

# Conus Hemangioblastoma with Holocord Syrinx not Associated with von-Hippel Lindau (vHL) Syndrome: A Case Report

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## ABSTRACT

We present a case of 40-year-old male who presented with backache, numbness and gradually progressive weakness of lower limbs of 6 months duration. Magnetic resonance imaging (MRI) revealed a homogeneously enhancing tumor in the conus with holocord syrinx suggestive of a hemangioblastoma. Hemangioblastoma of the conus medullaris is a rare pathology especially when it is not associated with vHL syndrome. Here we discuss surgical nuances of operating upon such a rare pathology and briefly review the literature.

**Keywords:** Conus medullaris tumor, Holocord syrinx, Spinal hemangioblastoma.

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**Conflict of interest:** None

Hemangioblastomas are benign tumors of the central nervous system which is most commonly seen in the cerebellum.<sup>1</sup> Location in the conus of the spinal cord is uncommon.<sup>2</sup> Here we present a case of a conus medullaris Hemangioblastomas with holocord syrinx not associated with vHL syndrome.

A 40-year-old male presented with a backache, numbness, burning sensation and gradually progressive weakness of lower limbs of 6 months duration. He also complained of difficulty in passing urine and constipation for the last 2 months. He had been diagnosed with pulmonary Koch's one and half years ago and had taken treatment for the same.

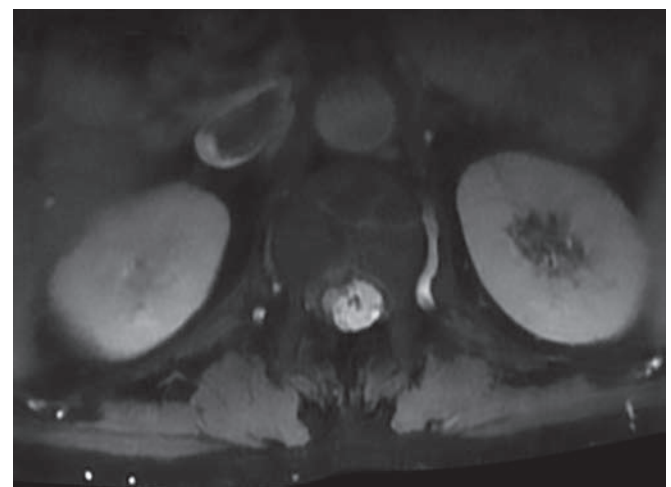
On examination, he had wasting of right lower limb and bilateral spastic paraparesis. Power in the right lower limb was 2/5 and that in left lower limb was 1/5. Power in right upper limb was 4+/5 and left upper limb was

5/5. Deep tendon reflexes were exaggerated in the lower limbs. Bilateral ankle and patellar clonus were present, and plantars were extensor. Graded sensory loss was present up to D9.

Routine blood investigations were normal. MRI revealed intramedullary intensely enhancing lesion in the conus region (Figs 1 and 2) with holocord syrinx from C2 to D11 (Fig 3). The lesion was hypointense on T1 and hyperintense on T2. Work up was done to rule out vHL syndrome. Ultra-sonogram (USG) abdomen was normal. No lesion was seen in retina on fundus examination.



**Fig. 1:** Sagittal MRI image showing intensely contrast enhancing lesion at D12-L1 level



**Fig. 2:** Axial MRI image showing intramedullary lesion at the level of conus

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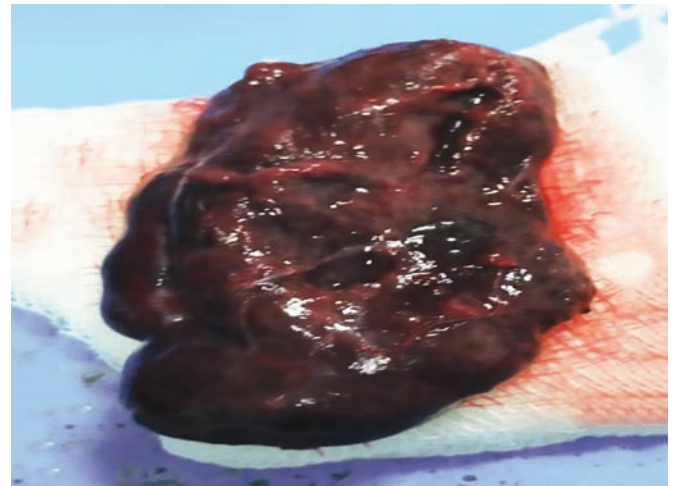
**Fig. 3:** T2 weighted sagittal MRI showing holocord syrinx at cervical, thoracic, lumbar region.

The patient was positioned in a prone position on Wilson frame. D12-L1 level was confirmed using image intensifier. After parts were painted and draped, vertical midline exposure was done at the desired level. Paraspinal muscles were separately subperiosteally and were retracted laterally using self-retaining retractors. D12-L1 laminectomy was then done. Vertical durotomy was made in the midline dorsal spinal cord under a microscope. Dural flaps were retracted laterally using sutures. Myelotomy was done in the midline using bipolar cautery at low power. The tumor was cherry red, vascular surrounded by prominent vessels (Fig. 4). Bipolar cautery at low power was used to coagulate the dominant arterial feeder followed by coagulation of a tumor to shrink it. The venous drainage was coagulated the last. The tumor was excised in en-bloc (Fig. 5). No attempt was made to excise the tumor piecemeal due to risk of heavy blood loss. Syrinx was not opened as these tend to resolve spontaneously.

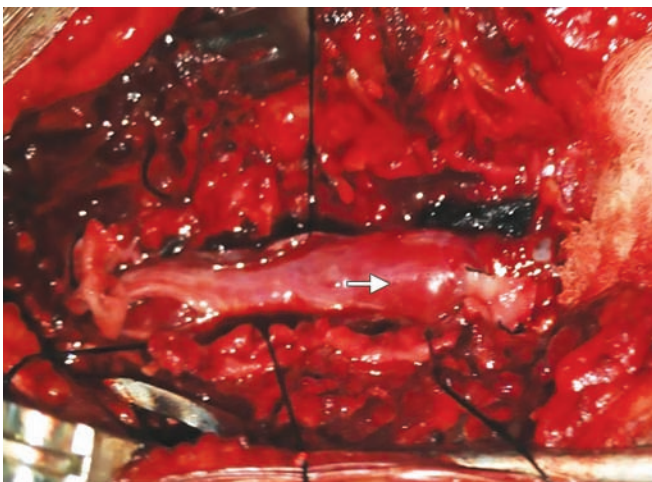
Postoperative period was uneventful There was no deterioration in the neurological status of the patient. The patient was lost to follow-up. Histopathology revealed a benign lesion consisting of stromal cells containing

foamy cytoplasm suggestive of hemangioblastoma (WHO grade 1) (Fig. 6).

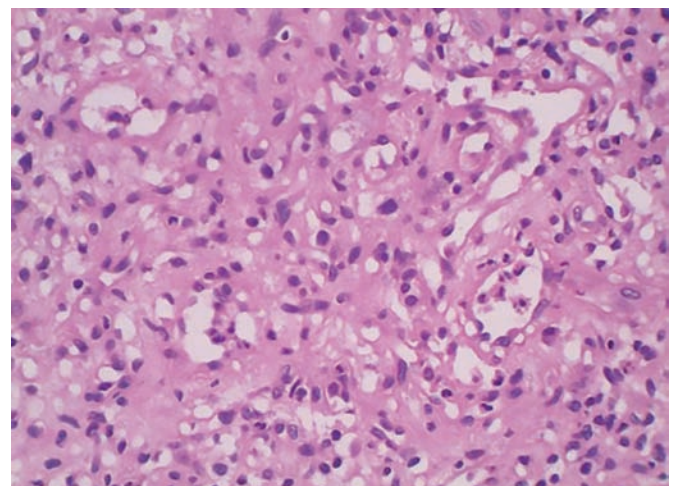
Hemangioblastomas are rare, benign and highly vascular tumors.<sup>1,3</sup> They usually occur in the cerebellum or retina as sporadic lesions and less commonly in the medulla or the spinal cord.<sup>4</sup> Incidence of spinal hemag-



**Fig. 5 :** Image showing the excised in toto specimen



**Fig. 4:** Intraoperative image showing cherry red vascular lesion



**Fig. 6:** Histopathology slide showing benign lesion with stromal cells containing foamy cytoplasm and hyperchromatic nuclei with large vascular spaces of various sizes suggestive of a hemangioblastoma

ioblastomas has been reported to be 1-5% of all spinal cord tumors.<sup>4</sup> They tend to be multiple when associated with vHL syndrome.<sup>1</sup> vHL syndrome is a heredo-familial autosomal dominant disease with incomplete penetrance. These tumors usually, occur in cervical or the thoracic region and location in the conus has rarely been reported earlier.<sup>2</sup>

Tumors in the conus usually present with back pain, sciatica and bladder bowel symptoms. In some cases, urinary symptoms may be the sole manifestation.<sup>3</sup>

MRI is imaging modality of choice which reveals low signal intensity on T1 weighted images with high signal intensity for cyst in T2 weighted images.<sup>4</sup> On Gadolinium-diethylene triamine penta acetic acid (Gd-DTPA) the lesion enhances brightly. Angiography may be done for visualization of vascular supply and to perform pre op embolisation.<sup>5</sup>

While hemangioblastomas are benign tumors (WHO grade I), they cause significant morbidity due to edema, extension into a cord or large cyst formation. In asymptomatic cases, the patient may be managed conservatively but symptomatic cases need to be operated upon. Hemangioblastomas usually have a well-defined plane of dissection and can be completely removed if situated posteriorly. Tumors with extension into the anterior part of the spinal cord may not be amenable to complete resection.<sup>4</sup>

Syrinx formation is seen with intramedullary spinal cord tumors such as ependymomas and hemangioblastomas. According to most authors, hemangioblastomas are associated with a syrinx in more than 50% of cases but presence of a holocord syrinx with hemangioblastoma is

rare.<sup>4</sup> Transudation of fluid from the pathological tumor blood vessels and obstruction to flow of cerebro-spinal fluid (CSF) is believed to be the mechanism for formation of syrinx.<sup>2</sup> It is believed that higher the tumor, greater is the incidence of syrinx. The presence of syrinx is associated with higher rates of resection and better recovery in postoperative period.<sup>2</sup> Syrinx usually resolves following excision of tumor and syringe-subarachnoid shunt is unnecessary.<sup>2</sup>

Hemangioblastoma of the conus medullaris with holocord syrinx is a rare entity. The tumor must be excised en-bloc to minimize blood loss. The associated syrinx usually resolves spontaneously.

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