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Published online 2022 June **Case Report**

Primary malignant melanoma of bone (clear cell sarcoma): A case report

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Abstract:
Melanoma is a malignant tumor that predominantly arises in the skin. In rare cases, however, it can manifest in the bone too. In this paper, a primary malignant melanoma of the lesser trochanter of the right femur is described.
In this case, the subject was a 19-year-old woman presenting with pelvic floor pain that lasted for 3 weeks.
Magnetic resonance imaging (MRI) showed that T2MRI has bone melanoma soft tissue edema that sustained to malignant melanoma.
Immunohistochemistry report revealed that tumoral cells were positive for HMB45, S100, which is suggestive of melanoma diagnosis.
The patient was admitted and underwent CNB. Since the result was negative, the patient received a wide local excision from the right lesser trochanter.
The histopathological examination of biopsy revealed fibro sclerotic tissue affected by malignant melanoma.
The patient was then referred to an oncologist for postoperative adjust chemotherapy and target therapy.

Keywords: case report- melanoma- clear cell sarcoma

Introduction

Melanoma is the most prevalent and most known form of cancer in the melanocyte cell, which often affects the skin and is regarded as a skin cancer with a high mortality rate. (1-4)

The third most common skin cancer, melanoma originates from neural crest cells on the neural tube (2, 5) which usually migrate to the basal layer of the epidermis in the skin, though some may migrate elsewhere such as urea, meninges, and mucosal surface (3).

The malignant proliferation of melanocytes often takes place in the skin, but it may appear in other organs including the esophagus, oral cavity, eyes, meninges and urogenital mucosal.

However, it can disseminate and metastasize to multiple sites including lung and bone (3).

Melanoma metastasis in the bone is not uncommon (2,6-7). It has been reported that 23 to 40% of malignant melanoma metastasis spreads to the bone (3).

The most common sites of bone affected are ribs and the spinal column (8). The disease can change the bone, leading to an increased risk of bone fracture (9).

While more than 90% of malignant melanoma has a cutaneous origin (10), primary osseous melanoma is a very rare variant of melanoma (2).

In the present report, a rare case of primary melanoma arising from the lesser trochanter is described in an adult female.

Case report:

The patient was a 19-year-old woman who presented with pelvic floor pain that lasted for 3 weeks. She had no history of trauma, infections or constitutional symptoms.

The examination did not reveal anything suspicious and the patient had no localized tenderness or limited range of motion at the hip joint. The physical examination also did not reveal any evidence of a skin lesion in her body. Moreover, no significant abnormality was observed in the plain radiography of bone(Fig.1)

Magnetic resonance imaging (MRI) indicated that T2MRI has bone melanoma, soft tissue edema that has transformed into malignant melanoma (Fig.2). The patient was referred for a CT-guided core needle biopsy. However, the CT-guided biopsy was negative for malignancy (false negative).

Accordingly, the MRI finding was interpreted as a tumor arising from the right lesser trochanter with soft tissue edema.

Computed tomography (CT) showed destructive lesions in the lesser trochanter, which in differential diagnosis were interpreted as osteoid osteoma, osteoblastoma, and malignant lesion(Fig.3).

We resected the lesser trochanter and the old tumoral lesion and investigated the pathology of

melanoma. The imaging revealed a fracture with an abnormal destructive lesion with an intramedullary extension, but no reactive sclerosis or periosteal reaction was identified.

Hence, differential diagnosis pointed to osteoid osteoma, osteoblastoma or Eg.

Immunohistochemistry (IHC) report showed the tumoral cells were immunoreactive positive HMB45, S100, Maogenine, TLE1, ki67, melan, and FL11 but they were negative for CD99, LCA, CKEA1, AE3, CD56, CD20, CD10, synaptophysin, TTF1, SALLa, SMA, OCT314 Desmin.

IHC findings suggested that S100 and HMB45 were strongly positive which indicated the diagnosis of malignant melanoma. This finding was with the diagnosis. Changes had all hallmarks of malignant melanoma. The patient was then admitted and underwent CNB. Since CNB was negative, the patient underwent a wide local excision in the right lesser trochanter.

CNB pathology report revealed hypocellular mass without evidence of neoplasm or inflammatory process. The specimen received in formalin consisted of multiple tan-brown soft and boney tissue fragments measuring 3×3×1 cm in length.

The histopathological examination of the biopsy specimen revealed fibrosclerotic tissue affected by malignant neoplasm consisting of a large cell with large nuclei, spacious red nucleoli and extensive pale eosinophilic cytoplasm arranged in a nest (Fig.4).

Some cells showed granular brown intracytoplasmic pigments. In some areas, crush artifacts with hyperchromic nuclei and scan cytoplasm were noted. The change was suggestive of malignant melanoma, but there was insufficient data on whether the tumor was primary or metastatic; therefore, the clinical study was recommended.

Though less likely, the primary clear sarcoma of bone could be considered as a differential diagnosis. A whole-body scan showed active lesions and differential diagnosis revealed metabolic and metastasis lesions.

The bone scan was performed in three phases. Finding bone scan was focal area of hyper flow hyperemia and delayed hyperactivity was noted in the lesser trochanter of the right femur.

There were also areas of increased focal radiotracer accumulation in the right side of the mandible, the left humeral head, the posteromedial aspect of the 7th rib on the left, and the anterior 5th rib on the left in the acetabulum.

No significant abnormality was detected in the rest of the skeletal system. An active abnormality lesion was reported in the lesser trochanter of the right femur.

In other bone lesions in the right side of the mandible, the posteromedial aspect of humeral head fracture on the left 7th rib, anterior rib and probably the right acetabulum were observed. Hence, a tissue exam was recommended to exclude primary or metastatic lesions or metastatic bone lesions.

Positron Emission Tomography - Computed Tomography (PET/CT) (PET/CT) was performed to rule out possible metastasis of malignant melanoma in the bone.

PET/CT report of chest, abdomen, and pelvic was compatible with FDG-avid metastatic disease in: the right soft tissue lesion in the maxillary sinus, multiple bilateral pulmonary masses and nodules, soft tissue nodules in the right breast, two chest-wall subcutaneous soft-tissue lesions, bilateral adrenal masses, multiple skeletal lesions. There was a non-FDG avid lesion in the right cerebellum.

To rule out metastatic disease, correlation with brain MRI is recommended. Clinical laboratory findings were essentially normal. The analysis showed local disease, a therefore a wide excision of lesser trochanter tumor was performed.

Intraoperatively, the tumor appeared to originate from the right lesser trochanter. Because there was no clinical evidence of lymph node involvement, the central node biopsy was not performed.

In addition, neoadjuvant chemotherapy was not administered for this patient. The patient could not recall any previous skin lesions and the evaluation of the total body skin was negative. The patient was referred to an oncologist for postoperative adjust-chemotherapy and target therapy. It is notable, after chemotherapy, when the patient lost her hair, a mole was seen on the patient scale, which cloud have been the primary source of the tumor.

She was followed up in our clinic for 12 –months. She eventually died of brain complication and multiple metastases 12 months after the initial diagnosis of melanoma in the right lesser trochanter.

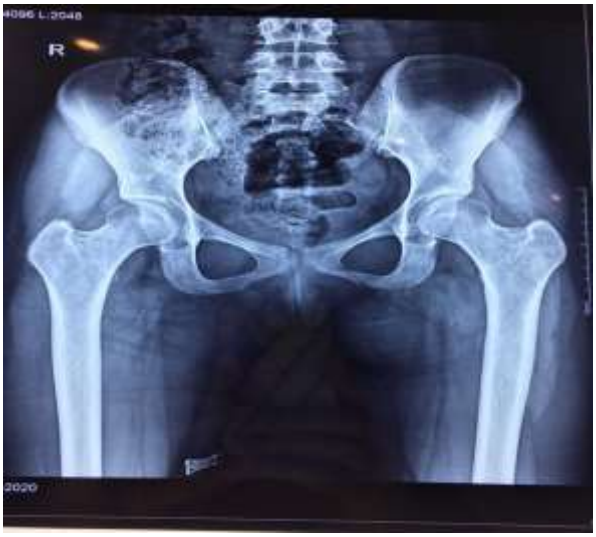

Fig.1.Radiography of patient showing any pathological finding

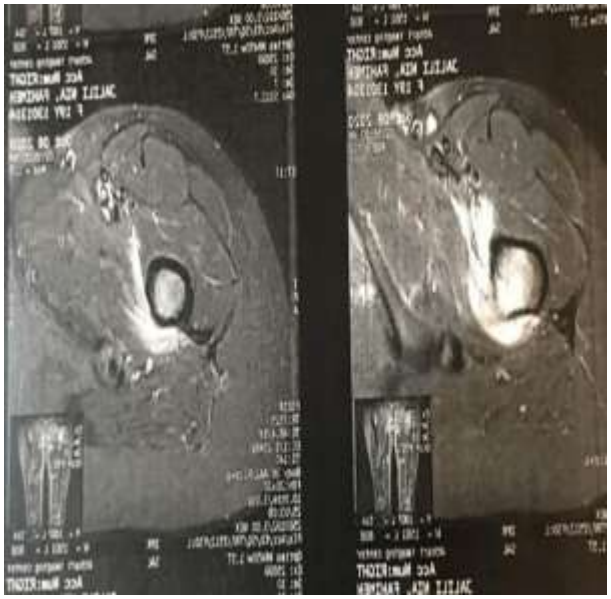

Fig.2. MRI showed a tumor arising from the right lesser trochanter with soft-tissue edema

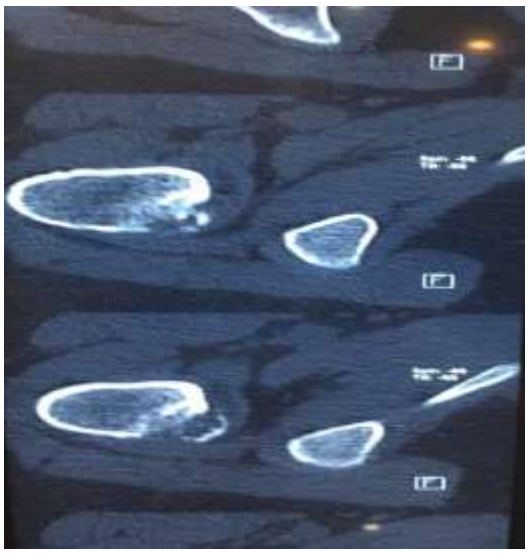

Fig.3. CT showed a destructive lesion in the lesser trochanter

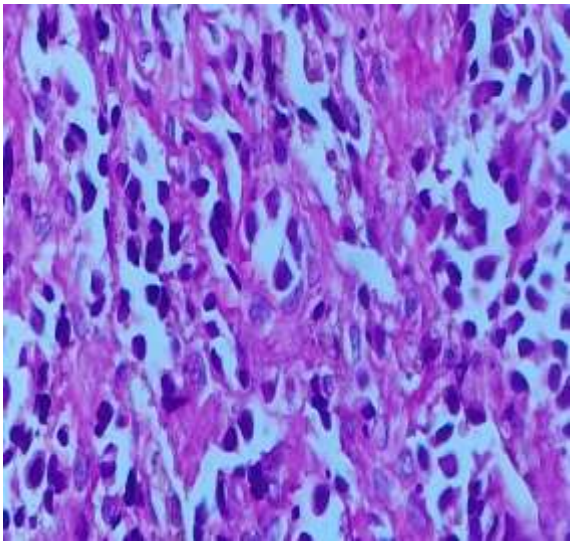

Fig.4. Histopathology image showing a large cell with large nuclei as well as conspicuous red nuclei and eosinophilic cytoplasm some cells with brown intracytoplasmic pigments

Fig.5. PET/CT report showed metastatic disease in:1. right soft tissue lesion of the maxillary sinus 2 multiple bilateral pulmonary masses and nodules 3. soft-tissue nodules in the right breast 4. two chest wall subcutaneous soft tissue lesions 5. bilateral adrenal mass 6. multiple skeletal lesions

Discussion

Melanoma is a common neoplasm of the skin. It may also arise from the mucosal surface or the site to which the neural crest has migrated.

It is not uncommon for melanoma to metastasize in the bone, though primary osseous melanoma is fairly rare. Melanoma rarely presents in the lesser trochanter. Malignant melanoma (the clear cell sarcoma) accounts for 1-3% of all malignant cases and its incidence is wising in the world (11).

Metastasis of melanoma in the bone marrow is rare and its dissemination is reported in only 5-7% of cases (12). In about 5-15% of patients initially presented with melanoma metastases, no primary tumor can be detected (13, 14).

However, in some instances, the primary tumor may be located in the skin and regress spontaneously (15). The prognosis of patients with metastatic malignant melanoma and metastases of unknown primary origin is known as that of a patient with overt primary melanoma (14).

Clear cell sarcoma is a malignant soft-tissue neoplasm first described by Einzinger in 1965 (18). It is fairly rare, accounting for less than 1% of soft-

tissue sarcoma. It rarely occurs in bones, but in 90-95% of cases, it is seen in the extremities. The feet and ankle are the most common primary sites, accounting for 33-43% of cases(19). Pain and tenderness are the main symptoms in 33-50% of patients (20).

The immunohistochemical study can help diagnose malignant melanoma.

Typically, melanoma reacts to Vimentin, S-100 protein, HMB 45, Melan-A, tyrosinase and microphthalmia transcription factor (16).

HMB-45 is a significantly more specific marker than S-100 protein (17). In Immunohistochemical studies, clear cell sarcoma is positive for S100. HMB-45 is also perceived in the granular cytoplasmic staining of the clear cell sarcoma.

One difficulty in the diagnosis of clear cell sarcoma is that its histologic characteristics are identical to the cutaneous melanoma. Osteogenic melanoma is a rare variant of melanoma described by Uramecher in 1984. Since then, three main theories have been proposed to explain this variant:

On presumes that osteoid formation is a response to a previous injury because in certain cases, lesions are observed in the site of previous resections. Contrary to this theory, various osteogenic melanomas are observed in the denovor lesion. Moreover, reactive osteogenic does not present cellular atypia or positivity for melanocyte markers (21). BREKKE (22) and Kubota reported a similar case with a different treatment plan.

Conclusion

In the present case, various radiographic analyses suggested that the tumor was interosseous in origin. Diagnostic analysis revealed no primary sites other than the lesser trochanter.

In addition, the patient was disease-free for 12 months after surgery, compared to the median survival of 3.6 to 8 months in a patient with skeletal metastases of malignant melanoma, including those with unknown primary tumors.

It is worth noting that the diagnosis of primary malignant melanoma of bone is often delayed due to nonspecific and radiologic factors.

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