POSTERIOR FOSSA BRAIN METASTASIS FROM PRIMARY OSTEOGENIC SARCOMA: A RARE ENTITY

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Introduction

The most common primary tumors associated with brain metastases are melanoma, lung, breast, kidney and colon carcinoma.[1] The occurrence of metastasis in childhood is more unusual, and only a few reports have described their clinical and radiologic features. Brain metastasisis a very rare consideration in osteogenic sarcoma. It has become a new challenge for pediatric oncologist due to change in the natural history of pediatric malignancies as a result of the introduction of effective systemic chemotherapy. Brain metastasis in osteosarcoma represents advanced stage of the disease and is reported to have a poor prognosis. We report a rare entity of posterior fossa brain metastasis from primary osteogenic sarcoma in a 16 year old boy along with pulmonary metastasis.

Case Report

A 16 year old boy presented with pain and swelling of the left knee. He also had shortness of breath. Plain radiograph at presentation showed an ill definedosteolyticlesion in the distal left femur with an aggressive pattern causing cortical erosion and periosteal reaction. Magnetic Resonance Imaging (MRI) demonstrated a large infiltrating tumor involving the distal end of the femur with a large soft tissue component with areas of necrosis in it (Figure 1). It was diagnosed to be osteogenic sarcoma on needle biopsy.

Concurrent contract enhanced CT Scanof the Chest showed multiple pulmonary nodules of varying sizes scattered in both lungs representing pulmonary metastases (Figure 2).Amputation of the affected limb was done and six cycles of palliative chemotherapy was given.

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Patient remained asymptomatic for 26 months and then presented with severe headache and vomiting. CT Scan of the brain with contrast revealed two soft tissue density space occupying lesions in the posterior fossa showing peripheral enhancement (Figure 3).For further characterization,contrast enhanced MRI brain was performed. MRI showed a large well defined rounded enhancing lesion involving cerebellar vermis and another small enhancing lesion in left cerebellar hemisphere close to left cerebello-pontine angle(Figure 4a, 4b) measuring 3.2 cm x 2.8 cm and 1.0 cm x 0.9cm respectively. These were associated with mild supra-tentorial hydrocephalus. In view of the history of known primary tumor,differential diagnosis ofmetastasesand primary tumor was made. Biopsy of the lesions confirmedbrain metastases. Due to progressive nature of the disease,patient died within three months.

Discussion

Osteogenic sarcoma is the most common osteoid producing malignant bone tumor[2].It usually occurs in children and young adults, predominantly involving metaphysis of long bones. Distal femur, proximal tibia and humerus are the most common sites involved. Axial skeleton is rarely involved and accounts for about 10%[3].Hematogenous dissemination is typical, primarily affecting lungs and bones.Involvement of the brain in sarcoma patients occur in 1-8% of the cases. Three percent of the brain metastasis is found to be from primary sarcoma[4,5].In the literature review, only 11 cases of primary intracerebral or meningeal osteogenic sarcoma has been reported without any skeletal involvement[6].Brain metastasis in primary osteosarcoma is rare with reported incidence of 1.8 – 5.6%[7].The two hypothesis regarding spread of sarcoma to brain arehematogenous dissemination and contiguous spread through skull bones into intracranial structures. There is another hypothesis of lung tumor emboli invading the brain in patients with prior pulmonary metastasis[8].

A large study evaluating brain metastasis in musculoskeletal sarcomas conducted by Akira Ogose et al[9]. detected only three cases of brain metastasis in 103 osteosarcoma patients. Of these, two had prior pulmonary and one had retroperitoneal metastasis. RejinKabudi et al7.detected brain metastasis in five of the patients with osteosarcoma and all had prior pulmonary metastasis.

Brain metastasis from osteogenic sarcoma typically occurs through anterior circulation involving grey white matter junction of supratentorial compartment[10].Uptillnow, 56 cases of osteosarcoma with brain metastasis have been published including this case. Location of metastasis varied and mostly occurred in cerebrum with most common involvement of frontal lobe. Only two cases were reported to involve the posterior fossa. Niazi et al[11]. reported metastasis in left cerebellar hemisphere in a 16 month old child presented with posterior fossa hemorrhage with a survival ofonly 6 days. Wroński et al[12].described posterior fossa metastasis along with cerebral metastasis. This case is the third reported case involving the posterior fossa (cerebellum) again with bad prognosis.

Brain metastasesrepresents advanced stage of the disease and estimated time interval varies between 20 – 30 months from presentation. It occurred after 24 months in the index case. Patients usually present with neurological signs and symptoms like headache, vertigo, paresthesia and visual field defects. Comprehensive neurological evaluation must be performed in these patients.Yonemoto et al.[13]and Marina et al[14].recommended periodic brain imaging for early detection and surveillance in

patients with metastatic disease at diagnosis or recurrence less than 12months. All patients with pulmonary metastasis must be evaluated for brain metastasis. It has been reported that the incidence of extra-pulmonary metastasis has been increased with the introduction of systemic chemotherapy as it prolongs the survival time of the patient resulting in higher incidence of brain metastasis[15].Therefore, CT Scan or MRI brain should be periodically performed in patients with pulmonary metastasis for early detection.

Treatment of brain metastasis is also challenging. Chemotherapy is not so beneficial as most of the patients have already been treated with chemotherapy. Patients with poor medical condition and distant metastasis may benefit from Radiation. Stereotactic radiosurgery may be used. Radiosurgery is also a safe and effective method for inoperable lesions[9].

Conclusion

The occurrence of Brain Metastasis in patients with bone and soft tissue cancers is a poor prognostic sign that suggests late stage disease. High degree of suspicion of brain metastasis is necessaryin all patients of osteogenic sarcoma with prior intrapulmonary metastasis. Properfollow up with neurological assessment and radiological workupwhen necessary is the key to early diagnosis and better survival of patients.

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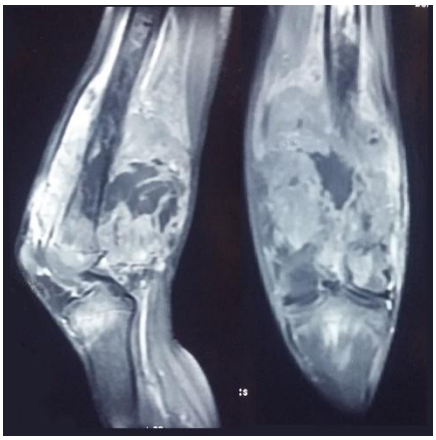

Figure 1: Sagittal and Coronal images of contrast enhanced MRI of left Knee joint demonstrating a large infiltrating tumor involving the distal end of the femur with a large soft tissue component showing enhancement and areas of necrosis in it

Figure 2: Axial section of CT Chest with contrast on lung window setting showing a well defined rounded pulmonary nodule in left lung suggestive of intra pulmonary metastasis

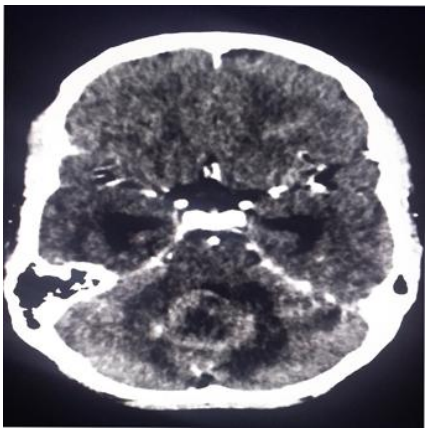

Figure3: Axial section of CT Brain with contrast demonstrating two soft tissue density space occupying lesions in the posterior fossa showing peripheral enhancement

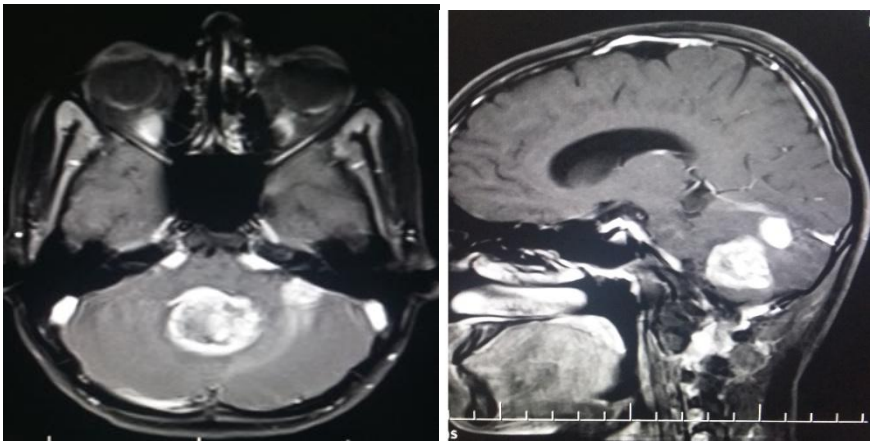

Figure 4: Axial and Sagittal images of MRI Brain with contrast demonstrating two well defined rounded enhancing lesions involving cerebellar vermis and left cerebellar hemisphere close to left cerebello-pontine angle representing brain metastasis.