Case Report

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Congenital sacral mesenchymal chondrosarcoma in a neonate: A case report and review of literature

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SUMMARY

Mesenchymal chondrosarcomas are rare malignant tumours in children, especially, in neonates. The authors present a case of congenital mesenchymal chondrosarcoma in a 1-day neonate located in sacrum. According to the authors' literature searches, this case is the first congenital sacral mesenchymal chondrosarcoma. We also reviewed the papers published in English literatures.

Key words: Mesenchymal chondrosarcoma, neonates, sacrum

INTRODUCTION

Mesenchymal chondrosarcom can arise anywhere in the body. Those arising from the soft-tissue are among the rarest tumours in childhood,[1] and fall under the term of soft-tissue sarcoma (STS).

STS comprise 11% of all childhood malignancies; only 2% of all childhood sarcomas are discovered in the new born period.[2,3] The number of different histological subtypes complicates attempts to define diagnostic and management criteria. Moreover, the effectiveness of treatment regimens including, surgery, chemotherapy, and radiotherapy is poorly defined.[2] These tumours carry a high malignant potential and tend to occur in young adults.[4,5] Although, most of the lesions are located in the skeleton, extra osseous mesenchymal chondrosarcomas arising from pancreas, kidney, Para pharyngeal space, thyroid, brain, and spleen have been reported.[4,6-13] Because of the rarity of these tumours, treatment approaches basically depend on information gained from single case reports or small case series. In this case report, we aimed to present the first case of congenital mesenchymal chondrosarcoma of the sacrum with a literature review.

CASE REPORT

A 1-day-old term male neonate presented with a left buttock mass. His Apgar scores were 10 at 1 min and 5 min following an uncomplicated vaginal delivery. The infant's weight was 2600 g. There was no pertinent family history.

Unfortunately, prenatal ultrasonography had been normal. After birth, a physical examination revealed a 5 cm × 6 cm mass located in the left buttock [Figure 1].

It was covered by mucus without skin, which could easily bleed. The physical examination of the neonate



Figure 1: Tumour before operation

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was otherwise normal. The reflexes were good and he had no feeding difficulties or respiratory distress. Examinations of other systems were unremarkable.

Computed Tomography [CT] scan was performed and showed a well-circumscribed mass 60 × 51 × 32 mm in diameter, localised in the gluteal region without a presacral element [Figure 2].

Ultrasonography evaluation of the abdomen was normal and laboratory findings were normal.

A sacrococcygeal teratoma was clinically suspected. Total excision of the mass that attached to the lateral aspect of posterior sacrum was carried out. In the operating scene, the tissue consistency was firm and sarcoma–like and it was completely excised [Figure 3] and sent for pathologic diagnosis.

Pathologic examination identified the tumour as a mesenchymal chondrosarcoma [Figure 4].

We prepared him for chemotherapy. Ten days post-operatively; he was noted to have many metastases on his back when he brought for suture removal [Figure 5].

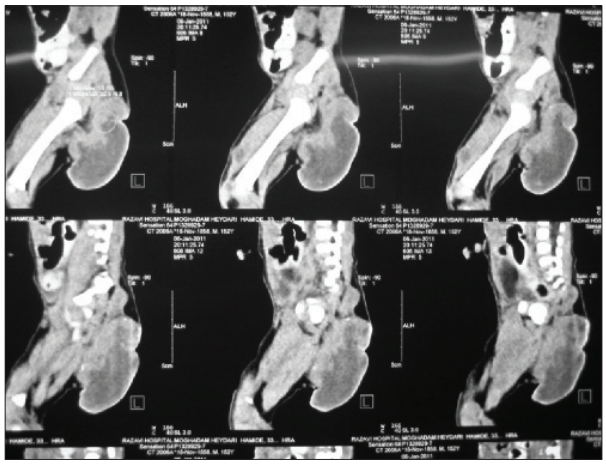


Figure 2: Tumour CT

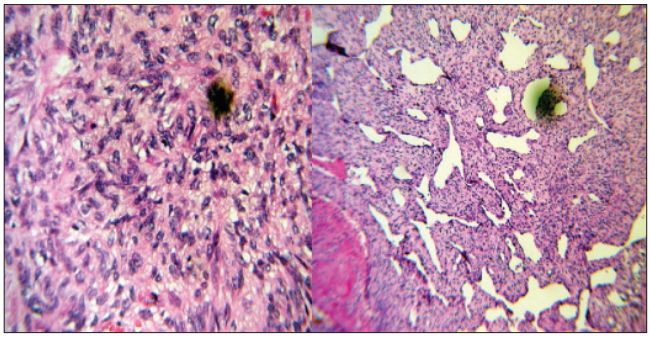


Figure 4: Pathology

We encouraged his parents to start chemotherapy as soon as possible, but unfortunately his parents decided not to perused chemotherapy. The infant died at 3 months old.

DISCUSSION

Mesenchymal chondrosarcoma diagnosed in the new born period is exceedingly rare, and for that reason, they are difficult to study and manage. The overall incidence of a neonatal solid tumour in one regional registry study was 5.7 tumours per 10^5 live births.[4] Comparisons between published studies are often difficult because of differences in age groups studied with some reports including, the first 3 months of life.[2] STS comprise a very small proportion of neonatal cancers within the 1st month of life. In a published report by Parkes *et a1*.,[4] they found only 6 malignant sarcomas in 170 neonates (3.5%) under 3 months of age in a 30-year study period.

A retrospective study of all STS diagnosed in the 1st month of life was conducted by 11 institutions of the



Figure 3: After operation

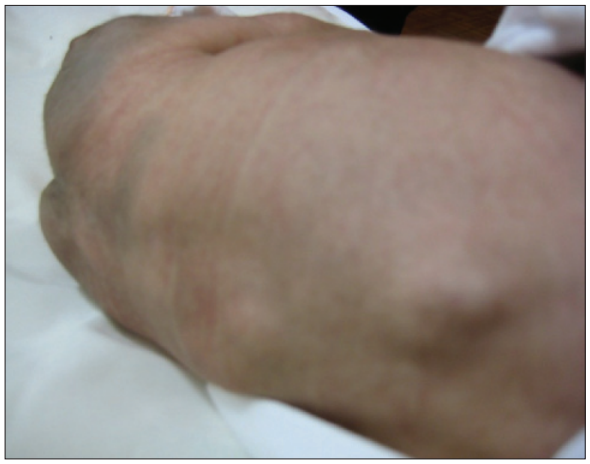


Figure 5: Back metastasis

Children's Cancer Group.[2] From 1971-1993, they found nine new born with non-rhabdomyosarcoma soft-tissue tumours and 32 neonates with STS. There were eight boys and one girl. Seven of their nine tumours (78%) were diagnosed at birth; most of their cases were in head and neck. In this report, 4 cases were mesenchymal sarcoma.[2]

Primary tumours of the sacrum include benign aggressive lesions such as aneurysmal bone cysts, osteoblastoma, giant cell tumours or low-grade malignancies such as chordoma and chondrosarcoma, which constitute 1-4.3% of all bone tumours.[10,14] Although, most of the classic chondrosarcomas are slow growing and low-grade tumours, mesenchymal chondrosarcomas are rapidly growing tumours with greater potential to metastasize.[15]

Mesenchymal chondrosarcomas can be encountered at any age. Their incidence tends to peak in the second and third decade.[1,4,5] The youngest age of a mesenchymal chondrosrcoma case presentation is 1 month, reported by Dantonello *et al*.[5] This was in the paravertebral region. The lowest age for a sacral mesenchymal chondrosarcoma is 10 years, reported by Küpeli *et al*.[14] Our case is the first case of congenital sacral mesenchymal chondrosarcoma. In a review by Kupeli,[14] most of cases of their study were female (25 of 42). Only 7 cases were reported in the spinal region and one case in the sacrum in a 10 years girl. Most reported cases are in head and neck.[14,16] Our patient is male with a tumour in the spinal region. Mesenchymal chondrosarcomas also can occur in other locations such as in the pancreas, kidney, Para pharyngeal space, thyroid, brain, and spleen,[4,6-13] as well as in bony structures such as maxilla, mandible, rib, vertebra, talus, ilium, tibia, femur, and skull bones.[17,5,7] Aprin *et al*.[18] described a 16-year-old female with a chondrosarcoma in the sacrum and S1 joint however, they did not specify the subtype of the chondrosarcoma. Although, there are 4 case reports in the literature reporting primary sacral mesenchymal chondrosarcomas in 10-17-, 20-, and 23-year-old patients,[19,20,14] the present case is unique for its sacral location and congenital presentation.

Radiological methods are necessary to delineate the extent of tumour and to provide a differential diagnosis. Magnetic resonance imaging[MRI] and CT scan are the best choices for mesenchymal chondrosarcoma diagnosis.[21-23]

Although, the pathogenesis of mesenchymal chondrosarcoma is not clear, the transition areas between mesenchymal cells and cartilage indicate precartilage mesenchymal origin.[8] Pathologically, it is characterised by undifferentiated small blue round cells mixed with cartilage.[24] Histopathological differential diagnosis of mesenchymal chondrosarcoma should include small round cell tumours such as lymphoma, neuroblastoma, rhabdomyosarcoma, and others such as osteochondroma, conventional chondrosarcoma, and hemangiopericytoma.[25] Immunohistochemistry and biomarkers such as cell differentiation[CD]99, sinaptophysin, leukocyte common antigen, desmin, Tdt, CD43, chromogranin, periodic acid–Schiff, and S100 may help to differentiate some of these tumours.[26] Complete surgical resection is the gold treatment for all subtypes of non-metastatic chondrosarcoma.[1,2,5] In a large series that also included adult patients, resection with wide margins was accepted as the gold standard of treatment.[27] In all paediatric mesenchymal chondrosarcoma cases, at least subtotal resection, but mostly complete resection, should have been undertaken[2,5,8,11,25,28-30] On the other hand, wide excision may cause significant morbidity depending on location and degree of invasion of the tumour. Bladder and bowel functional sequel (permanent urinary and anal incontinence) have been reported due to sacral resection in an adult with sacral mesenchymal chondrosarcoma.[15,19] There is no general agreement on whether additional chemotherapy, radiotherapy, or both are necessary after surgery[2,5] In our case, initially the tumour was removed completely. Due to metastatic spread in a short time after the surgery, we recommended chemotherapy but his parents did not wish to pursue this course of treatment.

Adjuvant therapy can be considered in patients with incomplete resection. Although, chondrogenic tumours are assumed to be relatively radio resistant due to lower growing rates, mesenchymal chondrosarcoma, with a higher fraction of dividing cells, can benefit from radiotherapy. Radiotherapy is especially, a valuable tool to ensure local control with macroscopic or microscopic residual disease after surgery.[1] Despite a scarcity of cases, chemotherapy seems to be efficient in mesenchymal chondrosarcoma. Gelderblom *et al*.[1] reported a higher chemo-sensitivity if the tumour contains a high percentage of round cells. This is also reported by Vencio *et al*.[31] Several authors have reported cases of mesenchymal chondrosarcoma responding to chemotherapy in paediatric patients.[5,8,11,28]

In a large series of chondrosarchomas, clear surgical margins and tumour grade were found to be statistically significant factors for local recurrence and systemic spread.[17] Ten-year survival rates for patients with mesenchymal chondrosarcoma are reported between 27% and 58% in the literature.[5,27,30,31]

Although, malignant tumours very rare in neonatal period, especially, sacral mesenchymal chondrosarcoma, paediatric surgeons may find it like as a sacrococcygeal teratoma presentation in a neonate as we find.

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