Spinal extradural Masson's tumor : Report of a rare case

Gopal Raman Sharma[1,2], Alok Deo[1], Maya Bhattachan[1], Samir Aryal[1], Ram Chandra Shrestha[1], Sumit Joshi[1,2], Damber Bikram Shah[1,2], Rakesh Pathak[3]

[1]Department of Neurosurgery, Dirghayu Neuro Hospital and Allied Sciences, Chabahil, Kathmandu, Nepal

[2]Department of Neurosciences, Nepal Mediciti, Bhaisepati, Lalitpur, Nepal

[3]Department of Pathology, Dirghayu Neuro Hospital and Allied Sciences, Chabahil, Kathmandu, Nepal

ABSTRACT

Extradural spinal Masson's tumor (MT) or intravascular papillary endothelial hyperplasia (IPEH) is one of the very rare spinal pathologies and only few cases have been reported in the literature. Here, we present a 43year young male patient having spinal extradural Masson's tumor at L3 spinal level. We review literature and discuss on its epidemiology, clinical scenario, diagnosis, management and outcome of this rare spinal pathology.

KEY WORDS

Extradural, Hemangioma, IPEH, Intravascular papillary endothelial hyperplasia, Masson's tumor, Spinal, Vegetate intravascular hemangioendothelioma.

*Corresponding Author |
Dr. Gopal Raman Sharma
Department of Neuroscienes
Nepal Mediciti, Bhaisepati , Lalitpur, Nepal
Mail address: gopal47@hotmail.com
gopalramansharma@gmail.com

INTRODUCTION

Masson's tumor (MT) is one of the rare neurological diseases of central nervous systems (CNS). It is also known as intravascular papillary endothelial hyperplasia (IPEH) or Masson's vegetated hemangioendothelioma. In fact, MT is not a tumor, it is a kind of pseudo tumor. It actually develops from reactive papillary proliferation of endothelial cells within blood vessels like vein, artery, vascular malformation and often accompanied by thrombus formation. Spinal MT is very rare and up to date, only 14 cases have been described in literature1-14. This case would be the 15th case of spinal MT and 4th case as secondary spinal extradural MT in world literature.

CASE REPORT

: A 43year old male, right-handed gentleman attended to our Neurosurgery OPD with history of low back pain for last 3 years. Pain was progressive and increasing in intensity. Later on, pain was radiating to both lower limbs which made him very difficult to walk. He used to stop for a moment after walking for a distance of 10 – 15 minutes due neurogenic claudication of both calf muscles. There was no history of incontinence of urine and stool. Clinical examination revealed local tenderness over the lumbar region with straight leg raising test (SLRT) of 60 degrees on both sides. There were no neurological deficits and sphincters involvement. CBC and ESR were within normal range, CRP was negative. Urine for Bence Zone's protein was negative, and other baseline laboratory tests were within normal limits. MRI of the thoracolumbar spine showed L3 vertebral body hemangioma with ventral extradural mass compressing the thecal sac and there was no evidence of instability (Figure.1). We decided go for laminectomy and removal of the extradural mass. A standard laminectomy of L2 to L4 was performed and biopsy was taken. Total excision of mass was not possible because of its high vascularity and tough consistency. After proper hemostasis wound was closed in anatomical layers. Postoperatively patient developed paraparesis of grade IV without sphincters involvement. Histological features were in favor of hemangioma (Figure.2). Immunohistochemistry (IHC) was suggestive of MT or IPEH with CD31, CD34, and ERG immunoreactivity (Figure. 3). Since the second attempt to remove this extradural mass was not possible due to its toughness and high vascularity, we recommended for a course of local radiation. On one month's followed up his neurological function was improved and able walk without support.

DISCUSSION

MT is not a tumor, actually a misnomer. it is a benign vascular mass characterized by the proliferation of endothelial cells within blood vessels and may be associated with organized thrombus17. MT is rare pathology commonly found in skin and subcutaneous tissues13,15,16. There are few anecdotal case reports of involvment of other organs of the body like renal, thyroids, lungs etc4. Occurrence of this disease in brain and spine is very rare12. MT involving the spine is very rare1-14.

MT was named after Dr. Pierre Masson, a Canadian pathologist who first described this rare pathology in 1923 as 'Hemangioedothelime Vegetant Intravasculaire'17. Afterwards, various terminologies have been used for this lesion such as, intravenous atypical vascular proliferation, pseudo angiosarcoma, intravascular angiomatosis and intravascular endothelial proliferation. In 1976 Clearkin and Enzinger used the terminology "Intravascular papillary endothelial hyperplasia (IPEH)" for this pathology18.

Actual pathogenesis of MT is not clear however, hypothesis is that the trigger may be the formation of a thrombus due to trauma, infection and radiation, followed by reactive proliferation of vascular endothelial cells19.

There are three varieties of MT: a) primary or type I (when it grows inside a normal intravascular space), b) Secondary or type II (when it grows within a preexisting vascular malformation like hemangioma, AVM and c) type III or undetermined (when it develops in extravascular hematoma)16,18,19. This type III is extremely rare and only one case has been notified in literature11.

Fourteen cases of Spinal MT have been reported in literature1-14 (Table.1). Age involved were usually young and male was predominant in those 14 cases. Clinical presentations are back and radicular pain, progressive neurological deficit, sensory impairment and bowel bladder involvement. Most of these clinical features are of progressive in nature.

Diagnostic test for MT is MRI of spines and the lesion is characterized by isointense on T1W1 sequences, hyperintense signal on T2 W2 images and homogenous enhancement on IV Gadolinium contrast. Often, CT scan of spines might be helpful to identify the involvement of posterior elements of spines. Most of these lesions are extradural compressing the thecal sac and may be associated with vertebral hemangioma 6,13,14. Rarely lesion may be located intradural and involving cauda equina 11.

In those 14 reported cases of Spinal MT commonly involved spines were thoracic (8), thoracolumbar (2), lumber (2), cervicothoracic (1) and multilevel (1). Majority of these lesions were located at extradural (12), one was intradural and in one case location was not described. Similarly 9 cases were primary, 3 cases secondary, one case undetermined and one case was not documented (Table.1).

Our case represents a secondary, extradural lumber Masson's

tumor which ranked by number 4 as a secondary extradural spinal IPEH.

Differential diagnosis of spinal MT includes schwannomas, ependymomas, tubercular granulomas and metastatic lesions.

Final diagnosis of MT is based on histopathological and immunohistochemistry features. Presence of dilated vascular space containing organized thrombus and reactive endothelial proliferation in a papillary pattern are the cardinal histological features16. Distinctive features of immunohistochemistry are the cells showing immunopositivity for CD31 and CD34, which are the markers for endothelial origin 8,11.

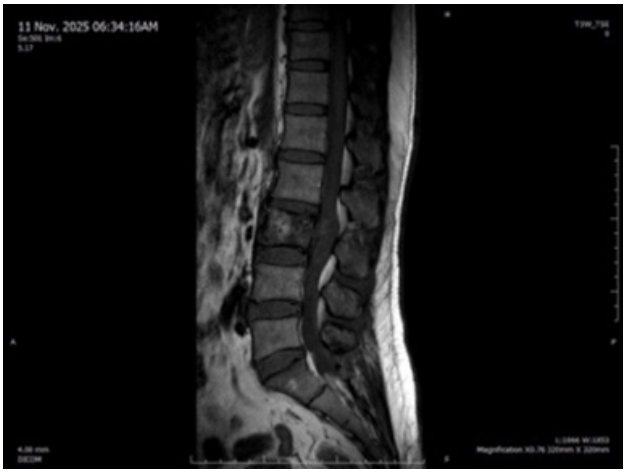
Surgery is the first line treatment if the patient is symptomatic. Aim of the surgery should be complete excision of the tumor after laminectomy/Laminoplasty with preservation of neural elements. Posterolateral instrumentation may be necessary if radiological features are suggestive of instability of spines. Out of 14 cases documented in literature 11 (79 %) patients had complete excision, 2(14%) had incomplete excision and 1(7%) was conservatively treated (Table.1). If complete removal is not possible, a course of local radiation like Stereotactic Radiosurgery (SRS), Gamma Knife (GK), Cyber knife (CN) may be considered to minimize recurrence 7. Preoperative embolization may be carried out to minimize blood loss during resection of MT 7. Beta- adrenergic antagonist has been used previously for skin MT, however it has not been documented as a treatment of choice for spinal MT15.

Long term followed up and result after the treatment of these spinal MT are not available at present. Recurrence of MT of spine is uncommon if completely excised however it may recur If the lesion is partially excised or if it arises in a primary subtype of vascular lesion16.

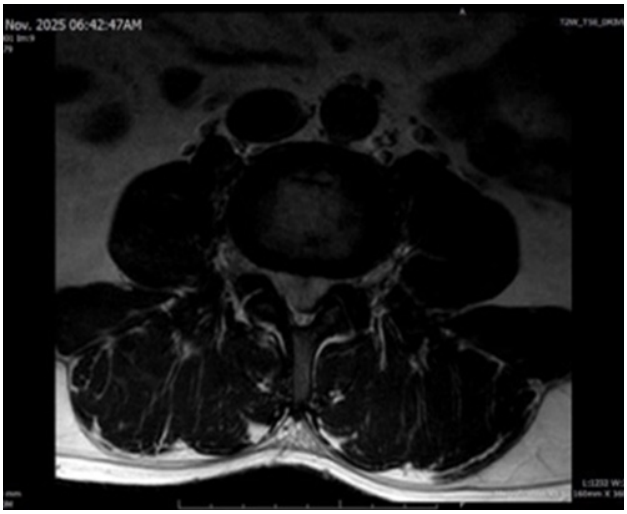
CONCLUSION

Spinal extradural MT is very rare pathology and they are benign vascular lesion. Complete excision should be the goal preserving adjacent neural element. Posterolateral fixation should be considered along with excision of MT if vertebral columns are unstable. Local radiotherapy should be considered for incomplete excision and recurrent.

Figure: 1. MRI of thoracolumbar spines



1a



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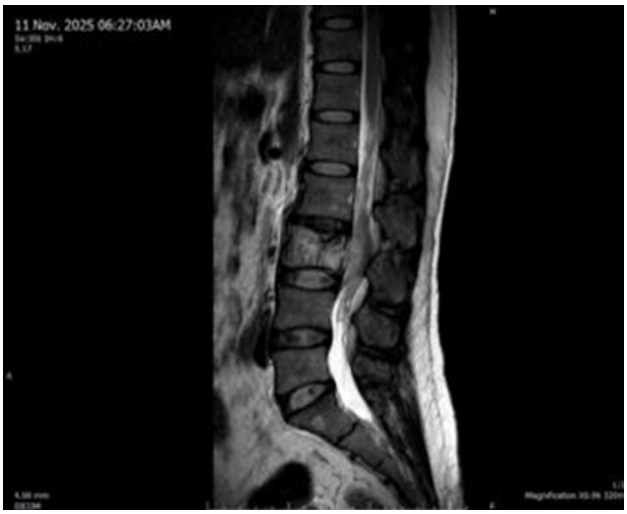

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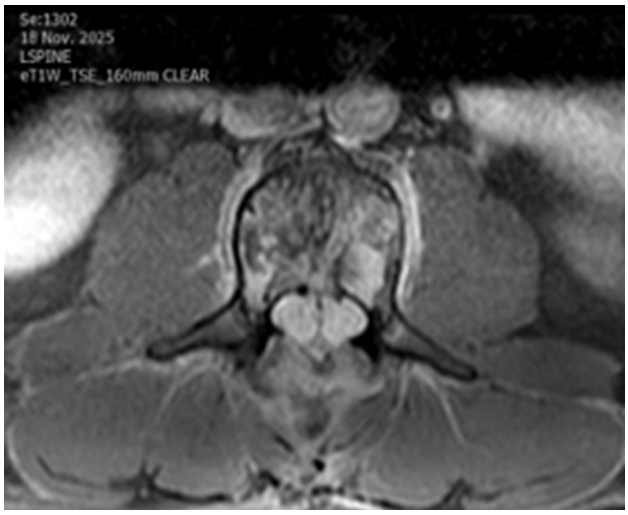
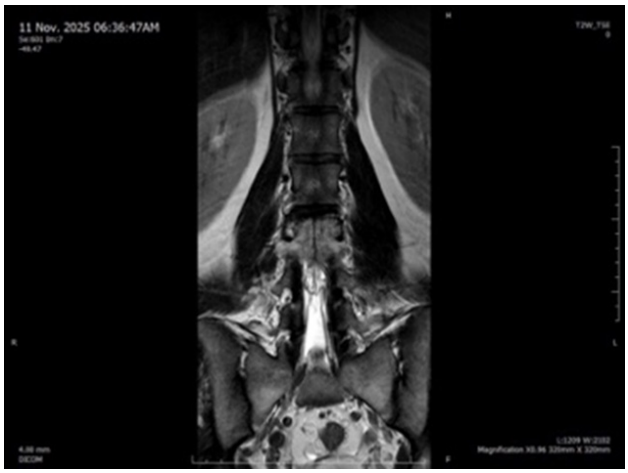


1d



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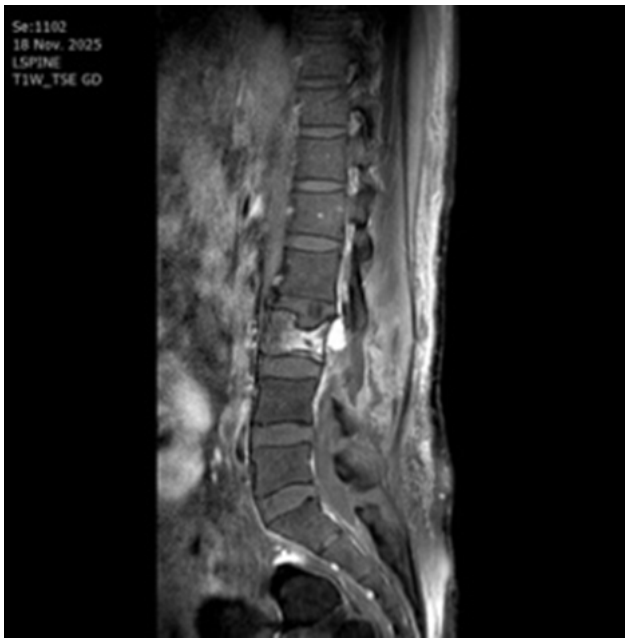

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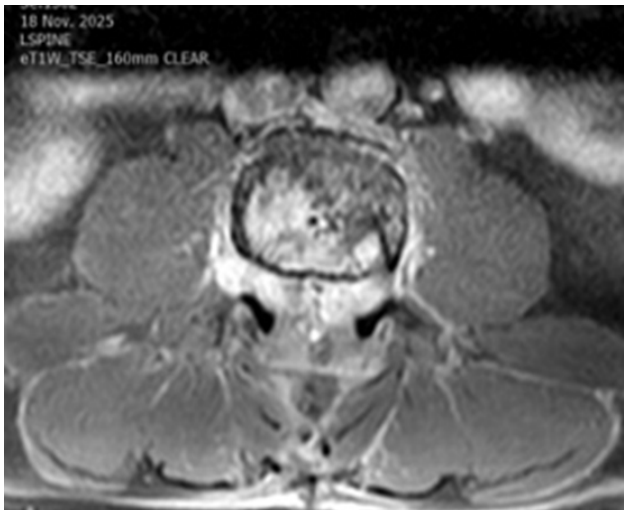
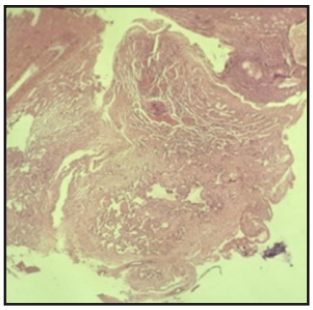


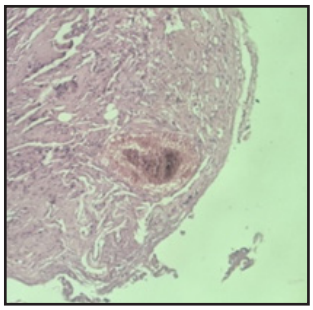
1g

Fig. 1a, b, c, d, e, f & g: MRI of thoracolumbar spines (sagittal, axial and coronal views) depicting ventral extradural mass causing severe canal compression with underlying vertebral hemangioma at L3 vertebral level.The lesion is hypointense on T1W1, isointense on T2W2 images and homogenously enhanced on IV contrast.

Figure: 2. Histology



2a



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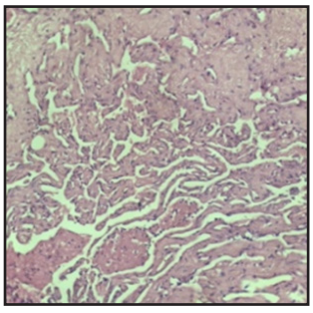

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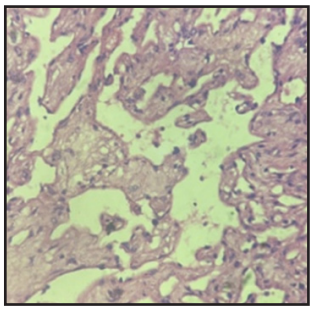
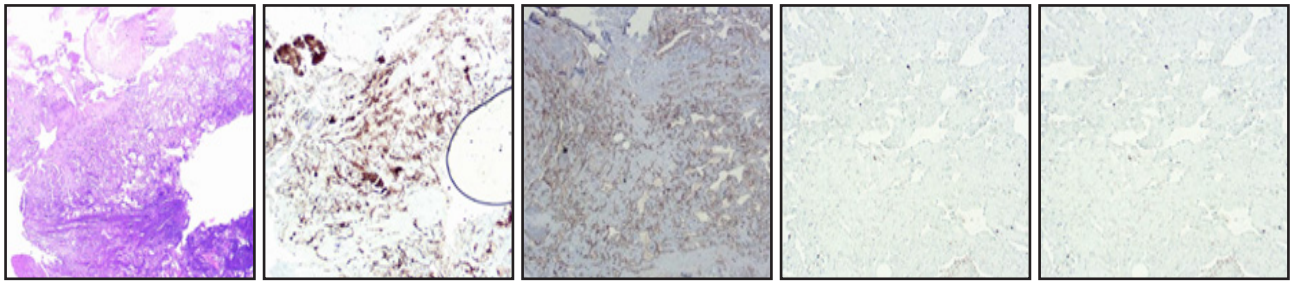


2d

Fig. 2 a, b, c & d: Well -formed and organized Papillae covered by single layer of flat endothelial cells projecting into the lumen of blood vessels associated with thrombosed blood vessel are classical histological pictures of Masson's tumor

Figure: 3. Immunohistochemistry (IHC)



3a.H&E **3b.CD31** **3c.CD34** **3d.ERG** **3e.Ki67**

Fig.3a, b, c, d, e: IHC revealed immunoreactivity for CD31, CD34 and ERG. Ki67 was immunoreactive for 2-3% of tumor cells. These IHC features are suggestive of Masson's tumor

Table no. 1 Reported cases of spinal Masson's tumors in the literature (till to date)

No	Authors/ Years	Age/ Sex	Clinical presentation	Radiological features	Subtypes	Immunohisto-chemistry (IHC)	Treatment	Outcome
1	Ali SZ et al 1994[1]	42 /M	Paraplegia	MRI: nonspecific T1/T2 signal changes. CT myelography- extradural mass	Primary (Type I)	N/A	Radical excision	N/A
2	Porter DG et al 1995[2]	16/ M	Mid thoracic pain with radiculopathy	CT myelography: extradural mass	Primary (Type I)	N/A	T6 laminectomy with T5/ T6 right partial facetectomy	N/A
3	Taricco MA et al 1999[3]	17/M	Left lower limb mono paresis with bladder dysfunction	Contract CT scan: hyperdense lesion MRI with IV contrast: T1 - iso, T2- hyperintense with homogenous contrast enhancement	Primary (Type I)	N/A	Laminectomy and radical excision of mass	:Pain relieved :Neurological function recovered
4	Petry M et al 2009[4]	47 /M	Diffuse low back pain	MRI: T1- Isointense and T2- hyperintense lesion	Primary (Type I)	N/A	No surgery	N/A
5	Lanotte M et al 2010[5]	33/M	Back pain with paraparesis	MRI: T1 – hypo and T2 – hyperintense epidural mass	Primary (Type I)	N/A	Laminectomy and excision of mass	Improved
6	Mozhdehipanah H et al 2013[6]	50/M	Spastic para paresis with sensory deficits	MRI: T2 – hyperintense mass	Secondary (Type II)	N/A	Laminectomy + Radical excision	Improved
7	Bhalla N at al 2013[7]	41/ M	Back pain with paraparesis	MRI: L1 spinous process and pedicles involvement with compression of cauda equina	Primary (Type I)	N/A	: Preop embolization :Incomplete excision :Radtiotherapy	Remained well
8	Singla N et al 2016[8]	40/M	Back pain with right lower limb numbness	MRI: Dumbell shaped mass	Primary (Type I)	Immunopositivity for CD31, CD34	Radical excision	Excellent recovery
9	Mann P et al 2016[9]	55/ M	?	T7-T10 extradural mass	?	N/A	?	N/A

10	Behera BR et al 2017[10]	32/M	Paraplegia	MRI: T1- hypointense , T2- hyperintense epidural mass at T4-T5 level	Primary (Type I)	N/A	Radical excision	Good improvement
11	Tanaka M et al 2018[11]	40/M	Low back pain with legs pain	MRI: hypointense with partial area of high intensity without contrast enhanced intradural mass at L2-L3 level	Undetermined (Type III)	Immunopositivity for CD31, CD34	Radical excision	improved
12	Oktar N et al 2019[12]	37/M	Right lower limb paresthesia with monoparesis	MRI: Dumbell shaped extradural mass at T4-T5 level	Primary (Type I)	N/A	Radical excision	Symptom free at 6 months
13	Gu HL et al 2021[13]	27/M	Neck pain and weakness of the extremities	MRI: T1 hypo and T2 hyperintense at C6-T1 level extradural mass with hemangioma contrast enhancement	Secondary (Type II)	N/A	Laminectomy & Radial excision	Symptom free at 6 months
14	Kostic A et al 2024 [14]	67/ M	Progressive paraparesis	MRI: T5-T6 bilateral epidural mass with spinal card compression	Secondary (Type II)	N/A	Total excision of mass with sec. stabilization	Gradual neurological improvement
15	Present case report	43/M	Low back pain with lower limbs paresthesia	MRI: T1 hypointense and T2 hyperintense extradural mass with enhancement at L3 level	Secondary (Type II)	Immunopositivity for CD31 and CD34	: Laminectomy and subtotal excision : Local radiation	Symptom free at one month

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