

## OSTEOSARCOMA IN NEONATE WITH RARE LIVER METASTASIS AND ABDOMINAL LYMPHADENOPATHY

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### SUMMARY

In this study, we report an extremely rare case of osteosarcoma in neonate with rarely metastasis and abdominal lymphadenopathy. The patient appeared to have progressive swelling of the distal right femur. Magnetic Resonance Imaging (MRI) of the right thigh revealed an eccentric lesion arising from the right femur. Computed Tomography (CT) of the thorax and abdomen showed liver metastasis and abdominal lymphadenopathy without lung involvement. It was confirmed to be osteosarcoma with focal rhabdoid features on histopathological examination.

**Keywords:** neonate osteosarcoma, liver metastasis, lymphadenopathy

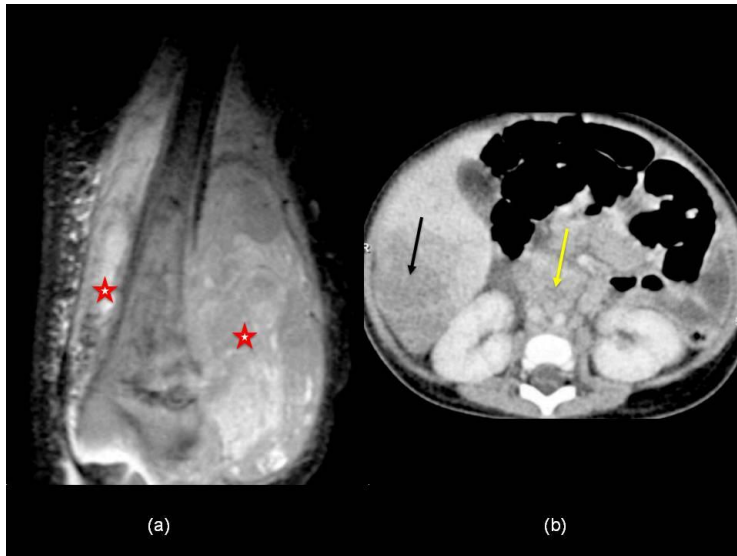
### 1.0 Introduction

Torus palatinus is a benign bony outgrowth arising from the hard palate which occurs in the midline. Different morphology of torus palatinus have been described namely flat shape, lobulated, (1,2) as well as unilocular or multilocular bony exostosis (3). It was more frequently seen in females than in males and in subjects aged 40 years and above (4). Patients normally present with incidental finding of bony outgrowth seen on the hard palate, prior to denture prosthesis assessment (2).

### 2.0 Case Report

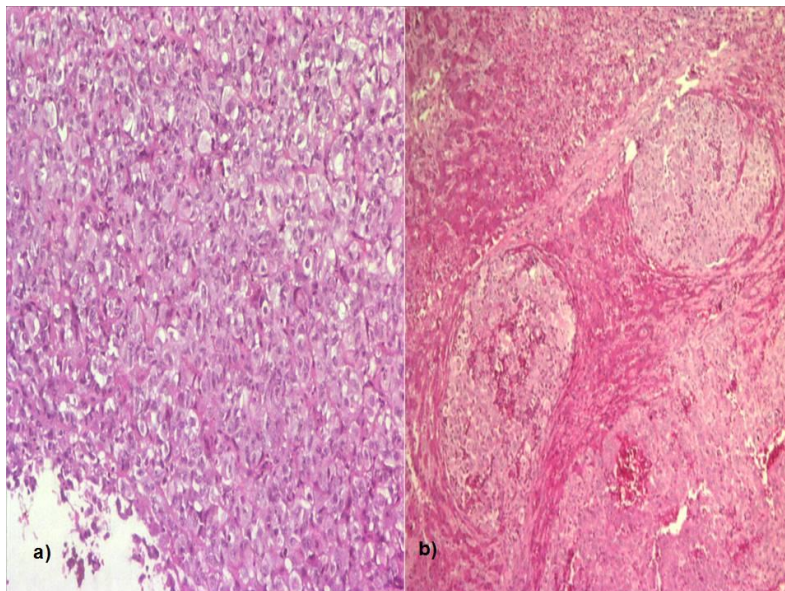
A 4-month-old girl presented with one-month history of swelling at the distal right femur which increasing in size associated with lost of weight and appetite since two month ago. MRI of the right thigh showed an eccentric lesion arising from distal metaphysis of the right femur and extends to the mid diaphysis and epiphysis (Figure 1a). This lesion has mixed to high signal intensity in T2 weighted images.

CT scan thorax and abdomen showed an ill defined hypodense area in segment VI of the liver with abdominal lymphadenopathy (Figure 1b). No lung lesion seen.



**Figure 1**

(a) MRI of the right thigh showing an eccentric lesion arising from distal metaphysis of the right femur and extends to the mid diaphysis and epiphysis. This lesion has mixed to high signal intensity (stars) in T1 weighted image post gadolinium coronal plane. (b) Contrast enhanced CT abdomen in axial plane showing an ill-defined hypodense area (black arrow) in segment VI of liver with multiple para-aortic and paracaval lymphadenopathy (yellow arrow).



**Figure 2**

a) Histology from bone biopsy showing tumour cells with high Nucleus: Cytoplasm ratio and round to spindle nucleus. Mitoses are seen easily. b) Histology from liver biopsy showing multiple nodules and entrapped ductules suggests that the liver lesion is a secondary tumour deposit. The immunohistochemistry results in keeping with osteosarcoma.

Needle biopsy was done at the distal femur. Histopathological examination showed tumour cells with high nucleus, mitoses and a distinct rhabdoid appearance. In other areas, there is osteoid production. Fragments of calcified and myxochondroid matrix, as well as necrosis noted. Features are those of an osteosarcoma with focal rhabdoid features (Figure 2a).

In Operation Theater, laparoscopy procedure was abandoned due to frank haemoperitoneum and preceded to laparotomy. Trucut and wedge biopsies were obtained from the lesion in liver which histopathological examination revealed high-grade sarcoma (Figure 2b).

Postoperatively, the patient's condition deteriorated. No further surgery was performed due to poor prognosis. Only one course chemotherapy was given. She died after one-month diagnosis osteosarcoma with liver metastasis and abdominal lymphadenopathy

### 3.0 Discussion

Osteosarcoma is characterised by the direct formation of immature bone or osteoid tissue by the tumour cells. The first peak is in the 10-14 year-old age group shows close relationship between the adolescent growth spurt and osteosarcoma (1). It is extremely rare below the age of 6 year especially in neonate.

The cause of osteosarcoma is unknown. However, irradiation and genetic influence have been implicated in its development. Like in this case, most likely is genetic influence even though the antenatal history was uneventful. It is apparent that two suppressor genes, p53 and Rb, have major roles in tumorigenesis in osteosarcoma. Approximately 3-4% of children with osteosarcoma carry constitutional germline mutation in p53. The majority of these cases with germline p53 mutations occur in patient with a strong family history of cancer or with family histories suggestive of the Li-Fraumeni syndrome (familial cancer syndrome). The strongest genetic predisposition to osteosarcoma is found in patients with hereditary retinoblastoma. In hereditary retinoblastoma, germline mutation of the Rb gene is common (2).

Osteogenic sarcoma usually metastasizes to a homogenous route frequently to the lungs (3). In both clinical and autopsy series, over 90% of patients with recurrent disease had pulmonary metastasis. Although extrapulmonary metastasis did occur, there were usually found in patients with preterminal diffuse disease or at the autopsy. They seldom occurred before the onset of pulmonary metastases (4). Bone is the second most common site of metastasis and usually become involved only after pulmonary metastases have occurred. But in this case, patient presented with liver metastasis alone without lung involvement that is seldom to occur.

Prior to biopsy, CT scan should evaluate the pulmonary metastases and bone metastases should be ruled out by bone scintigraphy. Giuliano *et al.*, (1984) reported that 59 of 111 patients (53%) developed metastatic osteosarcoma. However, only 36 (61%) of these 59 developed pulmonary metastases as their initial site of recurrence, whereas 23 (39%) developed both pulmonary and extrapulmonary disease alone. Among the extrapulmonary disease, only one case was involved the liver. Because bone scans or nonpulmonary studies were not done routinely, extrapulmonary metastases were found only after development of signs or symptoms of involvement.

Distant bone metastases represent the latest stage of disease and are associated with the poorest prognosis (5). Like in this case, where the patient presented with distant metastasis before the diagnosis of osteosarcoma was made.

Treatment, the 5-year survival rate people with localized osteosarcoma is in the range of 60% to 80% compared with 15% in early 1960s. If the osteosarcoma has already spread when it is first found, the 5-year survival rate is about 15 to 30%. Metastatic disease should be treated with a combination of chemotherapy and metastasectomy and repeated thoracotomies are indicated. The cure for uncommon cases in which metastatic disease at presentation is located is less than 5% (2). However, surgery was not done to this patient because of poor prognosis, only chemotherapy had been given to her. She expired after one month diagnosed to have osteosarcoma with liver metastasis and abdominal lymphadenopathy that is unusual to occur at this age group and rare to have an extrapulmonary metastasis prior to lung metastasis.

#### 4.0 Conclusion

Osteosarcoma of the infant is rare. We present a case of primary osteosarcoma in a 4-month-old girl infant with metastasis to liver and abdominal lymphadenopathy. Metastasis at presentation is uncommon, early diagnosis and aggressive surgical management will help to improve the patient's outcome.

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