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Case Reports & Case Series

Impetuous expansion of pure epidural capillary hemangioma in pregnancy: A rare cause of acute paraplegia

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ABSTRACT

Background: Purely epidural spinal hemangioma (PESH) represents a rare clinical entity. The majority of such reports focuses on the cavernous variety and the capillary type remains extremely rare. A minority of these lesions may cause acute clinical presentations, mostly from hemorrhage or less commonly due to tumor expansion. Acute paraplegia in pregnancy from PESH is extremely rare and may pose management dilemma.

Case report: A 25-year old pregnant lady, 34 weeks into her pregnancy, presented with one-week history of paraplegia with bladder bowel involvement. On MRI, a dorsal epidural contrast enhancing mass with transforaminal extension was noted at D10–11 level. An emergency caesarean section (CS) to deliver the baby was followed by tumor excision after 48 h. The patient gradually recovered and regained 3/5 power in her lower limbs at 6 months follow up.

Conclusion: PESH of capillary variety is a rare cause of acute paraplegia in pregnancy. Preoperative suspicion of this diagnosis is extremely unlikely and regardless of the timing of surgery, such acute paraplegias rarely improve completely. Fetal lung maturity is an important factor determining the surgical priority.

1. Introduction

Vertebral hemangiomas of spine are not rare. However, a minority of these tumors, the so called aggressive hemangiomas, would extend beyond the vertebral body and may compress the spinal cord [1]. The epidural tumor extensions in such cases are usually secondary to the vertebral involvement. There is however, a specific group of spinal hemangiomas that occur exclusively in the epidural space, without any involvement of the vertebral body. These so called purely epidural spinal hemangiomas (PESH) are almost always of cavernous variety [2,3]. There are > 100 reported cases of cavernous epidural spinal hemangiomas, however the capillary variety of PESH is much more uncommon. To best of our knowledge, ours is the thirteenth case reported in English literature.

Pregnancy is a state of physiologic hypervolemia. Moreover, there is an increase in serum progesterone during pregnancy. A combination of these two may lead to expansions/of vascular lesions/hypervascularized tumors circulation and sometimes occurrence of de-novo lesions [2,8]. Instances of acute paraplegia during pregnancy in such situations are not uncommonly reported.

Our literature review revealed two previous reports of PESH causing paraplegia during pregnancy. Moreover, only five cases of pure capillary hemangioma has been reported before in females, that too including a known case of endometriosis (Table 1). We present an interesting case of PESH of capillary variety showing impetuous growth and acute paraplegia in a pregnant lady. The pathophysiology, unique radiologic findings and management dilemmas of this rare condition are discussed with a review of literature.

2. Illustrative case

2.1. Clinical course

A 25-year lady, 34 weeks into her first pregnancy, was referred to our emergency department with acute onset low back ache for 1 week associated with numbness and profound lower limb weakness. This was accompanied by acute retention of urine and faecal incontinence for the same duration. There was no contributory past history.

On examination, she had spastic paraplegia (MRC grade 0/5), exaggerated lower limb deep tendon reflexes and a complete sensory loss to all modalities below D12 dermatome. There was no spinal tenderness or deformity. The clinical picture, at this point, indicated a diagnosis of

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Table 1
Previous reports of pure epidural capillary hemangioma reported in literature.

Author (year)	Age/gender	Presentation	Level	Foraminal extension	Histopathology	Excision
Gupta et al. [5] (1996)	50/M	3 months	T8–T10	Yes	Sheets of variably sized endothelium-lined vascular spaces within a fibrous stroma	Not mentioned
Badinand et al. [9] (2003)	40/F	1 year progressive (known case of endometriosis)	T2–T4	Yes	Composed of thin and irregular capillary vessels caught in low-attenuating fibrosis	Partial
JS Kang et al. [20] (2006)	56/M		T3–T4	Yes	Sheets of variably sized endothelium-lined vascular spaces within a fibrous stroma	Partial (intra thoracic segment was left behind)
Vassal et al. [18] (2011)	59/F	Progressive weakness and pain	T5–T7	Yes	–	Total
Hasan et al. [11] (2011)	57/M	2.5 years with worsening pain since for last 5 months	T12-L2	Yes	Composed of vessels of various calibres. The walls were lined by endothelium	Total
Seferi et al. [16] (2014)	58/M	3 months progressive weakness and pain	T2–T4	Yes	Vessel walls were internally lined from endothelium with a total lack of smooth muscular elements; continuous basal lamina participated in the lobular architecture of the entire structure.	Total
Gencpinar et al. [21] (2014)	17 months/ Female	Acute presentation where pain severity increased within a day	T3 to T7	No	Proliferation of capillary vascular channels.	Total
GP Ma et al. [22] (2015)	67/F	Progressive weakness and pain in 20 days	T4–T5	No	Fibrofatty tissue with a proliferation of vascular structures, which were generally of small size, seating areas with myxoid appearance. No cellular atypia was seen.	En bloc
Egu et al. [17] (2016)	60/F	Progressively increasing pain	L5-S1	Yes		Total
Garg et al. [14] (2016)	50/M	Sudden to start but then progressive in nature for 3 months	T5–T6	Yes	Fibrocollagenous and fibroadipose tissue showing a dilated irregular vascular channel lined by flattened epithelium separated by stroma	En bloc
Tunthanathip et al. [15] (2017)	15/M	4 months coccydynia	Conus lesion	No	A lobular architecture, with the lobules being composed of tightly packed capillary-sized vessels lined by a single layer of endothelial cells. Immunohistochemical staining that positive for CD34 and factor 8	Total
Rajeev et al. [13] (2017)	50/M	1 year with rapid a progression in 1 month	T12-L2	Yes	Composed of variable calibre blood vessels predominantly thin walled	Total
Present case	25/F	Acute onset painful paraparesis	D10–D11	Yes	Lobules of thin walled spaces lined by flat endothelium with large areas of hemorrhage and necrosis. The stroma showed presence of mild, mononuclear lymphocytic infiltrate	Total

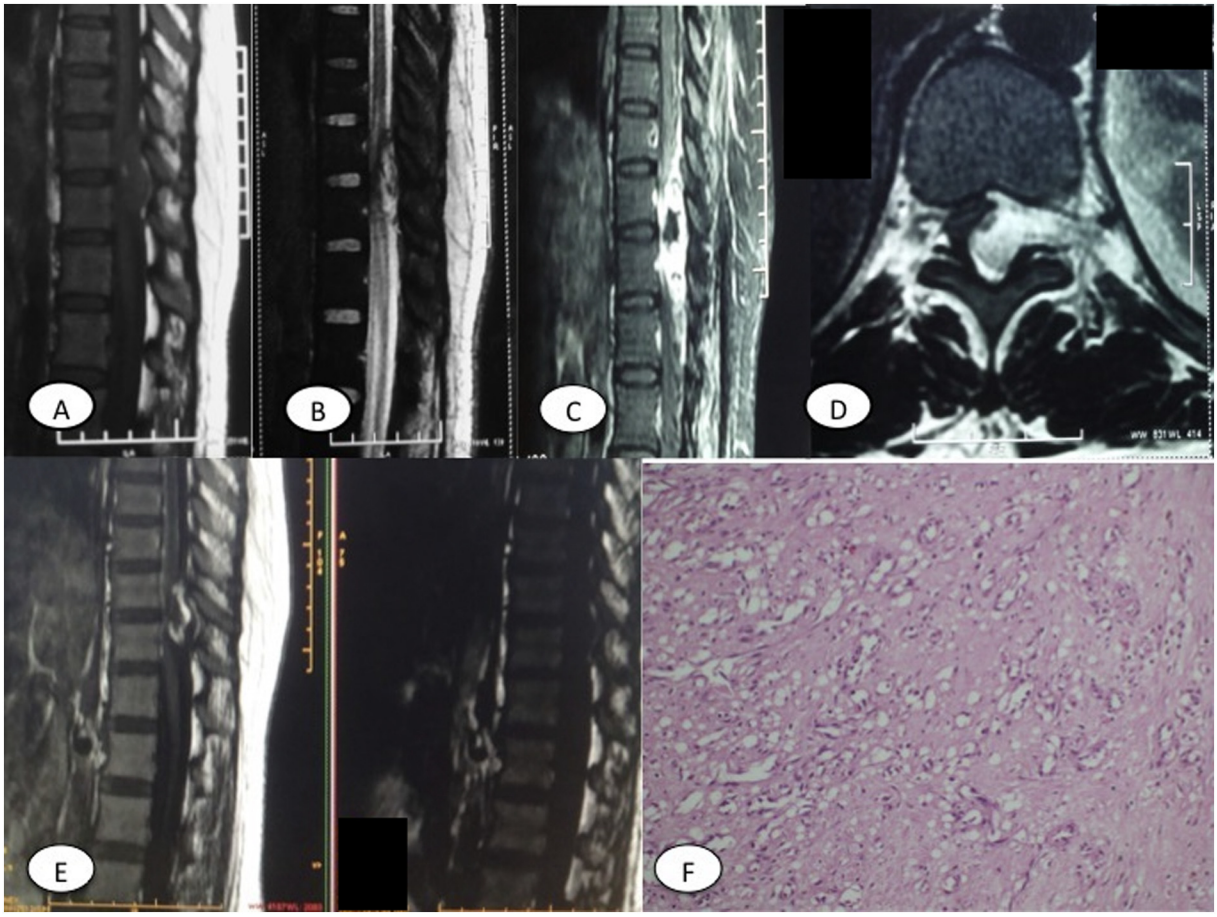

Fig. 1. MRI showed presence of an epidural lesion at the level of D10–D11 vertebrae, which was isointense on T1 sequence, with a hyperintense rim suggestive of presence of fat (A). It was T2 hyperintense (B) and showed intense contrast enhancement including the fat with a central non-enhancing core (C). Trans-foraminal extension was seen at Left D10–D11 neural foramen. A rim was seen on both out-of-phase and in-phase IDEAL images suggesting presence of both fat as well as tumor tissue containing water (E). Histological microphotograph showed tumor composed of lobules of thin walled vascular spaces lined by flat endothelium with large areas of hemorrhage and necrosis (F).

ruptured spinal cord vascular malformation.

2.2. Radiology

A gadolinium enhanced magnetic resonance imaging (MRI) of the spine showed a completely epidural, well defined mass lesion against D10 to D11 vertebrae with left D10-D11 transforaminal extension. The lesion was T1 isointense, with a hyperintense rim that suppressed on fat saturation images. The lesion demonstrated a rim which was seen on both out-of-phase and in-phase IDEAL images (iterative decomposition of water and fat with echo asymmetry and least-squares estimation) suggesting the presence of both fat as well as tumor tissue containing water (Fig. 1A–E). It was T2 hyperintense and showed intense contrast enhancement including the fatty component with a central non-enhancing core. The radiological possibility was an angiolipoma considering the fat signals inside the lesion and acute clinical presentation.

2.3. Management

After a multi-disciplinary team discussion, an elective caesarean section was done and a healthy boy weighing 2400 g with an Apgar score of 3, 6, 9 at 1, 5, 10 min respectively was delivered. Interestingly, the patient had 20% improvement in sensation and mild motor improvement (by grade 1 MRC) after delivery of the child.

Thereafter, patient underwent D9–D11 laminectomy with left D10–D11foraminotomy in prone position. Following laminectomy, a highly vascular, soft tissue mass was seen in the epidural space causing severe thecal compression. There were no bony changes and nerve roots were free. The vascular mass was excised in toto. After removal of the mass, the cord re-assumed its normal pulsations. The patient remained neurologically unchanged in the immediate post-operative period.

At 1-month follow-up, her lower limb power had improved to MRC grade 2/5, sphincteric functions improved significantly and there was further improvement in sensory functions in her legs. At 6-months follow-up, her lower limb power was MRC grade 3/5 and she had moderate lower limb spasticity. Follow-up MRI at 8 months showed some myelomalacic changes at the operative site without any residual mass/compression (Fig. 2A–D).

2.4. Histopathology

The tumor was composed of lobules of thin walled spaces lined by flat endothelium with large areas of hemorrhage and necrosis. The stroma showed presence of mild, mononuclear lymphocytic infiltrate. The picture was consistent with capillary hemangioma (Fig. 1F).

3. Discussion

Vertebral hemangioma (VH) is benign vascular hamartomatous malformations, being classified by the dominant vascular channel (capillary, cavernous, arteriovenous or venous) on histologic examination. When Virchow first described VHs as benign developmental

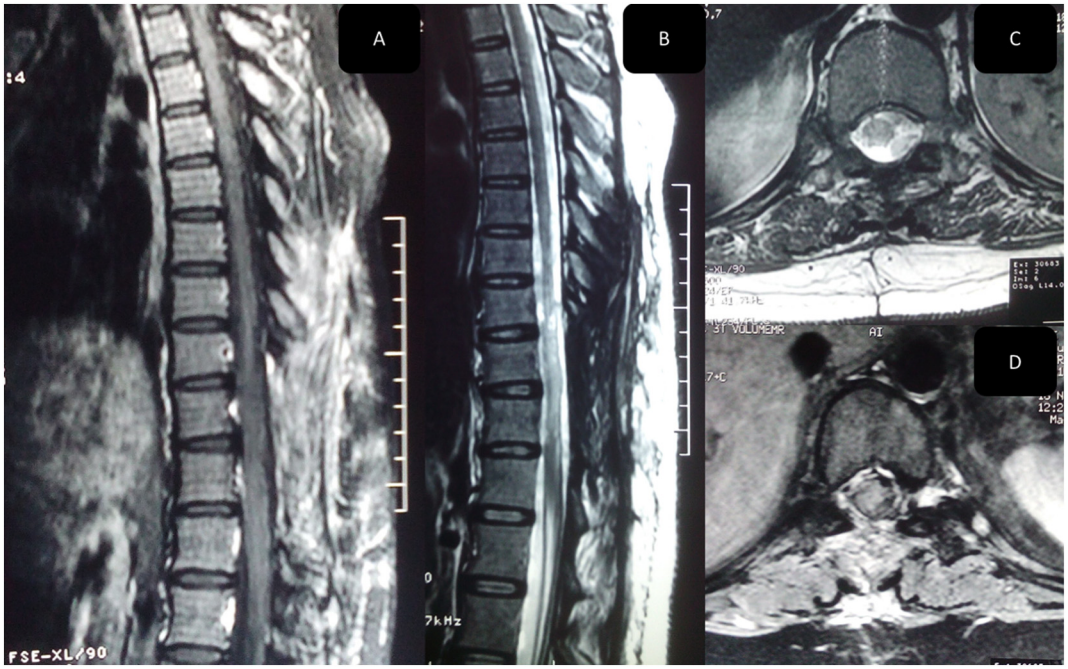

Fig. 2. (A) Shows T1 weighted imaging sagittal MRI and (B) T2 weighted imaging showing laminectomy defect with some myelomalacic changes at the operative site without any residual mass/compression. (C) Axial T2 weighted imaging and (D) axial T1 weighted imaging showing well defined thecal sac at operative site.

hamartomas of vascular origin; he was unaware of this completely extraosseous, epidural variety (0.22 in a million) [1,2]. Since only 1% of VH are actually symptomatic, very few cases are reported in literature [3]. The origin of these hemangiomas in spinal cord is supposed to be the meningeal coverings and vasa nervosum.

Symptomatic VH in pregnancy has been mainly described in the upper thoracic levels [3]. Furthermore, till now only about 30 cases of VH have been reported to be symptomatic in pregnancy [4]. It is estimated that the epidural hemangioma constitute approximately 4% of all spinal epidural masses and 12% of all extraosseous hemangiomas [5,6]. Purely epidural hemangioma of capillary variety is further rare with only 10 reported cases in the literature (Table 1).

3.1. Pathogenesis of PESH and cause acute symptoms in pregnancy

The similarity in embryological pathogenesis between cutaneous and epidural hemangioma suggests an important role of estrogen hormone on the rapid growth of these vascular lesions [7]. These hamartomata are known to grow by cellular hypertrophy and thereby distinguishing itself from malignant pathologies. It is known that estrogen increases the proliferation of hemangioma via estrogen mediated stem cells and estrogen receptor-alpha [8]. Unlike the cavernous variety, capillary hemangiomas are benign and never bleed. Thus, an aggressive clinical presentation of capillary hemangioma is extremely rare. Since most of the male cases have been reported before, the effect of pregnancy or hormonal imbalance still remains under discussed.

Apart from direct effects of hormones, the physiological state of hypervolemia in pregnancy favours the rapid growth of spinal vascular lesions [8]. This was further evidenced by partial symptom resolution after the child birth. Moreover, the growing uterus tends to compress the inferior vena cava, indirectly causing a venous hypertension, in the paraspinal venous plexus. The increasing venous pressure further leads to rapid growth of hemangiomas. Further, increase in plasma volume has deleterious effect of these lesions. What remains interesting is that the lesion manifested itself suddenly in a fashion characteristic of apoplexy in vascular malformations.

3.2. Difficulty in preoperative suspicion on neuroimaging

The radiological features of intradural extramedullary lesion with homogenous enhancement generally points towards neuroma, meningioma, hemangioblastoma, lymphoma or hemangiopericytoma. Moreover, the presence of foraminal extension is a hallmark of nerve sheath tumors [9,21]. Being a hamartoma, these lesions do contain non vascular tissues also - including fat, smooth muscle, fibrous tissue, and hemosiderin [9]. Presence of fat in these lesions is considered a good prognostic sign [10]. We misdiagnosed our case radiologically due to the fat content. Unlike similar lesions inside the central nervous system, the purely epidural hemangiomas lack peripheral hemosiderin ring, thus making a pre-operative diagnosis further difficult.

3.3. Histopathology and role of immunochemistry

Pathologically, hemangiomas can be capillary or cavernous types. At one side, cavernous type shows a large number of sinusoidal channels in collagenous tissue whereas the capillary hemangioma shows thin irregular capillary-sized vessels. Immunohistochemistry have promising role in differentiating these two and also from other more common epidural spinal pathologies like schwannoma or malignancy. Immunohistochemistry shows a positive reaction for CD31 and CD 34 but is negative for S100 and epithelial membrane antigen [11]. The capillary variety is further divided into 3 types by Alakandy et al. - metameric capillary hemangioma involving the skin, deep soft tissue and the spinal cord and solitary tumor without cutaneous lesions [12]. Cavernous hemangioma generally presents acutely (possibly due to intratumoral bleed) as compared to the capillary hemangiomas which presents in chronic progressive benign course.

3.4. Management dilemma: CS first or spine surgery first?

The decision depends mainly on the fetal maturity. If > 34 weeks, CS should be performed first because (a) after delivery there are chances of spontaneous recovery, (b) positioning during laminectomy becomes easier and (c) intraoperative blood loss is less. However, before 34 weeks, it is imperative to administer steroids for lung maturity

(theoretically it may also help in controlling spinal cord functions) and then under uterine relaxants. In such situation the surgical planning needs to be done in lateral position (laminectomy).

The pure epidural hemangioma has a favourable prognosis showing considerable improvement when the presentation is a chronic myelopathy. However, like our case and many others reported [19], once paraplegia sets in and some time has elapsed since its onset, the recovery is disappointing.

4. Conclusion

We have described an extremely unusual cause of paraplegia in pregnancy. Perhaps, pregnancy related hemodynamic alterations and hormonal changes led to sudden expansion of purely epidural spinal capillary hemangioma. These patients are best managed by a multidisciplinary team. The treatment of choice is urgent cord decompression but the decision has to take into account the age of gestation, fetal maturity and thus needs to be individualized. The prognosis in acute paraplegia, whether or not the culprit hemangioma bled, remains guarded.

Conflict of interest

The authors declare that the article and its content were composed in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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