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CASE REPORT

# A rare invasive hemangioma in cauda equina

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**Abstract**
*Purpose* Typical hemangioma of cauda equina with relative clear margin has been described in some case reports, but atypical hemangioma with invasion, infiltration, and augmentation of all nerve roots in the cauda equina area has never been reported. In this paper, we reported a rare case with invasive hemangioma in cauda equina, and analyzed its radiological appearance and treatment.
*Methods* We described an atypical case of hemangioma, which was revealed by MR and intraoperative photograph without clear margin and confirmed by histopathologic diagnosis.
*Results* The patient was received radiotherapy (5400 cGy/25f/5w) and continued to be clinically in good condition with follow-up MR after 24 months without further surgery.
*Conclusions* Surgery may be the first management modality for hemangiomas with severe or progressive neurologic deficits, and radiotherapy may be an alternative therapy to treat hemangiomas according to accurate histopathologic diagnosis.

**Keywords** Cauda equina · Hemangioma · Radiotherapy

## Introduction

Tumor of the cauda equina is a rare condition that can lead to major permanent neurological deficits, and the most common tumors in cauda equina are myxopapillary ependymoma (90%) and schwannoma [1]. Although typical hemangioma of cauda equina has been described in some case reports, atypical hemangioma is more rarely with vascular histogenesis and infiltration in all nerve roots of cauda equina. To our knowledge, no such cases have been reported so far, and now we report a case of rare invasive hemangioma in cauda equina.

## Materials and methods

A 51-year-old man presented a 5-month history of progressively worsening pain in his low back and legs, and he tried conservative treatment for pain control but failed.

Upon examination, he experienced radicular pain and sensory disturbances in both legs, and especially in perineal region without muscle weakness. Knee and ankle jerk reflexes were absent without positive pathologic reflexes. Straight leg raising test was positive in both leg, even leg movement during physical evaluation can exacerbate the pain.

According to MRI and dynamic contrast-enhanced MRI, the atypical tumor was indicated by intermediate signal intensity on T1 and T2-weighted images, and had relatively homogeneous enhancement after contrast. It revealed signal abnormal in all nerve roots of cauda equina (L1–L5) without clear margin demonstrating the tumor ingrowth in

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all nerve roots, and there was nothing found on X-ray and CT.

Given the patient's clinical history, and negative laboratory findings and bone marrow cytomorphologic examination, the preoperative diagnosis was intraspinal tumor invading and infiltrating all cauda equina which we had never encountered before and we cannot identify the specific type of tumor. The patient was treated with posterior decompression of lumbar spinal canal with dissection of the dura because of his serious neurological symptom.

The dura was extremely tense, and the nerve fibers bulged out immediately when a longitudinal incision was made in the center of the dura. The intraoperative findings revealed that tumor grew disseminated within all the cauda equina without separate tumor mass, and the enlarged nerve roots were firm in texture on which the tortuous vessels went around. We tried to do our best to remove the tumor but failed because the tumor invaded, infiltrated and expanded all the never roots inseparably, and we can't even distinguish clear margin of the tumor from the enlarged nerve roots. With the purpose of further treatment, we obtained a small part of vessels and nerve roots infiltrated by the tumor for histopathologic diagnosis with SEP/MEP monitoring (Figs. 1, 2, 3).

## Results

The patient had prompt relief of pain and numbness immediately, and had a good recovery over a period of 1 week after surgery with intravenous treatment (methylprednisolone and mannitol). The histopathologic diagnosis was cauda equina hemangioma, and the patient was received radiotherapy (5400 cGy/25f/5w) and continued to be clinically in good condition with follow-up MR after 24 months without further surgery.

## Discussion

The most common tumors in cauda equina are myxopapillary ependymoma (90%) and schwannoma, and other less common tumors are paraganglioma, intradural metastases, hemangioblastoma, meningioma, astrocytoma, spinal PNET, and ganglioglioma. In addition, some benign masses that mimic tumors are lipoma of the filum terminale, dermoid cyst, and epidermoid cyst [1–7]. In this case, atypical hemangioma is very rare with invasion, infiltration and augmentation of all nerve roots in the cauda equina area, and we haven't searched this kind of case in previous publication (Figs. 4, 5, 6).

Tumors are generally easy to diagnose according to MRI and dynamic contrast-enhanced MRI, and the tumors or benign masses presented above are usually solitary and sporadic, often associated with intense enhancement, intrinsic vascular changes (thrombosis; sinusoidal dilatation), cyst formation, haemorrhage, calcification and fatty degeneration [7, 8], and have characteristic appearance on MRI, so it may be the gold standard for the diagnosis of these kinds of tumors.

Typical hemangioma is mainly located in the vertebral bone of the spine, accounting for about 5% of all vascular lesions in the spine, and only about 3% of which is found inside the spinal canal sporadically with clear margin. Histopathologically, typical hemangiomas are vascular malformations consisting of vascular spaces lined with a

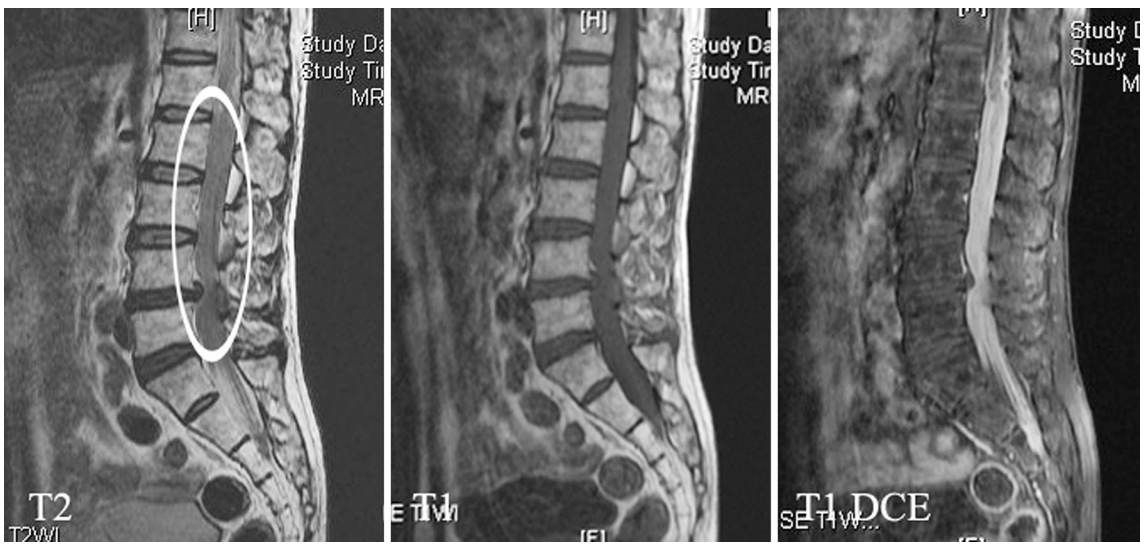


**Fig. 1** Preoperative sagittal MRI images (T2, T1-weighted, and T1-weighted enhanced images) (*white oval* showed all cauda equina with infiltration of hemangioma without clear margin)

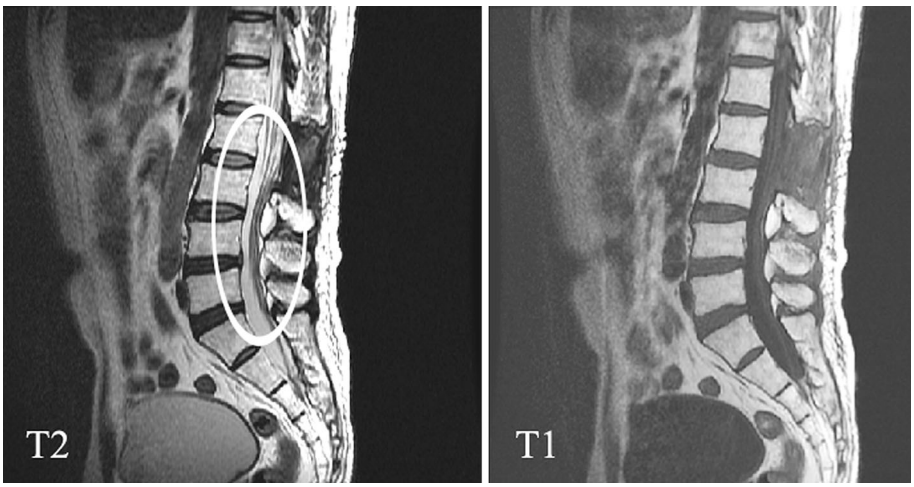


**Fig. 2** Follow-up sagittal MRI images (T2 and T1-weighted images) (*white oval* showed relative normal nerve roots)

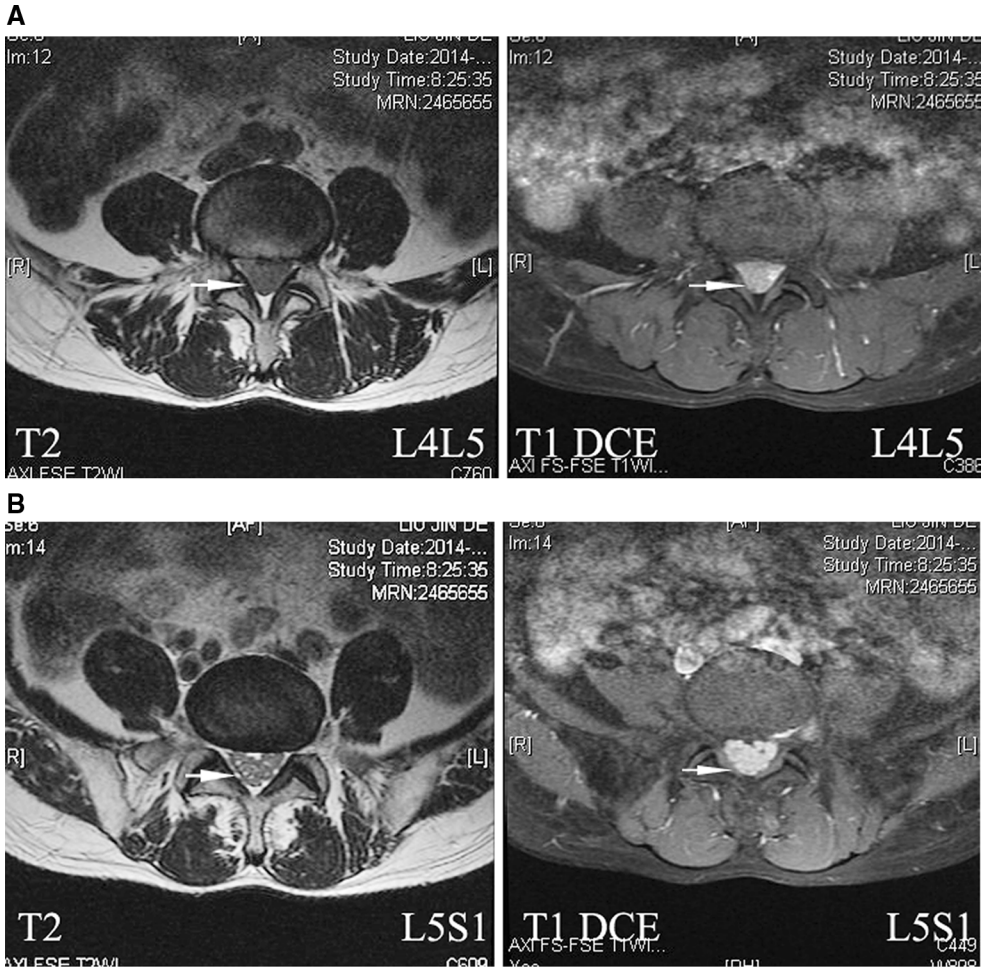


**Fig. 3** Preoperative MRI images on L4/L5 (**a**) and L5/S1 (**b**) level (*arrow* enlarged nerve roots infiltrated by hemangioma)

single endothelial layer without neural or glial elements. Cavernous hemangioma is comprised of closely packed, large dilated vascular channels without interposed neural tissue. Unlike cavernous hemangioma, capillary hemangioma is histologically characterized by a lobular architecture, with each lobule separated by septa of fibrous connective tissue and consisting of a myriad of small and very small capillaries lined by endothelial cells [9]. Therefore, the main difference between cavernous and capillary hemangioma is the dominant vessel size at

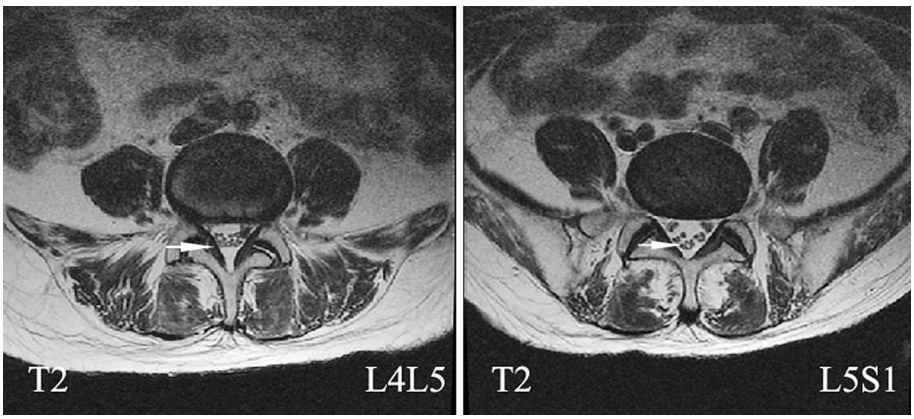


**Fig. 4** Follow-up MRI images on L4/L5 and L5/S1 level (*arrow* normal nerve roots)

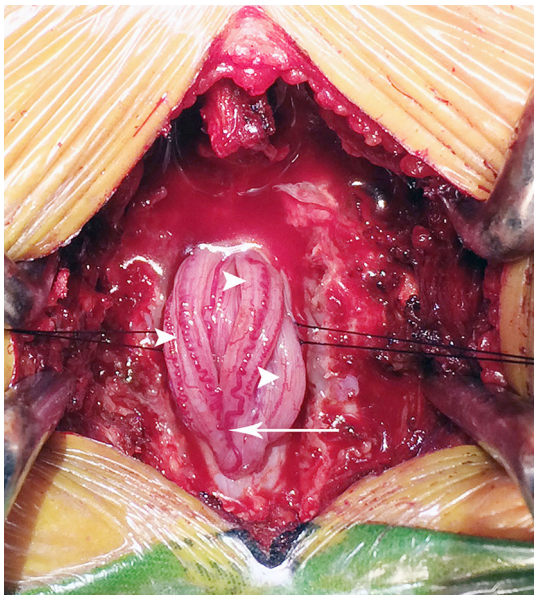

**Fig. 5** Intraoperative photograph showed hemangioma ingrowth and expand all cauda equina with tortuous vessels covering enlarged nerve roots, and the cauda equina just like the legs of octopus (*plain arrow* tortuous vessels; *arrowheads* enlarged nerve roots infiltrated by hemangioma)

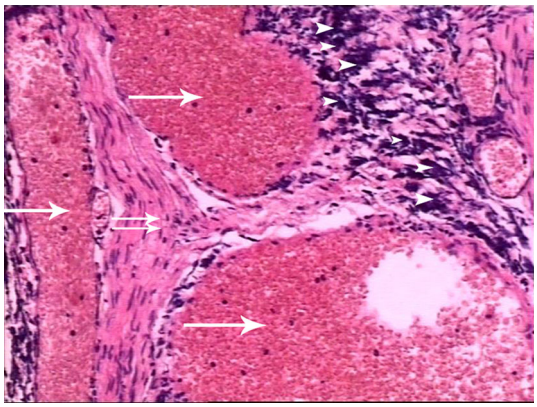

**Fig. 6** Microscopic histological appearance of the cauda equina hemangioma with infiltration of vessels and lymphocytes from the surgical specimen (*plain arrows* blood vessel malformation; *arrowheads* lymphocytes; *double arrows* nerve fibers)

microscopy. In our case, it is a much larger and poorly-circumscribed lesion within the nerve roots. The irregularly dilated blood-filled vessels arrange in a haphazard pattern lined by nerve fibers, and inflammatory cells could be observed around the vessels. Thus, it is totally different from the typical hemangioma.

MRI appearance of cavernous hemangioma inside the spinal canal shows a mixed signal on the T1 and T2-weighted images with a hypointense ring of hemosiderin in the T2-weighted images, and the enhancement is variable [6, 7, 10], and capillary hemangioma appears isointense or slightly hyperintense on T1-weighted, and hyperintense on T2-weighted images with strong homogeneous enhancement [9]. However, in this atypical case, the tumor was poorly-defined isointense on T1 and T2-weighted images with relatively homogeneous enhancement after contrast. It seemed that the tumor invaded, infiltrated and expanded all the never roots inseparably. Therefore, it is very difficult to obtain a correct preoperative diagnosis in our case based on the atypical radiological presentation, and these tumors mentioned above should be taken into account.

Recent studies suggested the neurologic recovery rate from surgical decompression for typical hemangioma in cauda equina with serious neurologic deficits was nearly 100% [11]. However, in this atypical case, misdiagnosis can potentially lead to serious complications even death, we had no choice but to choose decompression as our first treatment choice because of the patient's serious neurological symptom, unfortunately, we failed to complete excision of the tumor because we cannot even distinguish clear margin of the tumor from enlarged nerve roots during the operation, it revealed that the tumor grew disseminated within all the cauda equina.

The histopathologic diagnosis indicated that radiotherapy may be an alternative therapy to further surgical treatment because of impossible excision of the tumor,

which was introduced as a treatment for hemangioma in 1930, and may provide better results and fewer complication for subdural lesions. It can obliterate hemangiomas and control pain through vascular necrosis and/or anti-inflammatory effect [12, 13].

Radiotherapy alone may not be appropriated as the treatment for patients with severe or progressive neurologic symptoms, but may be suitable for mild and slowly progressing neurologic deficits in spite of the complications of radiotherapy including radiation-induced myelitis, necrosis and secondary malignancy. Heyd et al. [14] reported that neurologic symptoms completely resolved in 79% of patients after radiotherapy, more than 80% of those patients had undergone previous surgical interventions, and the potential for complications, especially as far as secondary malignancy, caused by radiotherapy might have been overemphasized. In this case, the patient was received radiotherapy (5400 cGy/25f/5w) and continued to be clinically in good condition with follow-up MR after 24 months.

## Conclusion

Hemangiomas of the cauda equina are rare condition that can lead to major permanent neurological deficits, and MRI may be the gold standard for the diagnosis of typical ones, but in this atypical case, histopathologic diagnosis is more important than MRI to guide further treatment. Surgery may be the first management modality for patients with severe or progressive neurologic deficits, even without histopathologic examination and accurate preoperative diagnosis. In our opinion, radiotherapy may be an alternative therapy to treat hemangiomas according to accurate histopathologic diagnosis.

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**Compliance with ethical standards**

**Conflict of interest** The authors declare that they have no competing interest, and the contents of the paper have not been presented or published previously.

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