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Case report

Endoprosthetic reconstruction for metastatic phaeochromocytoma in the distal femur: A case report

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ABSTRACT

Metastatic spread of malignant phaeochromocytoma is known to involve multiple organs including the axial skeleton. Its presentation as a solitary lesion in the long bones of the extremities is extremely rare. We report a unique case of solitary metastatic phaeochromocytoma presenting in the distal femur, 16 years after excision of primary abdominal tumor.

A 60 year old female, operated for adrenal phaeochromocytoma 16 years back was detected to have a bone tumor in her left distal femur. Chest and abdominal CT evaluation including bone scintigraphy confirmed the lesion to be solitary. Magnetic resonance imaging of the left femur revealed the tumor to be of aggressive nature. It involved whole of the distal femoral metaphysis with bone destruction, soft tissue extension and an impending pathological fracture. The tumor was histopathologically confirmed to be phaeochromocytoma. Patient underwent wide surgical resection of the tumor along with simultaneous endoprosthetic reconstruction. Following surgery, rehabilitation was rapid and effortless with patient ambulating independently. She had pain free full range of knee movements and resumed her daily activity uneventfully. On 18 months follow up the patient had no recurrence or complications.

Late and solitary skeletal metastasis of malignant phaeochromocytoma although rare, can occur in the distal femur mimicking a primary bone tumor. Wide surgical excision and simultaneous endoprosthetic reconstruction should be considered as a treatment option. This not only permits limb salvage and early rehabilitation, but also restores form and function of the limb.

1. Introduction

Malignant variant of phaeochromocytoma is rare and seen in about 10% of cases diagnosed with phaeochromocytoma.[1] Metastasis is often to multiple organs and commonly involves the axial skeleton.[2] Its presentation as a solitary metastatic lesion in the long bones is extremely rare and there is paucity of literature regarding management of such cases.[3,4] We report one such case of malignant phaeochromocytoma presenting 16 years after excision of primary tumor. This patient had a solitary metastatic lesion in the left distal femur. It was managed by wide surgical excision along with reconstruction of the distal femur and knee joint using modular endoprosthesis.

In this report, we stress the importance of considering wide surgical excision and simultaneous endoprosthetic reconstruction in patients with solitary pheochromocytoma.

2. Case report

A 60 year old female presented with pain and swelling of her left lower thigh and knee for six months. The pain was dull, localized, present at rest and aggravated on walking. It disturbed her sleep and restricted her daily activities. Walking distance was gradually reduced because of pain. She had noticed the swelling in her lower thigh with insidious onset and gradually increased to a size of about 10 cm in its largest diameter. The swelling restricted her terminal knee flexion causing difficulty in squatting. Patient had abdominal phaeochromocytoma in the past for which she had undergone adrenalectomy and nephrectomy 16 years back. She had not received any adjuvant therapy but was on regular follow up for initial few years. At her last follow up 5 years back,

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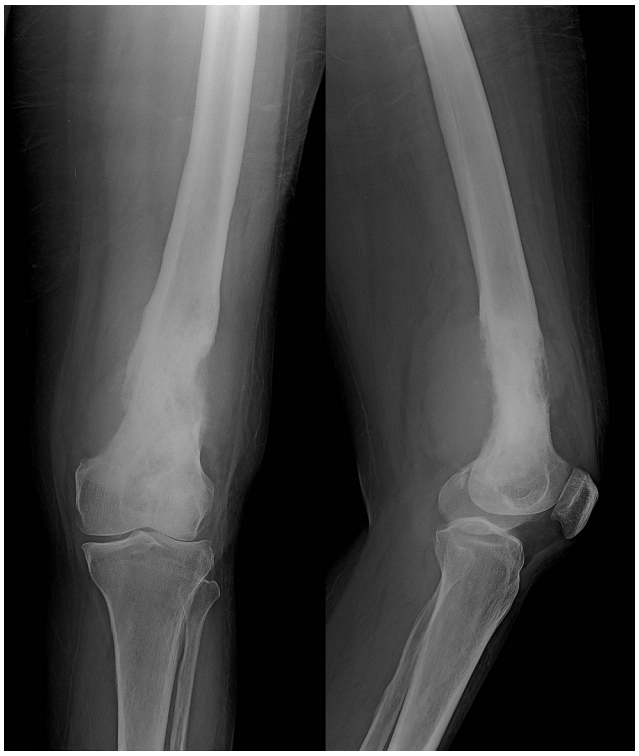

Fig. 1. Radiograph of femur with knee shows malignant bone tumor in the distal femoral metaphysis.

abdominal computed tomography (CT) and metaiodobenzylguanidine (MIBG) scans had ruled out presence of primary recurrence or secondary metastases with normal urinary vanillylmandelic acid (VMA) levels. Patient was a known diabetic and had poorly controlled hypertension.

On examination, patient had an antalgic gait with diffuse swelling over the anteromedial aspect of the left lower thigh and knee extending posteriorly. Skin over the swelling was stretched with visible prominent veins. Swelling had differential warmth and was firm to hard in consistency. It was fixed to the underlying bone but free from overlying skin. Knee had 10° of extension lag and free painless flexion up to 110°. There were no associated distal neurovascular deficits. Radiograph of the left femur including knee (Fig. 1) revealed an irregular predominantly sclerotic lesion involving the whole of the distal femoral metaphysis. The lesion had a permeative margin, cortical saucerization, sunray spiculed periosteal reaction and soft tissue extension suggestive of a malignant bone tumor. Magnetic resonance imaging (MRI) of the whole femur and knee (Fig. 2) showed an aggressive bone lesion involving the distal femur. It was seen extending from the femoral condyles up to 13 cm above knee joint without any skip lesions and with predominant posterior and medial soft tissue extension. CT scan of chest and abdomen confirmed absence of primary recurrence or secondary metastases. Whole body MIBG and technetium 99 m methylene diphosphonate (99mTc-MDP) scintigraphy revealed only a solitary lesion at the lower end of left femur (Fig. 3). Urinary VMA levels were within normal range. Core needle biopsy of the tumor was performed and histopathology studied. It showed polygonal cells with granular cytoplasm and

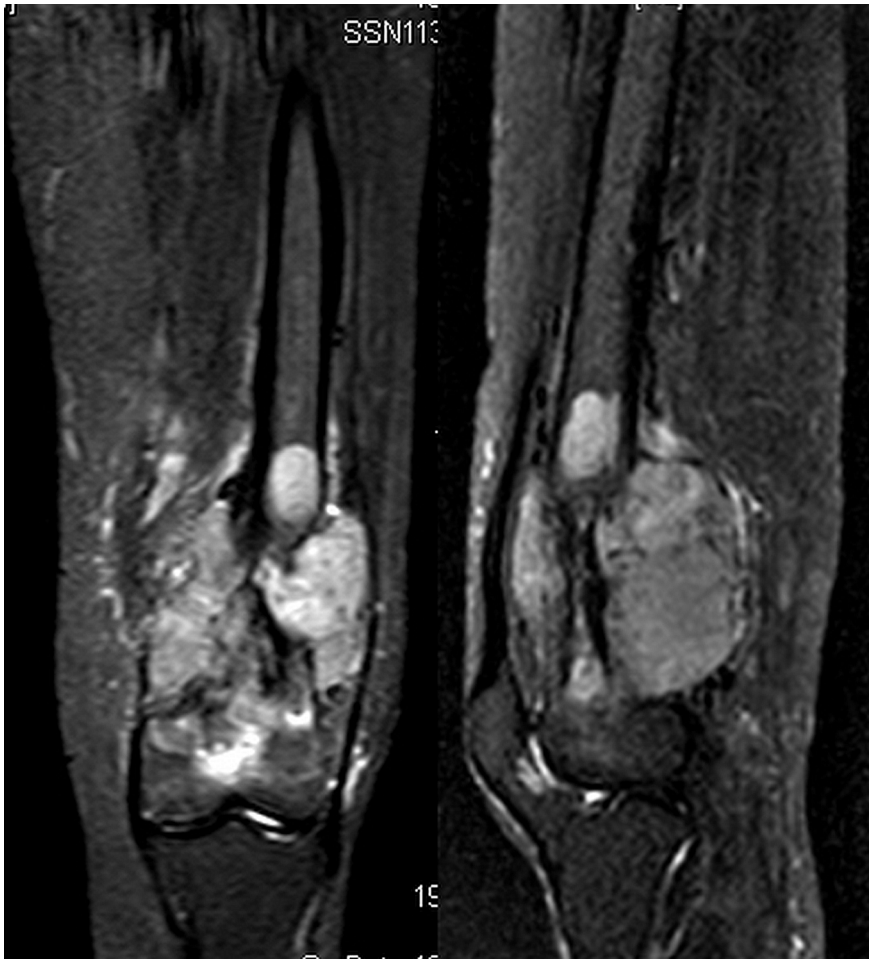

Fig. 2. MRI of the femur with knee shows intramedullary and soft tissue extent of the tumor.