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Case Report

Lymphangioma in the Epidural Space of the Thoracic Spine

Bok Yong Ha, M.D.,[1] Jun Bum Park, M.D.,[1] Young Min Kim, M.D.,[2] In Uk Lyo, M.D.[1]
Departments of Neurological Surgery,[1] Pathology,[2] Ulsan University Hospital College of Medicine, University of Ulsan, Ulsan, Korea

A rare case of solitary intraspinal epidural lymphangioma is described with a review of the literature. A 16-year-old boy was admitted to our hospital with a history of two-year of progressive paraparesis. Magnetic resonance imaging study revealed a 2 × 2 × 6 cm sized epidural cystic mass in the thoracic spine. Surgical total removal and biopsy were performed. The final pathologic report on the mass indicated lymphangioma.

KEY WORDS : Lymphangioma · Thoracic · Epidural.

INTRODUCTION

Lymphangioma of the soft tissue is fairly common, but lymphangioma presenting as a mass in the epidural space is extremely rare. Although magnetic resonance imaging (MRI) is considered the best modality for evaluating most soft-tissue tumors, it proved to be difficult to distinguish the present case from other tumors, such as schwannoma or meningioma before surgery using MRI.

CASE REPORT

A previously healthy, 16-year-old boy was presented with a history of two-year of progressive paraparesis. While sensation in the lower extremities were intact, motor examination of the lower extremities demonstrated paraparesis and increased deep tendon reflexes. The laboratory data and plain radiographs were normal. MRI revealed a well-enhanced epidural longitudinal cystic mass, compressing the dural sac, from the fifth to seventh thoracic vertebral bodies. This mass was relatively well-defined and displayed homogenous signal intensity on T2- and T1-weighted image but significant enhancement along the membrane on postcontrast images (Fig. 1). Tumor was resected through laminectomies from T5 to T7. Intraoperative findings showed that the tumor was entirely extradural and was not adherent to the osseous spine or epidural wall. The tumor was a thin-walled, transparent cystic mass filled with clear fluid (Fig. 2). The entire tumor was easily removed. No penetration of the tumor into the nerve root was found. The pathological specimen was found to be a characteristic cystic lesion lined by a single layer of flattened endothelial cells (Fig. 3A). Immunohistochemical stains for CD34 and D2-40 were positive (Fig. 3B, C). The patient completely recovered from paraparesis four months after the operation (Fig. 4).

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• Address for reprints : In Uk Lyo, M.D.
Department of Neurological Surgery, Ulsan University Hospital, College of Medicine, University of Ulsan, 290-3 Jeonha-dong, Dong-gu, Ulsan 682-714, Korea
Tel : +82-52-250-7130, Fax : +82-52-250-7138
E-mail : iulyo1@uuh.ulsan.kr

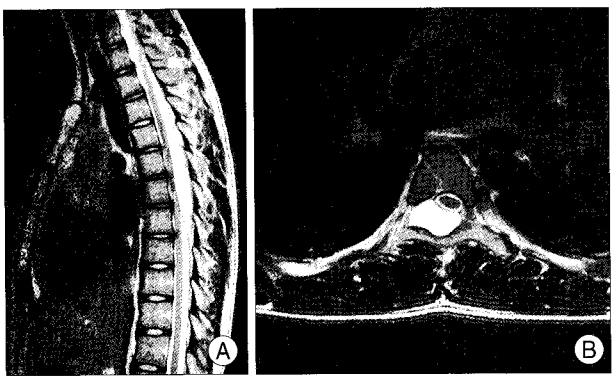

Fig. 1. Preoperative MRI. A : Sagittal MRI showing the extent of the tumor. B : Axial MRI showing the extent of the tumor, compressing the spinal cord to the left side.

DISCUSSION

Only a few cases of lymphnagioma of the spine have been reported[2]. Whether lymphangioma is a true neoplasm, a hamartoma, or lymphangiectasia remains controversial and the pathophysiological mechanism remains uncertain. Most lymphangiomas clinically manifest during childhood. The pathophysiological mechanism remains uncertain. Chervenak et al.[1] theorized that lymphangioma to be a malformation arising from sequestration of lymphatic tissue that had failed to communicate normally with the lymphatic system during fetal development. On the contrary, some lymphangiomas are considered to be acquired lesions arising from obstruction of affected lymphatic vessels[3]. The natural history of the typical lymphangiomas involves a slowly growing mass without malignant change. In our case, histological examination revealed a cystic lesion lined by a single layer of endothelial cells and strong positive immunohistochemical results for CD34 and D2-40. Destructive injuries from lymphangioma can be caused by replacement of inherent tissue as well as blockage of lymphatics and blood vessels that may result in organ failure, occasionally associated with chylothorax or chylopericardium[6]. The common differential diagnoses include schwannoma, neurofibroma, meningioma, sarcoma, and metastasis of carcinoma. Surgical resection is indicated when lymphangiomas cause neural compression and instability of the spine[5]. Alternative treatment methods, including radiation, chemotherapy, embolization, and sclerotherapy, have been applied to unresectable lesions[6]. Recombinant interferon therapy has shown promising results for disseminated lymphangiomatosis[4]. Although lymphangiomas are benign lesions, postoperative recurrence can occur, possibly due to incomplete resection of the mass[2]. Careful monitoring must be maintained to detect local recurrence.

CONCLUSION

We report an extremely rare case of lymphangioma occurring in the epidural space of the thoracic spine that was removed through laminectomies. Although lymphangioma of the epidural spine are very rare, they must be considered in the differential diagnoses for the masses occurring in the epidural space.

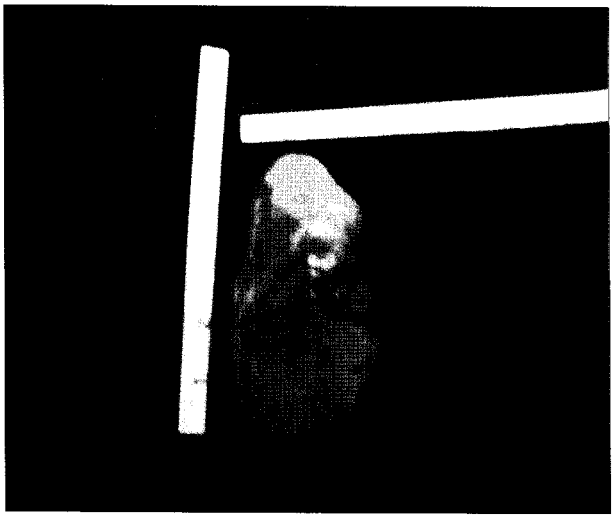

Fig. 2. The tumor is translucent, thin-walled mass measuring 2 × 2 × 6 cm.

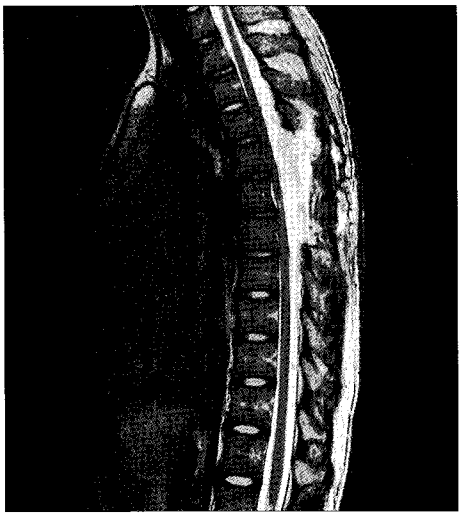

Fig. 4. Postoperative MRI.

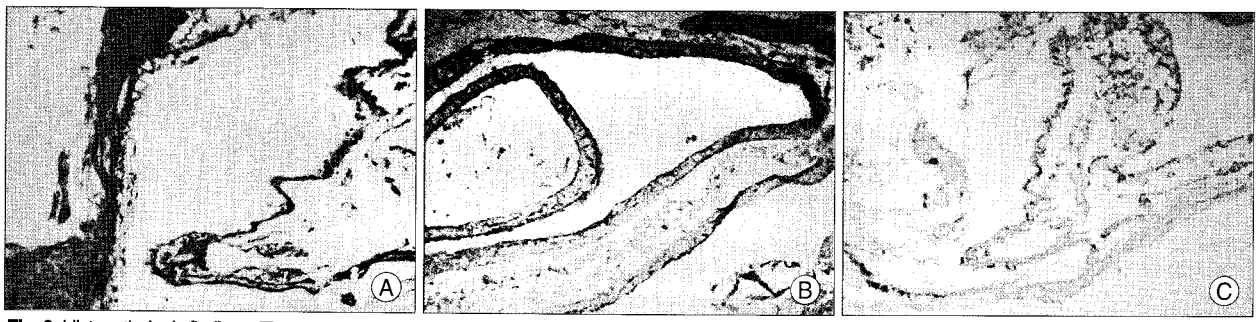

Fig. 3. Histopathologic findings. The tumor is characterized by a cystic lesion lined by a single layer of flattened endothelial cells (A : H&E, × 200). This mass is immunohistoreactive for CD34 (B : IHC for CD34, × 100) and D2-40 (C : IHC for D2-40, × 100).

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