Case Report

Spinal and Bilateral Breast Metastases of Embryonal Rhabdomyosarcoma

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A case of spinal and bilateral breast metastasis was reported. The primary tumor was an embryonal rhabdomyosarcoma of the left hand. Intralesional resection of metastatic spinal sarcoma was done with concurrence chemotherapy and radiotherapy. The literatures of these rare conditions were reviewed.

Keywords: *Metastatic sarcoma, Breast, Spine, Rhabdomyosarcoma*

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Metastatic sarcomas involving the spine are rare. Data in the literature regarding the treatment are limited[1].

Rhabdomyosarcoma is the most common soft tissue tumor in childhood.

This malignant tumor invades local structures and metastasizes to remote sites by lymphatic and hematogenous spread[2]. Metastases to the breast rarely occur and are mainly seen in adolescent girls. It sometimes has diagnostic problems. Early diagnosis is critical to prognosis and therapy[3].

The authors report a case of spinal and bilateral breast metastasis from embryonal rhabdomyosarcoma of the left hand and the literature is reviewed.

Case Report

A 16-year-old girl presented with a 5 cm. mass on the dorsum of the left hand first noticed 3 months earlier. Radiography showed an osteolytic lesion at the base of the first metacarpal bone (Fig. 1). Two months later, she developed back pain, numbness at anterior surface of both thighs and weakness of both hip flexors. In addition, there were multiple left cervical lymphadenopathy and mass at the left breast measuring 5 x 6 cm (Fig. 2). A biopsy of the hand lesion was obtained at the primary hospital. The histological examination confirmed round cell tumor. She was referred to Ramathibodi Hospital for proper investigation and management.

The patient was admitted. Investigation included a complete blood count, biochemical parameters, liver function test; these were all normal. Bone scan showed collapsed and increased uptake of L2 vertebral body. A CT chest and abdomen revealed diffuse pulmonary nodules, bilateral axillary and left cervical node enlargement. There were also bilateral multiple enhancing breast nodules, ranging from a few millimeters to 3.6 x 2.7 cm (Fig. 3). Plain radiography of the lumbar spine showed collapse of L2 vertebral body (Fig. 4). MRI of the whole spine was performed. There were multiple levels of spinal metastasis at C6, T1 and from T5 through S3 vertebral bodies (Fig. 5). The bone marrow aspiration revealed diffuse infiltrate non-hematologic malignancy, suggesting that there was metastatic CA involved bone marrow.

The patient underwent core biopsy from her left breast and posterior decompression and pedicular screws fixation at T12 to L3 (Fig. 6).The tissues were sent for histological examination and immunohistochemical study. The sections of the mass of the left hand showed small round blue cells with scanty cytoplasm. Immunohistochemical stains were positive for desmin, sarcomeric actin and negative for leukocyte common antigen, cytokeratin, HMB-45, leading to a

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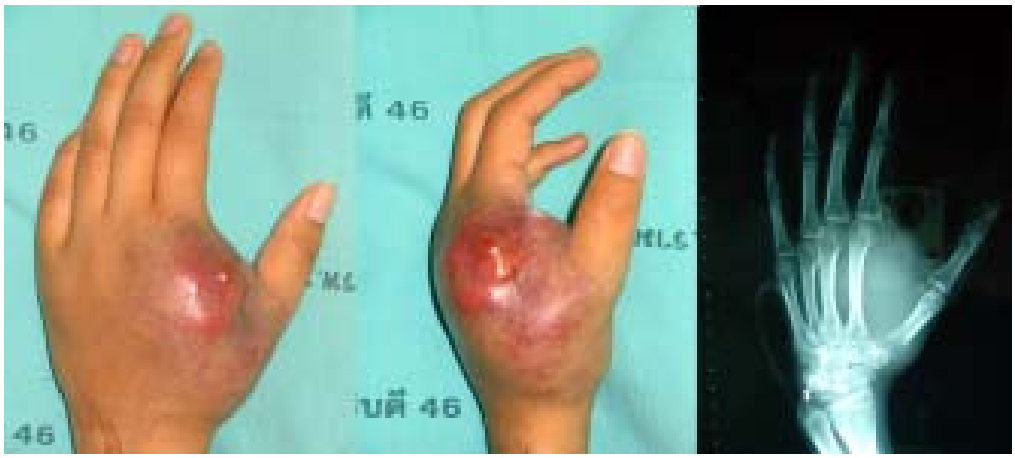

Fig. 1 The soft tissue mass and the radiography of left hand

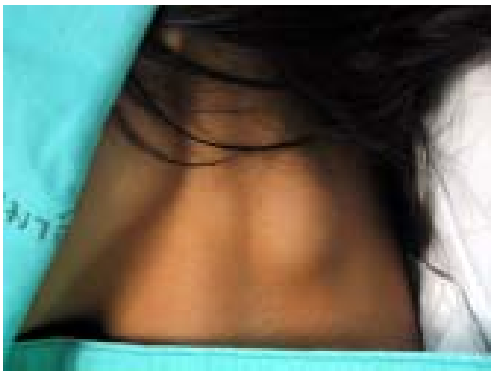

Fig. 2 Multiple left cervical lymphadenopathy

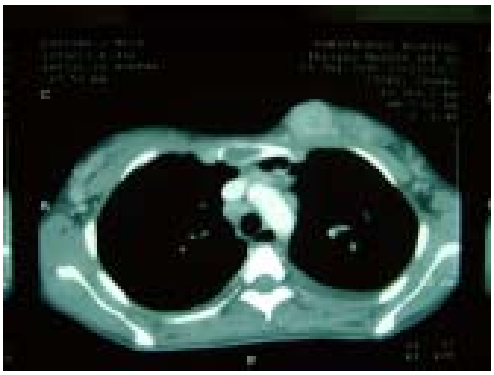

Fig. 3 CT chest at the level of breast masses

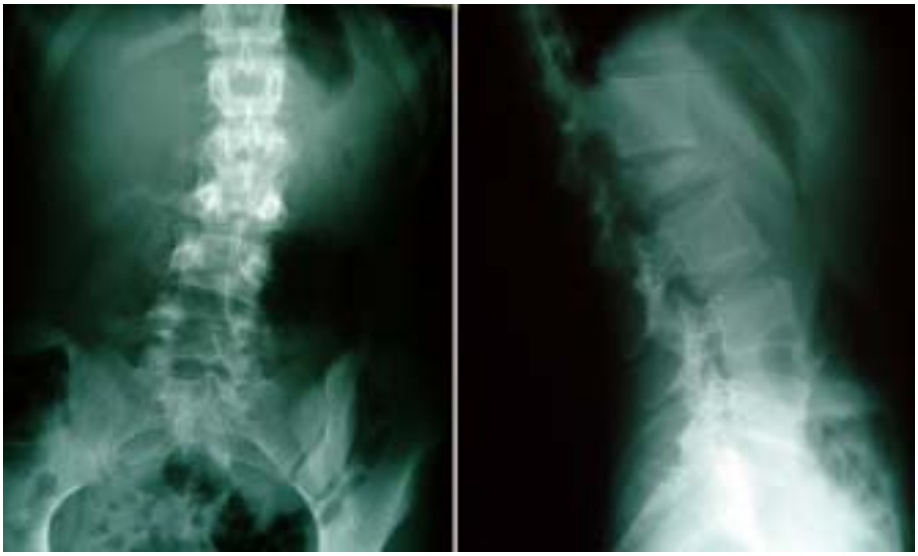

Fig. 4 Radiograph of lumbar spine preoperatively

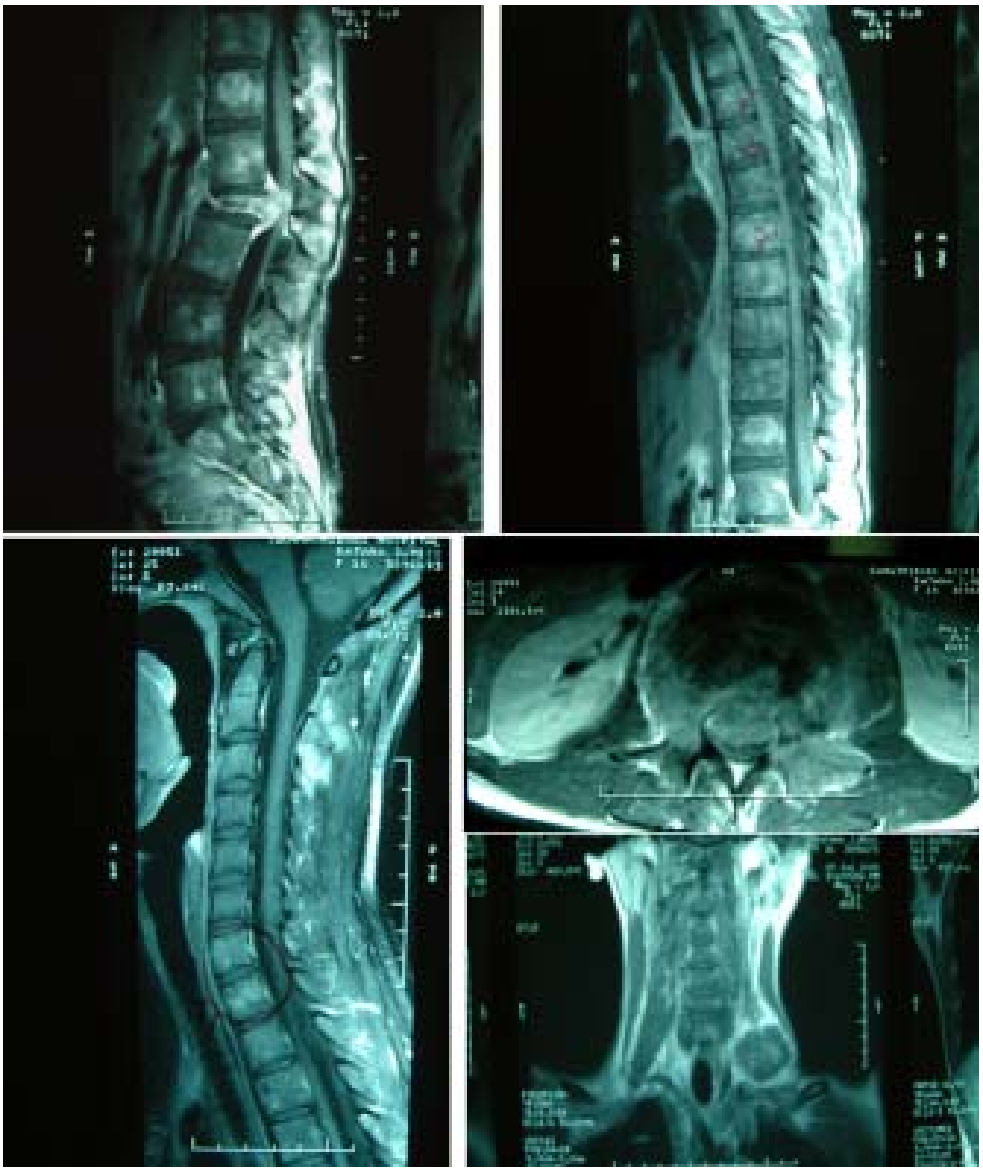

Fig. 5 MRI of the whole spine

diagnosis of rhabdomyosarcoma. The following incisional biopsy of the left breast showed non-cohesive groups of malignant small round blue cells, morphologically identical to the primary extremity tumor (Fig. 7, 8) The pathological diagnosis of all specimens confirmed metastatic embryonal rhabdomyosarcoma.

After surgery, the numbness at both thighs was improved and the muscle weakness returned to normal power. At the postoperative day 4, she could return to walk with a gait-aid. The radiotherapist was consulted for palliative radiation after the spinal fixation. Chemotherapy consisting of vincristine, adriamycin, and cyclophosphamide (VAC regimens) was administrated at two weeks after surgery. Five days after chemotherapy, she developed lower gastrointestinal bleeding and fever. The diagnosis was febrile neutropenia and thrombocytopenia. Although the proper antibiotics and symptomatic treatments were given, the patient's symptoms were not improved. Finally, she developed septic shock and she died at 3 weeks after surgery.

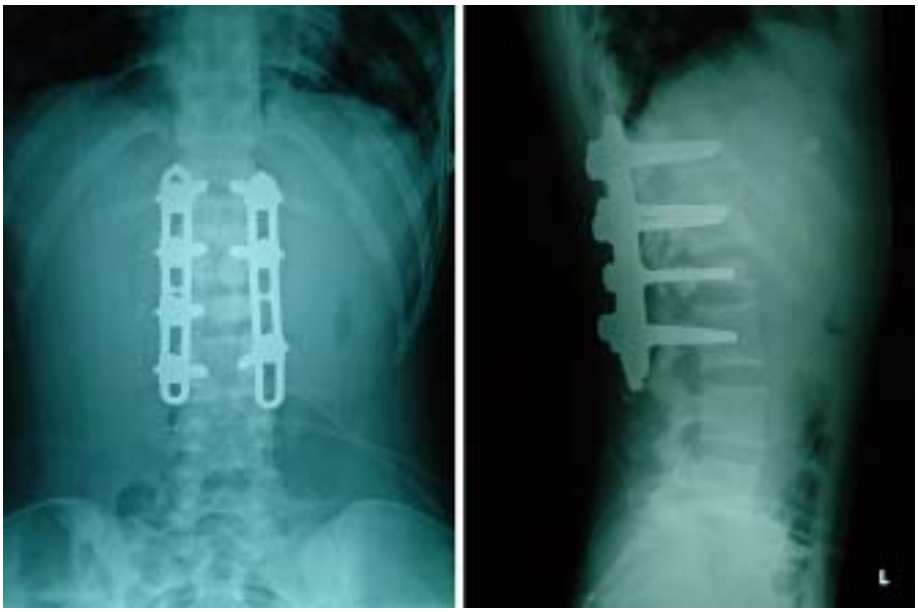

Fig. 6 Postoperative radiograph of the lumbar spine

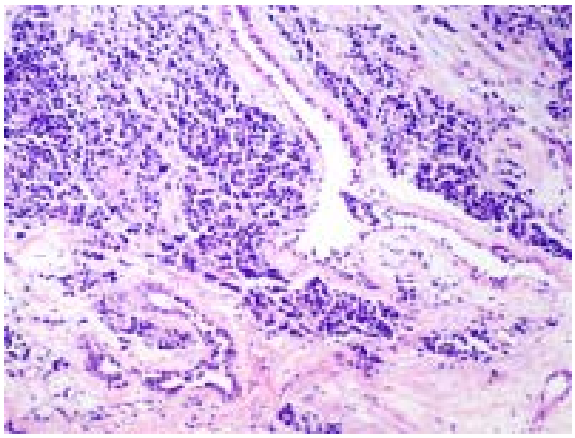

Fig. 7 The section of incisional biopsy of the left breast shows groups of malignant small round blue cells with scanty cytoplasm, morphologically identical to the primary extremity tumor, H&E (X100)

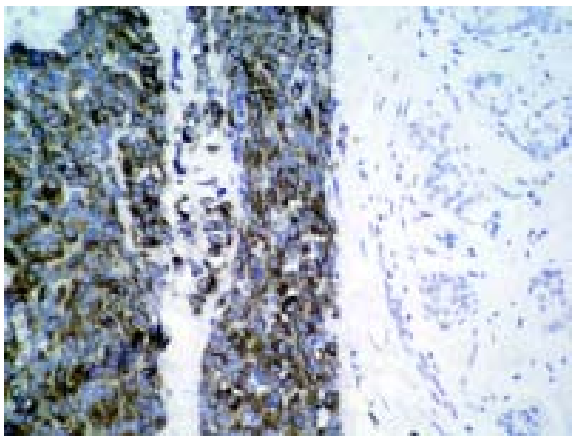

Fig. 8 The immunohistochemical stain for sarcomeric actin is positive in malignant small round blue cells (the left side of the picture) (X200)

Discussion

Rhabdomyosarcoma is the most common pediatric soft tissue sarcoma, comprising 50% of all soft tissue sarcomas and 10-12% of all malignant solid tumors in children[4]. Between 10-20% of patients with rhabdomyosarcoma will have metastasis at the time of diagnosis. This malignant soft tissue tumor can spread by three routes: direct invasion, lymphatic and hematogenous metastases[2,5]. The incidence of bony metastasis in rhabdomyosarcoma is not clearly defined. This tumor has no specific radiological features. Local bone invasion has been shown in 24% of cases, and bone metastases are typically ill-defined lytic lesions[5].

Primary and metastatic sarcomas of the spine are rare. Most are the case reports, and only a few series have reported the treatment outcome. Improvements in surgical techniques have resulted in better

decompression and gross total resection. In addition, the development of anterior instrumentation and posterior segmental fixation provide immediate stability.

The goals of surgery for patients with sarcoma involving the spine are improvement in neurological and functional status, relief of pain, local tumor control, and possibly cure. Although, En bloc resection with negative histological margins provides the best chance for local tumor control and long-term survival[6], Bilsky et al[1] report that fewer than 15% of patients had tumor patterns that were amenable to en bloc resection. They reported that the neurological and functional outcome of intralesional resection is excellent and comparable to those reported in series involving metastatic carcinomas in the spine. The drawback of this resection is high local recurrent rates, so radiation therapy is often used postoperatively in high-grade tumors.

Metastatic disease to the breast is unusual[7,8]. In children and adolescents, the secondary malignant tumors are more common than primary, most frequent being hematological (lymphoma, leukemia) malignancies and rhabdomyosarcoma[4]. Breast metastases from rhabdomyosarcoma are uncommon with an incidence of 6%[7]. Bilateral involvement is between 8 and 25%[9]. They occur mainly in adolescent girls with the most primary tumors being located in the extremities[3].

Alveolar type is the most common histological variant and there is strong association with breast deposits. These hematogenous metastases is believed to be due to increased vascularity and rapidly growing mammary tissue in the breast at puberty[3,8,10]. Disseminated disease is usually evidence at the time of diagnosis and the overall prognosis is poor. Early diagnosis in these patients has a major impact on prognosis and therapy[3].

Physical examination usually indicates benign painless, well-circumscribed, and freely movable breast lumps. Solitary lesions with nodular appearance are more frequent than multiple and diffuse involvement[3,4].

Rhabdomyosarcoma of the extremities have been reported as more aggressive. When arising in the hand or foot, they have a higher incidence of bony erosion and unusual anatomic sites of metastases, including breast, ovary, testis, pancreas, and kidney[11]. In the published cases of rhabdomyosarcoma of the hand or foot, 81-87% were alveolar subtype and they display aggressive clinical behavior.

In the presented case, the patient presented with a soft tissue mass and osteolytic lesion of the first metacarpal bone of the left hand. The histology confirmed embryonal subtype, although most cases in the literature arising in the hand or foot were the alveolar type. In the present case, she developed breast and spinal metastases. Intralesional resection and pedicular fixation were selected because of invasion of sarcoma to both pedicles and posterior element. The oncologist and radiologist were consulted for chemotherapy and postoperative radiation, respectively.

Conclusion

From the literature review, intralesional resections for metastatic spinal sarcoma are the appropriate palliative procedures although these have poor local control rates. En bloc resection is feasible in a small subset of patients. The authors found the association between rhabdomyosarcoma of hand or foot, breast metastases, and alveolar subtype. Therefore, patients presenting with a previous history of rhabdomyosarcoma, especially arising in the hand or foot of adolescent girls, should be thoroughly evaluated for occult metastases in the breast and other unusual sites[10,11]. Conversely, the possibility of a small primary rhabdomyosarcoma of the hand or foot should be investigated in patients presenting with metastatic rhabdomyosarcoma in these unusual sites.

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มะเร็งกระจายไปยังกระดูกสันหลังและเต้านมทั้ง 2 ข้าง โดยมีมะเร็งปฐมภูมิจากเนื้องอกแรบโดมัยโอซาร์โคมา

กันต์ แก้วโรจน์, พงศธร ฉันท์พลากร, วิเชียร เลาหเจริญสมบัติ, นภดล ลาภเจริญทรัพย์

รายงานผู้ป่วยมะเร็งกระจายไปยังกระดูกสันหลังและเต้านมทั้ง 2 ข้าง โดยมีมะเร็งปฐมภูมิจากเนื้องอกแรบโดมัยโอซาร์โคมา ชนิดเอ็มบริโอที่มือซ้าย ผู้ป่วยได้รับการรักษาผ่าตัดขูดเนื้องอกกระจายที่กระดูกสันหลังออกร่วมกับการให้ยาเคมีบำบัด และฉายรังสีรักษา จากนั้นทบทวนวรรณกรรมในภาวะนี้