

# A Case of Giant Intradural Extramedullary Capillary Hemangioma

Cheng-Zhang Shi, Jian Shen, Chu-Tian Zheng, Ren-Ya Zhan

Department of Neurosurgery, First Affiliated Hospital, Zhejiang University, School of Medicine, Hangzhou, Zhejiang 310003, China

To the Editor: Capillary hemangioma (CH), as a benign tumor of blood vessels, commonly appears in the skin or other superficial soft tissue throughout the body among pediatric population. Spinal intradural extramedullary capillary hemangioma (IDEMCH) is rare. According to the recent review by Bouali *et al.*, only forty cases have been reported in English literature,<sup>[1]</sup> and the size of the existing masses are generally around one vertebral length on the magnetic resonance imaging (MRI). In this paper, we report the first case of CH in the cauda equina that extends over two vertebral bodies, and discuss the diagnosis and treatment associated with such disorder.

A 73-year-old man survived a 5-year history of numbness in both legs, above the knee, but below the hip, and intermittent bilateral lumbocrural pain which could be exacerbated by walking and relieved by physiotherapy or rest. Roughly, a year ago, these symptoms got worsen. On admission, the patient complained of gait disturbance. The neurological physical examination revealed a weakness of the right leg, paresthesia of both lower limbs and limitation in straight leg raising test on both sides. MRI of the spine indicated a well-circumscribed mass with 6 cm × 1.5 cm × 2 cm in size at the level of T11-T12, in the intradural extramedullary region. The lesion appeared isointense relative to the spinal cord on T1-weighted image (T1WI), slightly hyperintense on T2-weighted image (T2WI) and homogeneous enhancement with the administration of gadolinium [Figure 1a-1c]. In addition, signal void and nodular enhancement of the dura mater were also identified. Subsequently, T11-T12 laminectomy was performed. Intraoperatively, a dark reddish nodular mass was found adhering to the cauda equina, and the surgical plane was ill-defined between the mass and the nerve roots. Finally, the lesion was totally removed [Figure 1d] with significant bleeding occurred during the separation process. Following surgery, the patient's muscle strength of lower extremities declined and his sensation diminished below the T12 dermatome. Four weeks later, the sensory disturbance improved significantly, and the motor deficit got partially relieved. Histologically, the lesion was enveloped by the fibrous tissues, and was separated into lobular arrangement, with each lobule dominated by capillary-sized vessels. Immunohistochemical studies showed CD31 and CD34 positive cells lining the blood vessels, and subendothelial layer appeared cells positive for smooth muscle actin [Figure 1e-1g]. All of these features satisfied the diagnosis of CH.

Hemangioma is a benign vascular disorder usually encountered in the skin or the subcutaneous tissues. Clinically, the lesion is separated into capillary and cavernous types according to the vessel size.<sup>[2]</sup> IDEMCH is rare, and due to the specific location, the natural history of IDEMCH has not been well described. According to the ISSVA system, CH of any organs is considered to be an infantile vascular tumor that appears after birth, progressively enlarge over a month and then regress rapidly.<sup>[3]</sup> However, this point of view seems questionable since Abe *et al.* had pointed that the spontaneous involution of central nervous system capillary hemangioma (CNSCH) is unlikely, and the GLUT1 protein, which is a diagnostically useful maker for infantile hemangioma, is not expressed within CNSCH.<sup>[4]</sup> To our mind, with the integration of the ISSVA system and the clinical character of IDEMCH, we believe that IDEMCH is more in line with the noninvolution type of the congenital hemangioma which present at birth and proportional growth without regression.

MRI is the main preoperative examination used for the visualization of IDEMCH. The lesion is commonly isointense relative to the spinal cord on T1WI, slightly hyperintense on T2WI and homogeneously enhanced after contrast injection.<sup>[1]</sup> The size of IDEMCHs was generally around one vertebral length on the sagittal images, except the case reported here, which extended over two vertebral bodies. Some special imaging features, including linear enhancement of the dura adjacent to the mass on contrast-enhanced T1WI and signal voids within dura on T2WI, were reported.<sup>[1,5]</sup> In our case, the spinal dura showed nodular enhanced with the administration of gadolinium. According to the literature, most of the IDEMCHs are radiologically mistaken for meningioma or schwannoma. However, in view of the conclusions of Abdullah *et al.*,<sup>[5]</sup> we believe that the presence of above-mentioned special MRI features within the dura mater could help raise the possibility of CH.

The treatment strategy of IDEMCH remains empirical. According to the literature, surgery is the only applied method for treating the

**Address for correspondence:** Dr. Ren-Ya Zhan,

Department of Neurosurgery, First Affiliated Hospital, Zhejiang University,  
School of Medicine, Hangzhou, Zhejiang 310003, China  
E-Mail: zjuneuros@163.com

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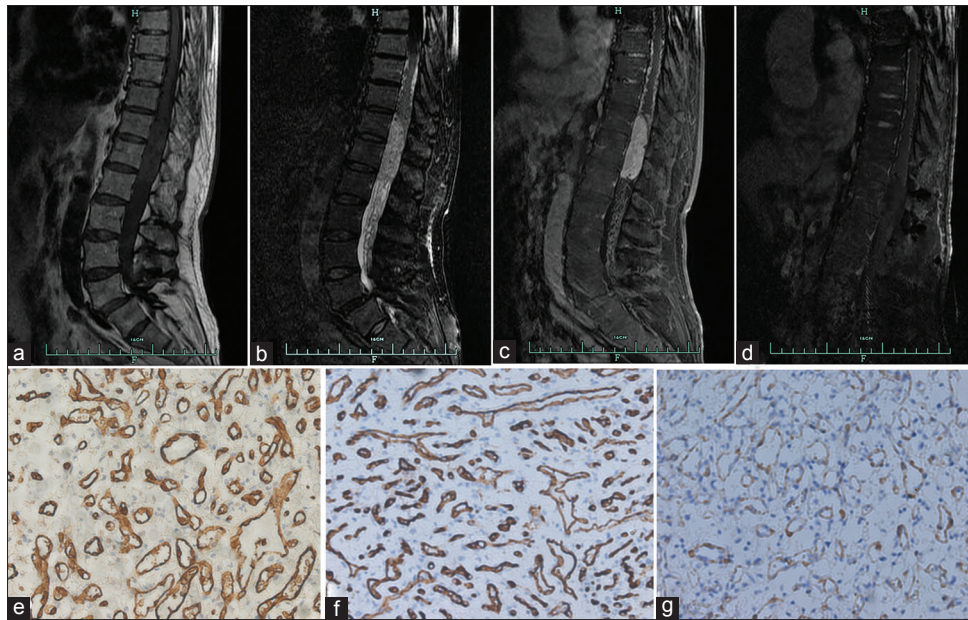
DOI:  
10.4103/0366-6999.198027

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**Received:** 24-07-2016 **Edited by:** Li-Min Chen

**How to cite this article:** Shi CZ, Shen J, Zheng CT, Zhan RY. A Case of Giant Intradural Extramedullary Capillary Hemangioma. Chin Med J 2017;130:251-2.



**Figure 1:** Magnetic resonance imaging appearance and pathological assessment: (a) cross-section on T1-weighted image (b) cross-section on T2-weighted image (c) Contrast-enhanced T1-weighted image (d) Contrast-enhanced T1-weighted image obtained 4 weeks after operation shows total removal of the lesion. (e and f) CD31 (left), CD34 (right) immunostaining shows positive cells lining the blood vessels (original magnification  $\times 200$ ) (g) smooth muscle actin immunostaining shows positive cells in the subendothelial layer (original magnification  $\times 200$ ).

lesion. The mass was exposed through standard posterior approach, *en bloc* resection was performed by identifying a plane between the mass and the spinal cord. In our case, the operative method and the resection technique are basically the same as those reported cases; however, significant bleeding occurred when detaching the tumor from the nerve root, causing an unsatisfactory prognosis. In comparison with the existing cases, we have identified that the lesion we reported was significant larger in size. Therefore, we believe that the risk of intraoperative hemorrhage could be associated with the tumor size, and preoperative embolization is necessary, especially when the lesion extends over two vertebral bodies.

In brief, CH are an extremely rare entity but should be considered in the differential diagnosis of intradural extramedullary lesions. The homogeneous enhancement of the mass combine with the signal changes of the dura mater on MRI could help raise the possibility of CH. Appropriate treatment appears to be gross total resection. In case that the lesion extends more than two vertebral bodies, preoperation embolization is recommended.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

### REFERENCES

1. Bouali S, Maatar N, Bouhoula A, Abderrahmen K, Kallel J, Jemel H. Intradural extramedullary capillary hemangioma in the upper cervical spine: First report. *World Neurosurg* 2016;92:581-7. doi: 10.1016/j.wneu.2016.04.131.
2. Takata Y, Sakai T, Higashino K, Goda Y, Tezuka F, Sairyo K. Intradural extramedullary capillary hemangioma in the upper thoracic spine: A review of the literature. *Case Rep Orthop* 2014;2014:604131. doi: 10.1155/2014/604131.
3. Lowe LH, Marchant TC, Rivard DC, Scherbel AJ. Vascular malformations: Classification and terminology the radiologist needs to know. *Semin Roentgenol* 2012;47:106-17. doi: 10.1053/j.ro.2011.11.002.
4. Abe M, Misago N, Tanaka S, Masuoka J, Tabuchi K. Capillary hemangioma of the central nervous system: A comparative study with lobular capillary hemangioma of the skin. *Acta Neuropathol* 2005;109:151-8. doi: 10.1007/s00401-004-0921-4.
5. Abdullah DC, Raghuram K, Phillips CD, Jane JA Jr., Miller B. Thoracic intradural extramedullary capillary hemangioma. *AJNR Am J Neuroradiol* 2004;25:1294-6.