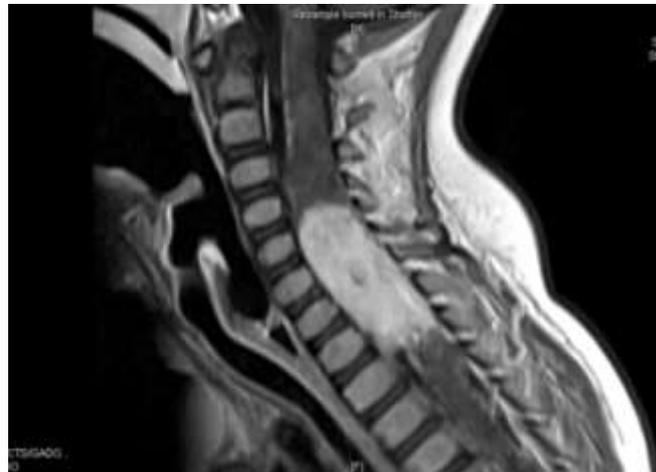
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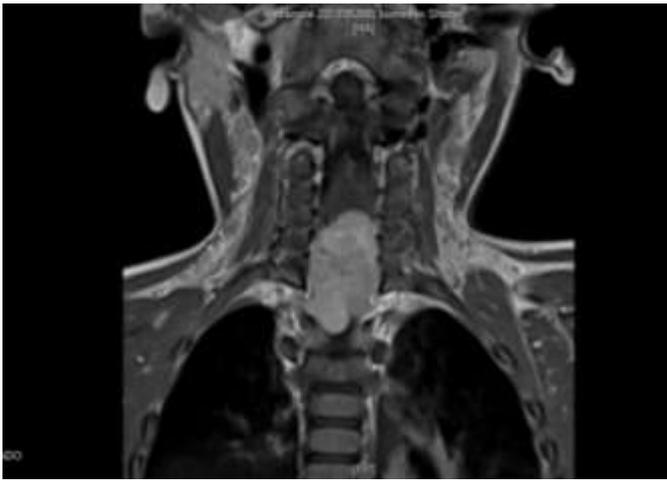
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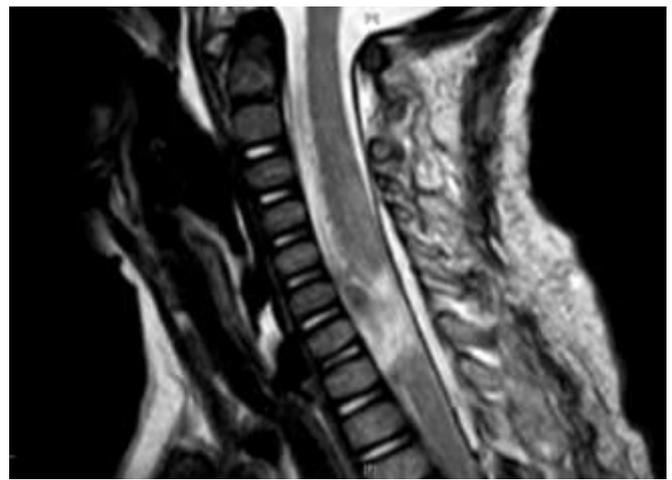
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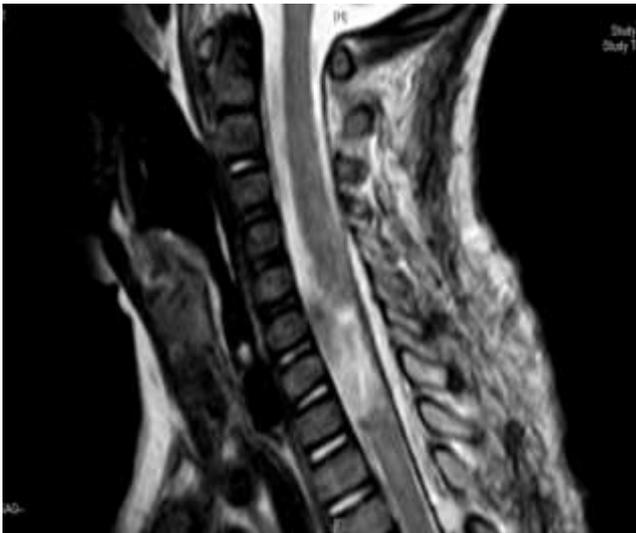


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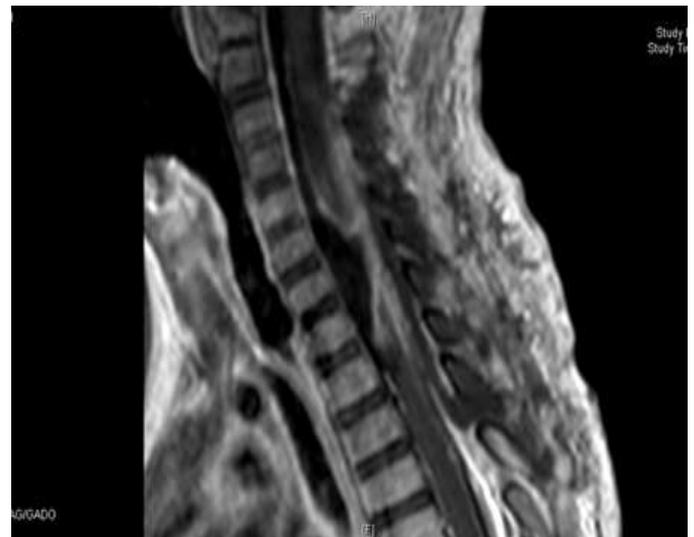


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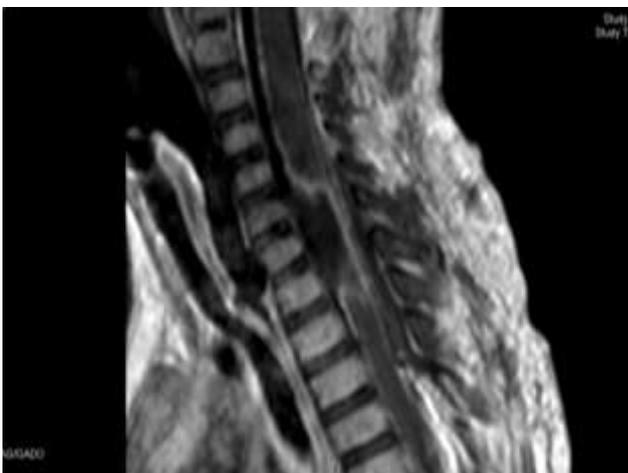
Figure 2 (a-f) Expansile intradural intramedullary and extramedullary tumour appearing isointense to cord on T1 W ,iso-hypointense with prominent vessels on T2W images .The mass shows significant enhancement on contrast .



a



c



b

Figure 3 (a-c) Follow up images showing post op defect with no evidence of residual tumour .

### Discussion :-

Myoepithelioma is a rare benign tumour . Myoepitheliomas account for 1.5% of all of the tumors in the major and minor salivary glands . Nearly 40% arise in the parotid glands. These are benign tumour showing myoepithelial differentiation.Clinical course is variant with lymph node metastasis at presentation .23.5% shows evidence of local recurrence with low malignant potential .Some of these lesions are aggressive (5,6).The use of CT for the assessment of parotid tumors is well established. Although much has been written on the imaging appearance of the

pleomorphic adenomas and Warthin tumors ,the detailed imaging features of myoepitheliomas of the salivary glands have not been much described in the literature(7,8).Diagnosis is based on the infiltrative nature and presence of dual glandular secretory and myoepithelial differentiation(8,9).

On CT ,these lesions are hypo- to iso-dense , well-demarcated with irregular in outline with marked enhancement on post contrast images .On MRI ,these lesions are iso-intense on T1 w images ,intermixed intensity on T2 w images and show marked enhancement on post contrast images .

### Conclusion :-

Spinal myoepithelioma should also be considered as a differentials when we see an intradural mass with extra and intramedullary component. These are rare benign tumors of the major and minor salivary glands. Timely diagnosis will help the patient in early management and recovery.

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