Metastasis of cardiac rhabdomyosarcoma at lumbar spine: case report

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We report a case and review literatures of a 78-year-old man presenting with progressive dyspnea for 1 month. A diagnosis of cardiac pleomorphic rhabdomyosarcoma with lung and scalp metastasis was made then surgical tumor removal was performed. One month later the patient complained of severe low back pain. After complete investigation, he was diagnosed with metastatic rhabdomyosarcoma at lumbar spine and adjacent paravertebral soft-tissue. He underwent a second operation resulting in a satisfactory early postoperative outcome. He passed away approximately 6 weeks later as a result of severe congestive heart failure and irreversible respiratory failure.

This is the first case report of metastatic cardiac pleomorphic rhabdomyosarcoma at lumbar spine which is assumed to be a rare and aggressive disease.

Keywords: *Metastasis, cardiac tumor, pleomorphic rhabdomyosarcoma, lumbar spine, case report*

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Case report

A 78-year-old man presented with progressive dyspnea for 1 month. The physician detected a 2.0x1.5 cm ulcerative nodule at scalp overlying the left fronto-parietal region without other obvious abnormalities. The radiologist further noticed a 5 cm mass at right paratracheal from a chest x-ray. Chest computed tomography (CT) was then performed and confirmed with computed tomography pulmonary angiography (CTPA) in addition with an echocardiography study revealing a 5.5x3.8x5.0 cm lobulated mass at the posterior segment of the upper lobe along with a 4.2x1.6 cm subpleural nodule at the lower lobe of the right lung and a 3.4x6.0x5.3 cm lobulated mass with inhomogeneous enhancement occupying the right ventricle as well as a 2.7x2.4 cm mass at the left ventricle. (Fig. 1) The initial diagnosis was suspected of cardiac tumor with lung and scalp metastasis. An open thoracotomy with cardiac tumor removal along with tricuspid valve repair was performed simultaneously with scalp tumor excision. Histopathology and special immunohistochemistry studies of both specimens indicated a cardiac pleomorphic rhabdomyosarcoma with scalp metastasis. The patient refused adjuvant chemotherapy and radiation therapy at thoracic and scalp because of personal believes.

One month later the patient gradually felt pain at his lower back. The muscle strength was 4/5 in the lower extremities with intact sensation and negative Babinski sign from physical examination. Plain lumbar x-ray revealed an osteolytic lesion at the body of L3, and magnetic resonance imaging (MRI) of the lumbar spine suspected of bony metastasis at L3 vertebra associated with a 1.5x4.2 cm posterior epidural mass at L2-3 and a bilateral paraspinal mass with extension into the left L2-3 neural foramen causing spinal canal stenosis with compression of cauda equine nerve roots. (Fig. 2A-2D) A diagnosis of suspected metastatic rhabdomyosarcoma at lumbar spine and paravertebral soft-tissue was made. Decompressive laminectomy at L2-3 with pedicular screw fixation from L2 to L4 with vertebroplasty to L3 vertebra was performed 3 days later with satisfactory early postoperative results without any immediate complications. (Fig. 2E-2F) The histopathological features of the lesion showed pleomorphic vesicular-chromatic spindle-shaped and polygonal-shaped cells arranged in fascicular pattern with strong immunoreactivity to desmin and cytokeratin corresponding with metastatic pleomorphic rhabdomyosarcoma. Technitium-99m (Tc-99m) bone scintigraphy was performed later and the finding was increased radiotracer uptake at L2-L3 without bony metastasis elsewhere, however the patient insisted of refusing adjuvant radiation therapy at lumbar spine. Therefore, his lung metastatic condition was getting worse and massive pleural effusion was developed 4 weeks later. The patient passed away approximately 6 weeks postoperatively as a result of severe congestive heart failure and irreversible respiratory failure.

Discussion

Cardiac tumors are considered to be rare diseases with the incidence ranging from 0.0017% to 0.28% in autopsy studies.[1] Primary lesions are

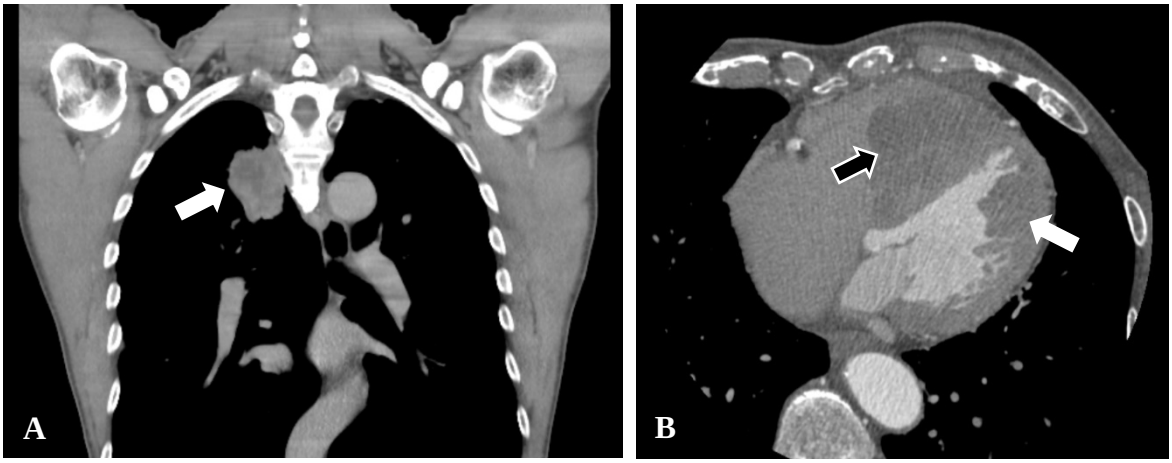

Fig. 1 Chest computed tomography (CT) shows lobulated mass (white arrow) at posterior segment of the right upper lung. (A) Computed tomography pulmonary angiography (CTPA) demonstrates masses at the right ventricle (black arrow) and left ventricle (white arrow). (B)

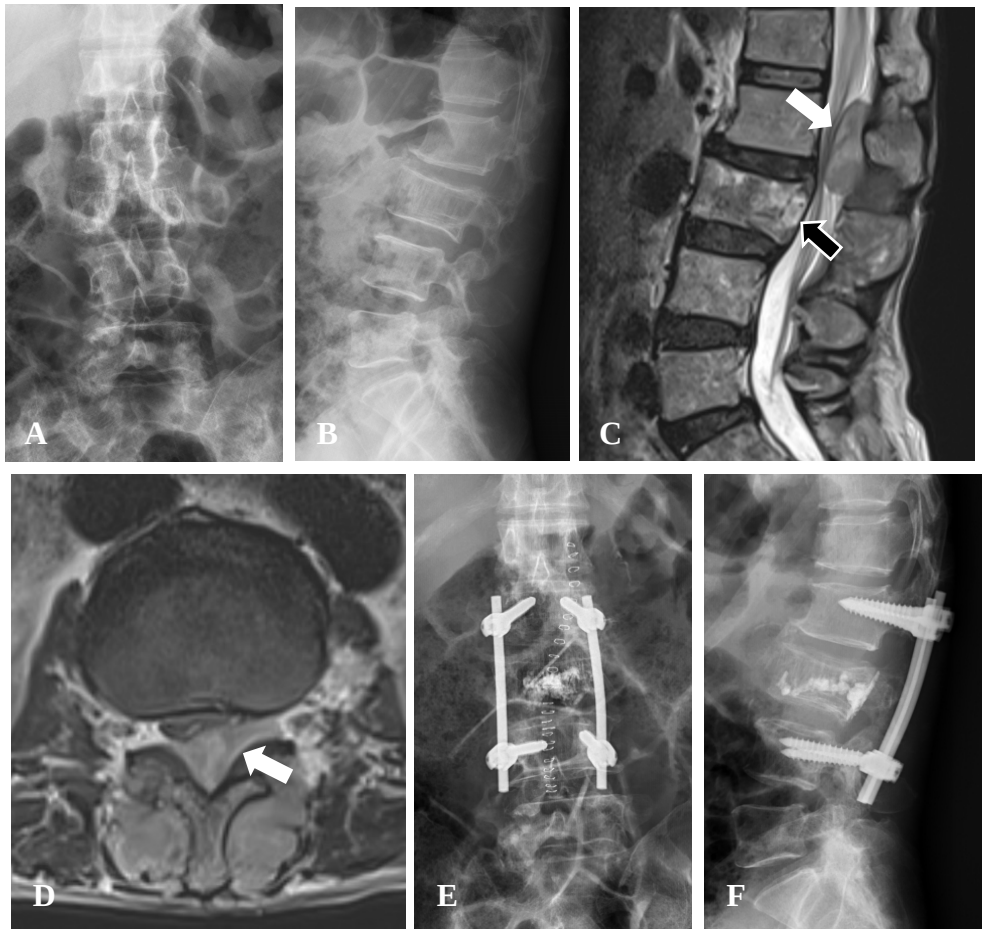

Fig. 2 Plain lumbar x-rays demonstrate an osteolytic lesion at the body of L3. AP view (A), Lateral view (B) Magnetic resonance imagings (MRI) of the lumbar spine depict bony metastasis at L3 vertebra (black arrow) and an epidural mass at L2/3 level (white arrow). Sagittal view (C), Axial view (D) Postoperative x-rays reveal spinal instrumentations at L2-L4 level and vertebroplasty at L3 vertebra. AP view (E), Lateral view (F)

less frequent than metastases to the heart. The majority of cardiac malignant neoplasms in adults are sarcoma, particularly angiosarcoma, which is the most common in adults and has the worst prognosis.[2, 3] Rhabdomyosarcoma is considered to be the second most frequent primary cardiac tumor in adults and the most common in children which makes up approximately 20% of all primary malignant cardiac tumors.[4] Rhabdomyosarcoma can be classified into 3 main types including embryonal, alveolar, and pleomorphic. Even though patients with cardiac rhabdomyosarcoma can present with varieties of cardiac symptoms, they are usually at the advanced stage of the diseases. Tiyapant A., et al. reported the first case of cardiac rhabdomyosarcoma in Thailand with clinical presentation of cardiac tamponade.[5] The patients can also present with skeletal pain and bone lesions from metastases on radiographs without evidence of a primary tumor.[6] Transthoracic echocardiography remains the widely available screening method for the early diagnosis of a cardiac tumor. On the other hand, computed tomography (CT), magnetic resonance imaging (MRI), and positron emission tomography (PET) are demonstrate better soft tissue characterization of the neoplasm.[7]

Surgery is the method of choice for treatment of cardiac rhabdomyosarcoma in order to confirm the diagnosis, relief of cardiac symptoms, and prolongation of life. Heart transplantation can offer long-term survival and can be considered for selected patients.[8] Chemotherapy or radiotherapy should be considered for an adjuvant treatment. Patients with pleomorphic rhabdomyosarcoma are recommended to have adjuvant radiotherapy over the primary site, even after complete removal of the tumor.[9] Despite an aggressive treatment, adult rhabdomyosarcoma is still becoming a highly malignant tumor with a significant incidence of metastasis.[10] The overall 5- and 10-year survival rates are approximately 31% and 27% respectively.[11]

Skeletal metastasis from soft-tissue sarcoma is uncommon, regardless of extremely rare cardiogenic sarcoma in origin. Yoshikawa H, et al. reviewed 277 patients with soft-tissue sarcoma and then reported that 28 patients (10.1%) had metastases at an average of within 18.6 months after time of admission. Eighteen patients (64.3%) of those with metastases had lesions at spines and pelvic bones.[12] The median survival of patients with bone metastases in soft-tissue sarcomas is 6 months after diagnosis of bone metastases.[13] A pooled analysis from United States and European Cooperative Groups obtained data from 788 metastatic rhabdomyosarcoma patients and performed univariate analysis. They revealed that 3-year disease free survival was significantly and adversely affected by alveolar histology, age, and unfavourable site of the primary tumor. By multivariate analysis, disease free survival was strongly associated with all previous mentioned factors except histology.[14]

Surgical resection is one of the most important modality for the treatment of metastatic spinal neoplasm. Although en bloc resection is the standard treatment for malignant primary neoplasms of the spine, the concept of metastatic treatment is slightly different. Goals of surgery include restoration of neurological function, oncological control, pain control, and deformity correction and stabilization. Risks and benefits must be discussed among healthcare professionals, patients, and the family so that everyone has an understanding of the treatment and expected outcomes.[15] Rao G., et al. had studied 110 surgeries from 80 patients with either primary or metastatic sarcomas of the spine, of which 98 surgeries were intralesional resections (89%) and 11% were en bloc resections. Overall, the patients had a median survival time at 20.6 months, while the median survival for primary spinal sarcoma and metastatic sarcoma patients were 40.2 months and 17.3 months, respectively. Tumors with high grades have been identified from multivariate analysis as an adverse predictor of overall survival.[16]

Radiotherapy for spinal metastasis from soft-tissue sarcoma should be considered either for adjuvant after surgery or definite palliative treatment. Although higher doses of radiotherapy (35 fractions of 1.8 Gy) tend to have benefits for eradication of microscopic diseases without concerning of spinal cord damage in lumbar lesions; however, lower doses of 10-fractions of 3 Gy could be used for palliative purposes.[17] On the other hand, chemotherapy is not widely administered because this tumor is considered relatively chemoresistant but might have a role in selective cases. Bisphosphonate treatment, including pamidronate or zoledronic acid, could delay the first skeletal related event and might have a positive effect on overall survival.[13]

There were few literature reports about metastatic cardiac sarcoma at vertebras.[18-21] As far as we know, this is the first case report of metastatic cardiac pleomorphic rhabdomyosarcoma at lumbar spine. Although it is an extremely rare condition, it should always be considered in the differential diagnosis in patients who have history of sarcoma and present with spine lesions.

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รายงานผู้ป่วยมะเร็งทุติยภูมิที่กระดูกสันหลังส่วนเอวแพร่กระจายมาจากมะเร็งในหัวใจชนิดแรบโดมัยโอซาร์โคมา

ชินดนัย หงสประภาส, พบ, วิชาญ ยิ่งศักดิ์มงคล, พบ, พงศ์ศักดิ์ ยุกตะนันทน์, พบ

รายงานผู้ป่วยและบทความปริทรรศน์ของผู้ป่วยชายอายุ 78 ปีที่มีอาการหอบเหนื่อยมากขึ้นมา 1 เดือน ได้รับการวินิจฉัยว่าเป็นโรคมะเร็งในหัวใจชนิดแรบโดมัยโอซาร์โคมาแพร่กระจายมาที่ปอดและหนังศีรษะ ผู้ป่วยได้รับการผ่าตัดเอาเนื้องอกออก หลังจากนั้น 1 เดือนผู้ป่วยมีอาการปวดหลังอย่างรุนแรง ภายหลังจากการตรวจเพิ่มเติมโดยละเอียด พบว่ามีการแพร่กระจายของมะเร็งมาที่กระดูกสันหลังส่วนเอวและเนื้อเยื่ออ่อนข้างเคียง ผู้ป่วยได้รับการผ่าตัดอีกครั้งโดยมีผลการผ่าตัดเบื้องต้นเป็นที่น่าพอใจ แต่ผู้ป่วยเสียชีวิตในเวลาประมาณ 6 สัปดาห์ต่อมาเนื่องจากภาวะน้ำท่วมปอดและระบบทางเดินหายใจล้มเหลว

รายงานผู้ป่วยนี้เป็นฉบับแรกที่มีการรายงานถึงผู้ป่วยมะเร็งทุติยภูมิที่กระดูกสันหลังส่วนเอวแพร่กระจายมาจากมะเร็งในหัวใจชนิดแรบโดมัยโอซาร์โคมาซึ่งถือเป็นโรคที่พบได้ยากและมีความรุนแรงมาก

คำสำคัญ : *มะเร็งทุติยภูมิ, มะเร็งในหัวใจ, มะเร็งแรบโดมัยโอซาร์โคมาชนิดพลีโอมอร์ฟิก, กระดูกสันหลังส่วนเอว, รายงานผู้ป่วย*