Neurological Deficit from Metastatic Low Grade Liposarcoma

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Myxoid liposarcoma is a malignant soft tissue tumor with a relatively indolent natural history. It commonly occurs in the extremities with extrapulmonary metastatic potential. Round cell liposarcoma, recently, is widely agreed to be the aggressive counterpart of the myxoid liposarcoma, mostly arising in adverse, local recurrence, and metastatic lesions. Metastatic liposarcoma with pure myxoid element is unusual. The authors report an extremely rare case of a 47-year-old Thai male patient having a metastatic pure myxoid liposarcoma to the thoracic vertebrae, one year after primary tumor documented on the left thigh. Histology of the primary and metastatic sites shows only myxoid pattern. Round cell feature was not documented. To the best of the authors' knowledge, this is the second case report of metastatic myxoid liposarcoma without round cell morphology.

Keywords: Myxoid liposarcoma, Round cell liposarcoma, Neoplasm metastasis

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Myxoid liposarcoma is the well-differentiated sarcoma mostly arising in the extremities^(1,2). This tumor usually metastasizes to bones and other tissues without pulmonary involvement. To date, it is accepted that round cell morphology is the spectrum of myxoid liposarcoma with poor prognosis. Round cell element should be found in aggressive, recurrence, or metastatic myxoid liposarcoma^(3,4).

Metastatic myxoid liposarcoma without round cell element is very unusual. The authors report herewith the extremely rare case of pure myxoid liposarcoma metastasizing to the multiple thoracic vertebral bones with neurological deficit in a 47-yearold Thai man.

Case Report

A 47-year-old male with a 7-year history of schizophrenia experienced a mass on the left thigh for several weeks. He came to the King Chulalongkorn Memorial Hospital because the mass was rapidly growing. The physician diagnosed him to have a soft tissue tumor and, then, the patient underwent marginal excision. The surgical specimen revealed a large soft tissue mass, measuring 20 x 12 x 15 cm, partially covered by striated muscle. Serial sections demonstrated nodular, soft, and gray yellow with myxoid cut surfaces (Fig.1). The tumor was also grossly close to surgical resected margins. The histology showed plumped spindle cells with scant cytoplasm in the myxoid matrix. Multivacuolted lipoblasts commonly occurred. The background disclosed typical and prominent arborizing thin-wall capillary networks orga-



Fig. 1 Tumor excised from the left thigh. Cut surface is nodular, with gray yellow and myxoid appearance

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nized in a plexiform pattern (Fig. 2, 3). Hemorrhage and necrosis were rarely demonstrated. Undifferentiated round cells were not seen in the additional extensive sampling sections. Pathological report was myxoid liposarcoma, grade 1, according to the United States National Cancer Institute (NCI) grading system. For TNM staging, the patient was classified as stage IIb (T2N0Mx). Unfortunately, the patient was lost to follow-up postoperatively.

One year later, he had gradually increasing low back pain and numbness on both knees and legs for a month. The patient was taken to the King Chulalongkorn Memorial Hospital again because he developed sudden nocturnal back pain, was not able to walk, had urinary urgency, and fecal incontinence. Physical examination revealed sensory loss on the chest including upper abdominal wall, decreased muscle power of the lower extremities with spasticity, and lax anal tone.



Fig. 2 Arborizing vasculature and lipoblasts set in the myxoid background



Fig. 3 Lipoblasts containing multiple vacuoles in the cytoplasm with pleomorphic nuclei

Emergency magnetic resonance imaging (MRI) was performed and showed multiple osteolytic lesions with spinal cord compression at level T4-T12. Chest x-ray was clear. Decompressive laminectomy was done. The histological findings expressed similar feature as aforementioned without round cell pattern in all representative sections. Local recurrence was not identified on the left thigh.

Discussion

Myxoid liposarcoma is the second most common subtype of the malignant adipose tissue tumor after well-differentiated liposarcoma, accounting for approximately 30% to 40%^(1,2). Clinically, they tend to occur in the lower extremities with a peak incidence ranging between the third and the fifth decade. If completely excised, they prone to locally recur. However, the metastatic rate is approximately 30%⁽³⁾. Extremity myxoid liposarcoma have a unique high incidence of extrapulmonary metastases with unknown reason. A peculiar tendency to metastasize to the soft tissue is observed and should not be interpreted as multicentricity^(4,5).

According to the World Health Organization, it is widely agreed that round cell liposarcoma, a highgrade malignancy, is a poorly differentiated counterpart of myxoid liposarcoma, which is low grade⁽⁶⁾. Smith et al has been studied the clinicopathologic correlation of myxoid/round cell liposarcoma on the extremities and found that the patients with round cell component of more than 5% in their initial tumor had a statistically significant by higher rate of metastasis or death due to disease⁽⁷⁾. Fukuda et al detected that round cell lesions were the potentially malignant elements, regardless of their proportion. Round cell component is found increasingly in the recurrence and metastatic tumor⁽⁸⁾.

Regarding to the present patient, marginal excision alone was unsatisfactory. Adjuvant chemotherapy or post-operative radiation had to be done for decreased risk of local recurrence and metastasis. Wide excision is the appropriate surgical procedure, if it does not disturbed anatomic relation and organ function. After treating the primary tumor, the following periodic chest and abdominal imaging studies should be considered, including additional spinal region correlated with neurological complaints.

However, our patient was lost follow up and presented with multiple vertebral bone metastasis one year later. Local recurrence at the primary site was not occurred. The histologic feature of the primary tumor of the left thigh and vertebral metastatic lesions showed pure myxoid liposarcoma without evidence of adverse round cell component. This is an exceedingly rare clinicopathologic feature. To the authors' knowledge, this is the second case report of metastatic pure myxoid liposarcoma. The first case was published by Ogose A et al⁽⁴⁾ in 2001, that patient experienced intraspinal extradural metastasis at T8-T11 after diagnosis of myxoid liposarcoma in the left forearm.

In conclusion, the authors describe the unusual feature of metastatic myxoid liposarcoma to vertebral bones with spinal cord compression, one year after the primary site was documented on the thigh. Microscopically, it showed a pure myxoid feature, which had a well differentiated form, without high-grade round cell morphology.

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ระบบประสาทผิดปกติจากมะเร็งเนื้อเยื่อไขมันเกรดต่ำที่แพร่กระจาย

มานะ ทวีวิศิษฏ์, วรนุช ธนากิจ

ตามธรรมชาติของมะเร็งเนื้อเยื่อไขมันแบบมิกซอยด์นั้นมักจะลุกลามไม่รุนแรง มะเร็งชนิดนี้เกิดขึ้นบ่อย บริเวณแขน ขา และมักแพร่กระจายไปตามเนื้อเยื่อต่างๆของร่างกายโดยที่ไม่มีรอยโรคที่ปอด ปัจจุบันเป็นที่ยอมรับแล้วว่า มะเร็งเนื้อเยื่อไขมันแบบเซลล์รูปร่างกลมก็คือกลุ่มมะเร็งเนื้อเยื่อไขมันแบบมิกซอยด์แบบหนึ่งที่มีการทำนายโรคที่เลว มักพบในพยาธิสภาพที่รุนแรง, เกิดเป็นซ้ำ, หรือรอยโรคที่แพร่กระจาย รายงานผู้ป่วยมะเร็งเนื้อเยื่อไขมันแบบมิกซอยด์ 1 รายที่มีลักษณะแปลกออกไปและพบได้ยาก ผู้ป่วยเป็นซายไทยอายุ 47 ปี ได้รับการวินิจฉัยว่าเป็นมะเร็งเนื้อเยื่อไขมัน แบบมิกซอยด์ที่ต้นขาซ้าย และในเวลา 1 ปีต่อมา มะเร็งมีการแพร่กระจายไปยังกระดูกสันหลังระดับอกหลายตำแหน่ง จนเกิดอาการผิดปกติทางระบบประสาท จุลพยาธิวิทยาของมะเร็งเริ่มแรกและที่ลุกลามแพร่กระจาย พบแต่ลักษณะมิกซอยด์ ไม่พบลักษณะการเปลี่ยนแปลงเป็นเซลล์รูปร่างกลมแต่อย่างใด ลักษณะที่แปลกของ มะเร็งดังกล่าวพบในผู้ป่วยรายนี้เป็นรายที่ 2 จากการทบทวนวารสารที่ผ่านมา