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UNUSUAL PRESENTATION OF OSTEOGENIC SARCOMA IN A CHILD- A CASE REPORT

Pathology

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ABSTRACT

Osteogenic sarcoma is the eighth most common childhood malignancy, comprising 2.4% of all malignancies in pediatric age group, and approximately 20% of all primary bone malignancies. It commonly occurs in the long bones of the extremities near metaphyseal growth plates. We present this case of diaphyseal osteogenic sarcoma of right radius in a child, because of its unusual presentation and diagnostic problem faced clinically and radiologically. The tumor was diagnosed on the basis of fine needle aspiration cytology (FNAC) and confirmed by histopathology. Long term follow up of Limb salvage surgery and neo adjuvant chemotherapy showed excellent results in this patient with unusual presentation.

KEYWORDS

Osteogenic Sarcoma; Radius; Fine needle aspiration cytology (FNAC); Biopsy.

INTRODUCTION

Osteogenic sarcoma is amongst common malignancies affecting young adults. The most common sites are the femur (42%, with 75% of tumors in the distal femur), the tibia (19%, with 80% of tumors in the proximal tibia), and the humerus (10%, with 90% of tumors in the proximal humerus). Other likely locations are the skull or jaw (8%) and the pelvis (8%) [1]. Rachel et al in their study of epidemiology of bone cancer in 0-39 years old in Northern England, found incidence rates of 2.5 per million per year in age group of 0-14for Osteogenic sarcoma[2] . Osteogenic sarcoma in diaphysis of radius is a very rare entity. Kenan Abdelwahab IF et.al. reported one case of parosteal Osteogenic sarcoma involving diaphysis of radius[3] .

CASE REPORT

A nine year old male child, after being hit by a cricket ball, developed pain and swelling in right forearm. The attending doctor at the primary health centre made a provisional daignosis of green stick fracture of shaft of radius and the child was put in an above elbow Plaster of Paris [POP] Cast for 6 weeks. After the POP was removed, the patient did not have any relief of pain & swelling. At that stage, the treating doctor suspected delayed union and referred the patient to our hospital.

On clinical examination at that stage, a localized swelling (5x5 cm.) at junction of middle one third & lower one third of right fore-arm was noticed. There were no signs of acute inflammation and the skin over the swelling was normal. There was significant tenderness in the affected area. There was no abnormal mobility at the site of swelling. There was no associated lymphadenopathy.

LABORATORY FINDINGS: Routine Investigations were normal. The ESR was slightly raised (26mm/1STHour by Westergren Method). The Kidney Function Tests and Liver Function Tests were normal except for the Serum Alkaline phosphatase which was raised (230 U/L) (normal range of 50 – 136 U/L). CRP was negative. The Montoux Test, after 48 hours of tuberculin injection (5 T.U.) was negative (Indurations less than 5 mm).

RADIOGRAPHY: X-rays showed an expansile lesion involving diaphyseal region of radius with presence of significant periosteal reaction. A provisional diagnosis of bone tumor was made and possibility of Ewing' sarcoma, Tuberculosis, Sub- acute osteomylitis etc. were suggested at this stage. The child was subjected to MRI scan of right forearm. The MRI showed an expansile lesion with a breach of cortex on the volar surface of radius near lower1/3 region of the diaphysis of radius. There was lytic destruction of lower shaft of radius with altered marrow signals and large soft tissue component and periostitis [Picture 1]. A likely diagnosis of Ewing's Sarcoma and remote possibility of tuberculosis was suggested by radiologist. Further investigations were advised to reach for final diagnosis.

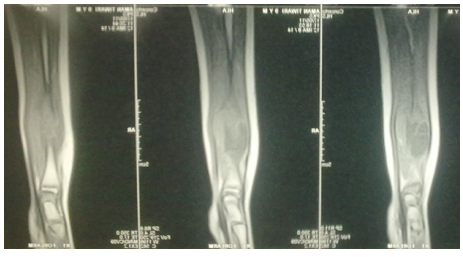

Fig. 1: MRI picture showing tumor in diaphysis of radius with extra osseous compartmental spread.

CYTOLOGIC FINDINGS: In view of cortical breach and extra compartmental spread of lesion, fine needle aspiration of the lesion was done using a 22-gauge needle. Smears were stained by Giemsa stain. Smears were richly cellular showing malignant tumor cells with eccentric nuclei and moderate amount of pale blue cytoplasm. There was moderate degree of nuclear pleomorphism. In between cluster of tumor cells, eosinophilic osteoid like material was seen. At places tumor giant cells and osteoclast type of giant cells were also seen. Hence a diagnosis of malignant mesenchymal tumor with possibility of osteogenic sarcoma was made. [Picture 2a]

HISTOLOGICAL FINDINGS: Biopsy was carried out under general anesthesia. Extra osseous spread of tumor along with breach of cortex was noted while doing surgery. The tumor tissue was fragile and bled significantly. Multiple dark brown soft tissues pieces were obtained. These were fixed in 10% formalin and sent for histo-pathological examination. Sections were stained with Haematoxylin & Eosin (H&E) Stain. Sections showed blood filled spaces lined by thin endothelial cells. These vascular spaces were separated by thick connective tissue septa showing presence of a large number of malignant tumor cells with moderate degree of nuclear pleo-morphism. The tumor cells were having eccentric nuclei with moderate amount of eosinophilic cytoplasm. At places, in between tumor cells, there was presence of eosinophilic osteoid material. Few osteoclast type giant cells were also seen. The findings were consistent with Telangiectatic osteogenic sarcoma [Picture 2b].

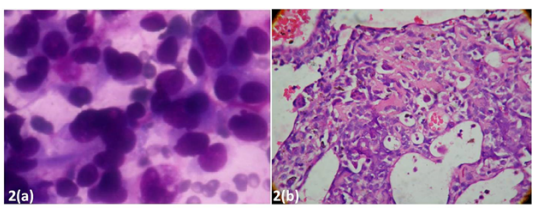

Fig. 2a: Cytological smear showing malignant tumor cells having eccentric nuclei with moderate degree of nuclear pleomorphism and eosinophilic osteoid like material (Giemsa stain, 40x10) Fig. 2b: Biopsy sections shows blood filled spaces lined by thin endothelial

cells separated by thick connective tissue septa, showing presence of a large number of malignant tumor cells with moderate degree of nuclear pleomorphism with presence of eosinophilic osteoid material. (H&E stain 10 x40)

FOLLOW UP: After the diagnosis of Telangiectatic osteogenic sarcoma was established, a CT scan of chest was also carried out. Contiguous axial slices were taken from lung apices to lung bases after intravenous administration of iodinated non-ionic contrast agent. CT scan of the chest was normal. The bone scan carried out at this stage demonstrated lesion confined only to forearm. The child was put on neo-adjuvent Chemotherapy. In view of extra compartmental spread without distant metastasis, the parents of the patient were given detailed information on treatment options and likely outcome. Detailed information about limb salvage procedure in the form of wide excision of the radius leaving behind only upper 1/3rd of radius as well as details about below elbow amputation were provided to the patient's parents. They opted for limb salvage surgery. En-block excision of the tumor was carried out, sacrificing radius from wrist joint to upper 1/3 and middle 1/3 junction (more than 5 cm. away from tumor margins). The wrist joint was stabilized using a long plate across wrist joint, fixing ulna to 3rd metacarpal. The patient was kept on regular follow up and is disease free at six years post limb salvage procedure. The bone scan at six years follow up is normal. The patient had developed manus varus deformity at wrist. Centralization of hand and correction of deformity is planned later at skeletal maturity.

DISCUSSION:

The Osteogenic sarcoma is rare in first decade of life. Christensen in his series of one thousand bone tumors listed 29 cases of bone sarcoma below the age of ten years [4] . Hayles, Dahlin and Coventry while reviewing their experience of this tumor over a 50-year period at the Mayo Clinic, found 129 instances of osteogenic sarcoma in patients below the age of 16 years. There were only 30 patients of age 10 years or less [5]. The involvement of radius by this tumor is extremely rare, irrespective of age. Our patient, a nine year old boy, got medical attention because of injury on forearm. The doctor at the primary health centre treated him for green-stick fracture of shaft of radius. The clinical, laboratory or radiological studies did not point towards the diagnosis of tumor or osteomylitis at that stage. The possibility of delayed union was considered by treating physician as the patient did not have relief after adequate period of immobilization in the plaster. So the patient was referred to our centre.

The radiological examination done at six weeks after injury at our centre and the subsequent MRI carried out pointed to the diagnosis of malignant tumor suggestive of Ewing's sarcoma. The final diagnosis was established by FNAC coupled with histopathology. Thus despite the patient seeking immediate medical attention for the trauma, the diagnosis was not suspected initially because of misleading clinical picture. M.Smida et.al. also reported a rare case of juxtacortical Osteogenic sarcoma arising from superior metaphyseal area of radius in a child of 12 years. They also emphasized that diagnosis and treatment was delayed because of misleading and wrongly reassuring radiological and histological aspects [6] .

The diaphyseal involvement in a long tubular bone by osteogenic sarcoma is extremely rare. Harvard Ellman et al. reported a rare case of diaphyseal osteosarcoma of femur in a 13 year old girl [7]. Kenan Abdelwahab IF et.al. reported a case report of Osteogenic sarcoma in diaphysis of radius [3] . In our patient also, the tumor was confined to diaphyseal area of radius with cortical breach on volar surface of radius, letting the tumor spread outside osseous compartment.

The FNAC established the Diagnosis of malignant mesenchymal tumor with possibility of Osteogenic Sarcoma. Abderwahab have also reported a case of de-differenciated osteosarcoma of radius in a 35-year-old woman. That case was reported because of rarity of involvement of radius and presence of two distinct components on Pathological examination: a cartilaginous low- to moderate-grade osteosarcoma on the surface and a high-grade Osteogenic sarcoma in the intramedullary component [8]. In our patient, the Diagnosis of Osteogenic Sarcoma (Telangiectatic Variant) was confirmed by Open Biopsy. The telangiectatic variant of osteosarcoma is rare. Its biological behaviour, treatment and prognosis are controversial [9] .

The diaphyseal Osteogenic sarcoma is very aggressive and patient usually has pulmonary metastasis at the time of presentation [9]. In our case, there was no distant metastasis, probably as the patient had reported early because of accidently sustaining injury around that region. However, despite being so early, the tumor had already crossed the osseous compartment with cortical breach and had spread to the surrounding soft tissue. This was seen preoperatively on MRI Scan and confirmed while performing open biopsy. The patient, after neo adjuvant chemotherapy, was managed by limb salvage procedures.

CONCLUSION:

Diaphyseal osteosarcoma of radius is extremely rare. The early detection of tumor is usually accidental and requires high degree of suspicion by treating doctor. Irrespective of the age and location, diagnosis is based on the cytology and histopathology as sometimes radiological and imaging studies can be misleading. A successful outcome with long life expectancy, good functional limb and high patient satisfaction is possible with multidisciplinary approach provided timely diagnosis is made.

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