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Hemangioendothelioma Lumbar Spine and Ribs: Case Report

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ABSTRACT

Background: Spine hemangioendothelioma is a rare low-grade vascular neoplasm that is characterized as intermediate between benign hemangioma and high-grade angiosarcoma, affecting 1 in 1,000,000 people worldwide. It has been described throughout the body with lung, liver, skin, bone and spine being the most frequent sites.

Case presentation: A 31-year-old male, complained of mid and lower back pain for 2 years, which was on and off. 5 months ago pain increased after lifting heavy object, which affected his walking and sitting. No weight loss or night sweating. He came to Dr. Solaiman Fakeeh Hospital as second opinion regarding the Lumbar 1 (L1) tumor. The patient was diagnosed with pseudomyogenic hemangioendothelioma according to radiological images and histopathology report in other center.

Conclusion: Through our experience in this case in the management of spine hemangioendothelioma tumor resection should be attempted whenever possible. Given the propensity of these lesions to recur, intralesional or subtotal resection should be followed by radiotherapy.

Keywords: Hemangioendothelioma, Spine, Radiotherapy.

INTRODUCTION

Spine hemangioendothelioma is a very rare neoplasm that has been described to have histological features between hemangioma and high-grade angiosarcoma [(1)].

Weiss *et al.* [(2)] described it first in 1982 as a vascular neoplasm of both bone and soft tissue [(2)]. It is commonly seen in soft tissues but can also be located in other organs such as lung, pleura, spleen, heart, liver, and bone [(3, 4)]. Primary hemangioendothelioma of the bone is rare, and it represents only 1% of all malignant tumors of the bone [(3)]. These tumors can be asymptomatic and discovered as an incidental finding. However, local pain is the most common presentation of spine hemangioendothelioma [(5)].

CASE PRESENTATION

A 31-years-old male, non-smoker, unknown to have any medical illness, complained of mid and lower back pain for 2 years, which was on and off. 5 months ago pain increased after lifting heavy object. Pain continued by the same intensity, intermediate, altered his sleep and increased with turning, twisting, bending and standing from sitting that affected his mobilization, sitting and daily activities. No numbness, no weight loss or night sweating and no constitutional symptoms.

Physical exam:

- Normal gait.
- Average height and weight.
- In brace (thoracolumbar brace).
- Previous incision of open biopsy.
- Mid and lower back tenderness.
- Decreased trunk flexion, extension and twisting due to severe pain.
- Right T12 distribution sharp pain.
- No neurological deficit.
- Intact sensation and motor 5/5 in all limbs.
- Reflexes were normal.
- No urinary or bladder issues.

Drug allergy: No known drug allergy.

- **Clinical Warning:** No clinical warning.
- **PMH:** Unknown.
- **PSH:** Open biopsy of lumbar spine (L1).

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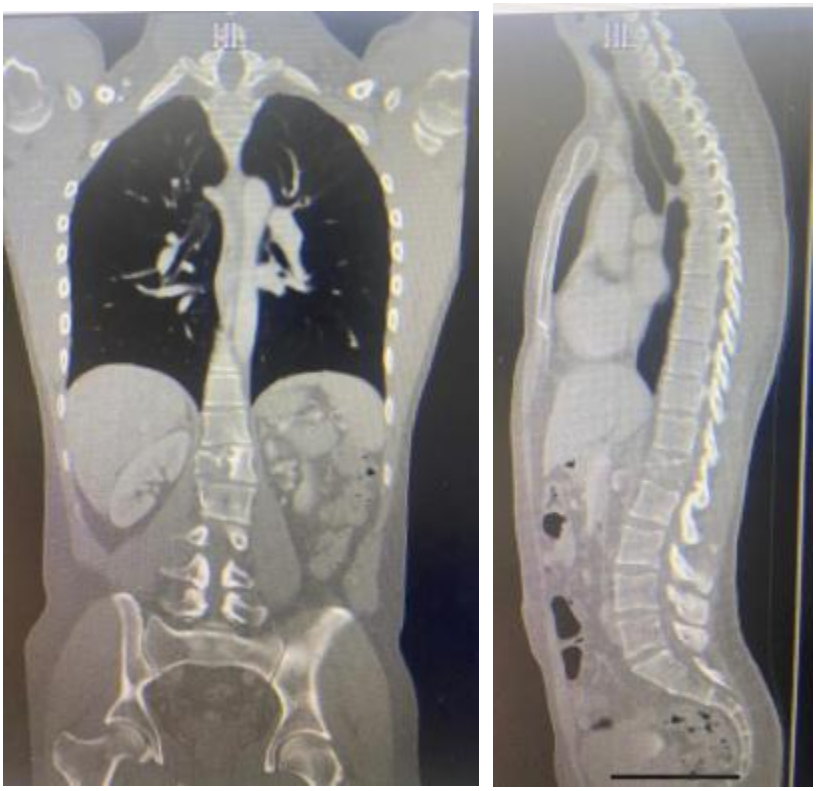

Figure (1): Pre-oprative CT scan

Radiology:

Ct scan report of thoracolumbar spine showed:

- Loss of normal height of L1 vertebral body with large multi cystic expansile lytic lesion affecting almost the 2/3 of the vertebral body extending toward the right lateral mass and posterior elements.
- A similar lesion was seen affecting the spinous process of the preceding vertebra at T 12 level.
- Multilevel atypical hemangiomas affecting body of L1 vertebra, spinous process of T12.
- Metastasis versus primary tumor.

MRI report of whole spine showed:

- Suspicious lesions affecting L1 vertebral body, T12 spinous process, which could be metastasis or unknown primary.
- Abnormal right paravertebral heterogeneous enhancement.
- Normal spinal canal.

CT chest, abdomen and pelvis for staging:

- Multi levels ribs head lesion
- No pulmonary nodules, consolidation or lung masses.
- No lymphadenopathy.
- Stable size of segment VIII arterial enhancing lesion measures 2.5 cm, which showed fading on portal venous and delayed phase, with centrally delayed enhanced scar, could be hemangiomas or tiny FNH.
- Stable another smaller lesion arterial enhancing foci scattered through the right and left lobe (at least 8, each measures 3-5 mm).
- No other significant finding.

Labs:

- All routine investigations are within normal limit.
- Histopathology report showed pseudomyogenic hemangioendothelioma (specimens were taken from right rib 12, D12 spinous process, L1 body)

Surgery details:

Operation:

Lumbar posterior corpectomy and cage with screws, posterior transpedicle percutaneous screws from D11 to L3.

Operation summary:

- L1 transpedicle vertebroctomy right side spinous process and lamina spinous process of T12, solera system 6.5 *45, expandable T2 altitude 23 to 29, rods 4.75.
- Patient prone position, induction of general anesthesia, motor evoked potential, cefuroxime 1000 mg intravenous, then shift patient to Jackson table, draping of lower back under sterile technique.
- Midline incision, Insert mini invasive T10 to T12 minimal invasive vouger system 6.5 *45.8 screws, then identify the L1 and T12, remove the spinous process lamina and facet.
- Pars from right side, isolate the nerve root of L1 reach to L1 vertebroctomy, insert L1 expandable cage, 23 to 29 mm, reduce the height kyphosis, and the meticulous hemostasis. layer by layer closure.
- Complications: No.
- Collected patient tissue: Excised lesion with multiple pieces, as difficult to be excised as one piece.
- Blood loss: Minimal

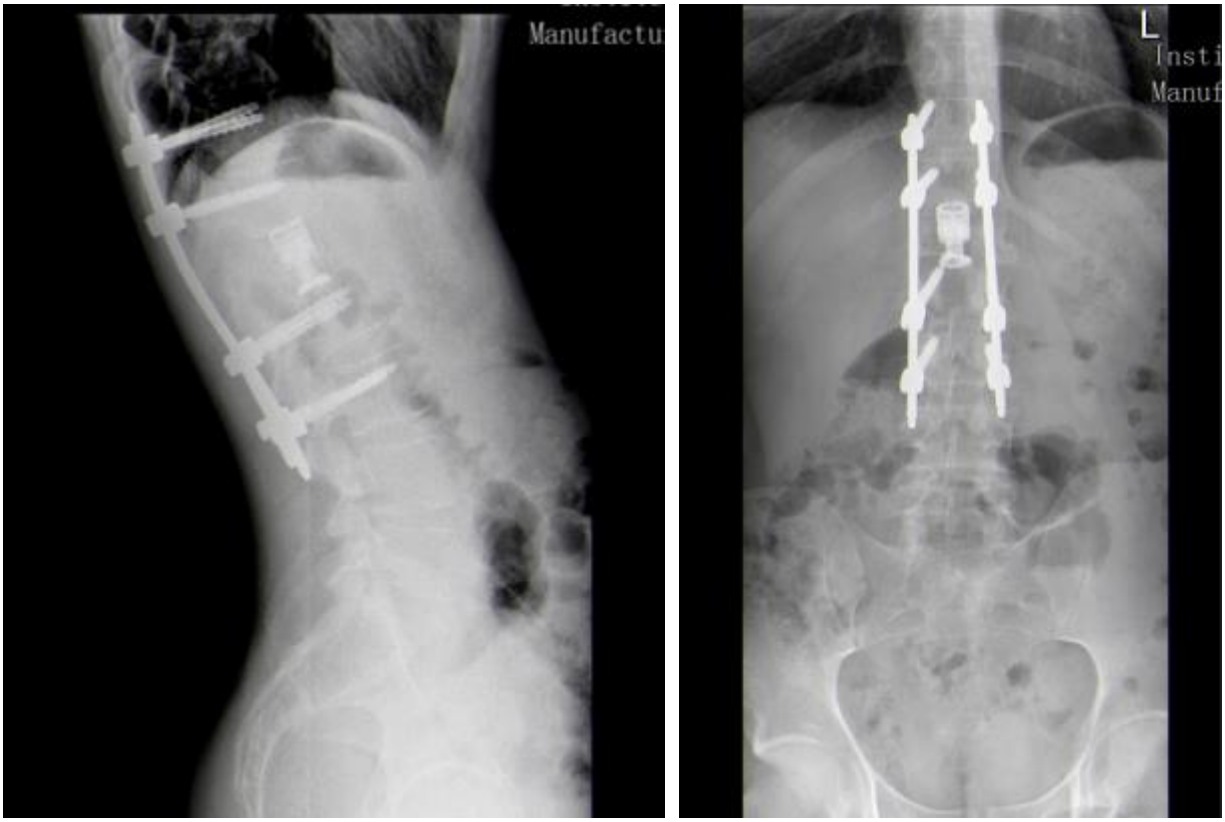

Figure (2): Post-operative X-ray.

Post-operative:

- Patient came for follow up post-surgery, wound was healed, X-Rays and MRI of whole spine did not show any gross residual lesion.
- Case have been discussed pre-operative and post-operative in tumor board meeting and agree that patient has pathological fracture in L1, T12 and apparently has involvement of the ribs head.
- The patient diagnosed with pseudomyogenic hemangioendothelioma as radiological images and histopathology.
- Recommend to do embolization of tumor before surgery to prevent bleeding.
- All his paper and images were reviewed and the conclusion was that he did not have any metastasis. The liver lesions were investigated and turned to be not of significance.
- As this type of rare tumor with possibility of local recurrence and low incidence of distant metastasis, and no clear recommendation in NCCNR guidelines, so the recommendation is to receive radiotherapy by tomotherapy, to protect the spinal cord from serious side effects, and decrease the chance for recurrence (its tolerance 50 GY).
- Patient received radiotherapy.

Ethical approval:

An approval of the study was obtained from Dr Soliman Fakeeh Hospital (Jeddah, Saudi Arabia) academic and ethical committee. The patient and his relative were informed that the case was took as case report for publishing and he was accepted. This work has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for studies involving humans.

DISCUSSION

Since Spine hemangioendothelioma is a rare disease and its natural history is unpredictable, there is no standard of care for treatment. Treatment options include preoperative embolization and surgical resection, which can be followed by radiotherapy and/or chemotherapy. Therapeutic management should begin with biopsy. CT-guided biopsy is preferred as a laminectomy and an open biopsy may make it difficult to achieve a wide resection afterwards. Patients benefit from spinal stabilization following surgical resection to prevent pain [6].

Radiotherapy for spine hemangioendothelioma is used in partially excised lesions and when it is difficult to access the lesion [5, 7, 8, 9]. **Campanacci *et al.*** [10] recommended the use of postoperative radiotherapy to decrease the risk of local recurrence after surgery. In the study reported by **Kelahan *et al.*** [11], one patient had local progression of spine hemangioendothelioma two months after surgical stabilization. For that reason, the patient subsequently was treated with localized radiotherapy, at 3 months follow-up there was significant regression of the epidural tumor.

In our case, MRI spine postoperative showed no gross residual (myogenic endothelioma), as this type of tumor is very rare tumor with possibility of local recurrence and low incidence of distant metastasis.

Radiotherapy is recommended and because there is no clear recommendation in National Comprehensive Cancer Network guidelines for such tumor. **Ma *et al.*** [1] reported that four patients who underwent surgical resection followed by radiotherapy; in three cases recurrence could not be detected after a long follow-up period.

CONCLUSION

Through our experience in this case in the management of spine hemangioendothelioma tumor, resection should be attempted whenever possible. Given the propensity of these lesions to recur, intralesional or subtotal resection should be followed by radiotherapy. We suggest initial monitoring of patients with spine hemangioendothelioma with interval CT scans to observe disease behavior. If initial imaging demonstrates slow progression of spine hemangioendothelioma in the bone without evidence of metastasis, minimally invasive surgery is recommended, and adjuvant radiotherapy may be considered.

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