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Case Study

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A RARE CASE OF IDIOPATHIC MASSIVE SWELLING IN THIGH OF A CHILD: A CASE REPORT

Dr. Nidhi Sharma[1]* and Dr. Anil Kumar Sharma[2]

[1]A.M.O., M. S. Shalyatantra, Ayush Department, H. P.

[2]Professor, MD Ayurveda, Department of Shalyatantra, R.G.G.P.G. Ayurvedic College and Hospital Paprola H.P.

***Corresponding Author: Dr. Nidhi Sharma**

A.M.O., M. S. Shalyatantra, Ayush Department, H. P.

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ABSTRACT

A 13 year old male child came at R.G.G.P.G. Ayurvedic college and Hospital Paprola with pain and idiopathic swelling over thigh region with no history of any trauma which was noted incidentally by him 4 days back. There was no history of any kind of trauma and fall. On clinical examination there was massive swelling on left thigh and the local temperature was raised. Patient also had fever 20 days back. After radiological evaluation, in which periosteal reaction was seen and CT left thigh, which was suggestive of osteomyelitis mid shaft femur the provisional diagnosis was made as osteomyelitis left femur. But after further workup and higher investigations the diagnosis was changed and the final diagnosis was concluded as Ewing's sarcoma. Ewing's sarcoma is an aggressive sarcoma of bone and soft tissue. It is considered a prime example of paradigms of translocation positive sarcoma genetically rather simple disease with a specific and neomorphic potential therapeutic target.[1] The treatment of Ewing's sarcoma relies on multidisciplinary approach, coupling intensive neoadjuvant and adjuvant chemotherapies with surgery or radiotherapy for control of primary site and possible metastatic disease. Current 5-year overall survival for patients with localized disease is 65-75%. Patients with metastasis have a 5-year overall survival <30%, except for those with isolated pulmonary metastasis approximately 50%.[2] So, before concluding the final diagnosis, higher clinical investigations should be done in case of suspicion, to prevent the diagnostic errors which may result in devastating consequences.

KEYWORDS: Idiopathic swelling, swelling Thigh, Ewing's sarcoma, osteomyelitis.

INTRODUCTION

Any case of a pediatric patient having idiopathic swelling over the thigh usually heads the diagnosis of most of physicians towards osteomyelitis but this is very important to differentiate it with other pathologies like Ewing's sarcoma, Osteoid osteoma and Osteosarcoma as the misdiagnosis may lead to a serious complicated pathological condition and injustice to the patient. Osteomyelitis is one of the most difficult and challenging problems encountered in orthopedic. The ravaging effects of osteomyelitis on the bone and its neighboring joints has been always a matter of concern for orthopedic surgeons.[3] It is a serious infection of bone that can either acute or chronic.[4] On the other hand, Ewing's sarcoma is the highly malignant tumor occurring between the age of 10-20 years affecting long bones mainly Femur and Tibia in which diaphysis being the most common site. Likewise, osteosarcoma is the second most common and highly malignant primary bone tumor of mesenchymal cells characterized by formation of bone by the tumor cells occurs between 15-25 years of age having lower end of femur its most common site. These conditions usually presented with the same complaints i.e. pain and swelling over thigh region often associated with fever and become noticeable due to swelling. Cortical destruction, new bone formation and the periosteal reaction being the common radiological feature among three.[5] All these life threatening pathological condition have poor prognosis which should not be ignored during making the diagnosis so higher investigations other than x-ray should be done to conclude the diagnosis.

MATERIALS AND METHODS

Aim and objective

1. The main aim of this study to clinically differentiate between osteomyelitis, Ewing's sarcoma, osteosarcoma.
2. To conclude the final diagnosis after higher investigations.
3. To prevent the injustice with the patient due to misdiagnosis and negligence.

CASE REPORT

A 13-Year-old child presented with the complaint of pain and swelling over left thigh which was noted incidentally

by him 4 days back. There was no history of any kind of trauma or fall. On clinical examination, local skin was normal and had no scar related to previous trauma or injury. Massive swelling was present on left thigh. The tenderness was diffuse over left thigh and the local temperature was raised. Patient also had fever 20 days back. (Fig.1-2)

On radiological evaluation x-ray showed irregularity in cortical region of mid shaft left femur along with bowing deformity and periosteal reaction. On further workup**, CT left thigh** showed diffuse periosteal reaction along the mid shaft of the left femur with adjoining subperiosteal/intramuscular fluid collection with effacement of intramuscular planes. No cortical breakage was noted and suggestive of osteomyelitis of midshaft femur with adjoining subperiosteal / intramuscular abscess. (Fig.3-5)

MRI lt. thigh showed multiple intramedullary altered signal lesions in diaphysis of left femur with cortical thickening upto 8mm and concentric uplifted periosteal layers with breaks within with extensive similar characteristic soft tissue surrounding entire diaphysis resulting in soft tissue extension s/o sarcomatous etiology ? ewing's sarcoma, ??osteosarcoma.

For further investigations and management patient referred to higher institute and there biopsy was taken and the **Oncopathological/ Histopathological** report showed positive IHC markers i.e. NKX2.2 – immunoreactive in 76-100% tumour cells (score4+), CD99 –immunoreactive in 76-100% tumour cells (score4+), Vimentin- immunoreactive in 76-100% tumour cells(score4+), Ki-67 Immunoreactive in 35-40%tumour cells (score 2+), ERG - non immunoreactive in tumour cells (score 0) suggestive of Ewing sarcoma.

Whole body FDG- PET Scan was done to rule out any metastasis which showed b/l cervical level II lymphnodes (Left) noted inflammatory. Muculoskeletal - Ill defined heterogenous soft tissue mass with cystic internal areas seen encasing the proximal epimeta and diaphyseal regions of left femoral shaft with ill defined fat planes and infilterating the adjacent muscles of the left thigh with cortical erosions and associated periosteal reaction. No extension into knee joint seen. No abnormal hypermetabolism elsewhere in the body and organs under view in the present study.

CECT Scan of Thorax study reveals no significant abnormality.
Final diagnosis was made as Ewing's sarcoma and treatment has been given to the patient accordingly.

DISCUSSION AND RESULT

In current medical practises, idiopathic swellings like these are frequently misdiagnosed, while malignancies like Ewing's sarcoma tend to be easily overlooked. In this instance, the patient was initially diagnosed with osteomyelitis, but after additional testing and investigations, the final diagnosis was determined to be Ewing's sarcoma.

Ewing's Sarcoma, a highly malignant tumour that can occasionally last up to 30 years, frequently affects long bones (in two thirds of cases), primarily the femur and tibia.[3] The long bone's diaphysis is the most typical location. The entire medullary cavity, or a sizable portion of it, is affected by the tumour. The tumour tissue is soft and grey-white, and may be thin, nearly like pus. The periosteum may be raised, the bone may enlarge, and sub-periosteal new bone is frequently formed in layers. Early on, the tumour ruptures through the cortex and spread into soft tissues. It is made up of sheets of small, quite uniform cells that resembles lymphocytes. The cancer cells frequently surround a central clear area, forming a ***pseudo-rosette***. It quickly multiplies and spreads through the blood to the lungs and other bones.[5]

Clinical characteristics

Both pain and inflammation are evident in the patient. Prior to onset, there could have been trauma, but this is typically incidental. It is often accompanied by fever, in which case osteomyelitis can be mistaken for it. Usually, diaphysis swelling is present.[5]

Radiological characteristics

In a typical example, a long bone's midshaft possesses a lytic lesion with cortical destruction and new bone growth in layers that resemble an onion peel. The tumour may be in the metaphysis in atypical presentations, where it may be mistaken for osteomyelitis. It could mimic a soft tissue sarcoma and predominately involve soft tissues with minimal cortical damage. With barely any new bone production, it is largely a lytic lesion in flat bones.[5]

The tumour is extremely radiosensitive. Chemotherapy and radiation are two treatments options. The prognosis is quite bad. Secondaries from bone to bone are usual. Five-year survival, which was formerly barely 10%, has recently increased to 30–40% because to powerful chemotherapy treatments.

Using the information in Table 1, it can be distinguished from other bone tumours. It can be distinguished from chronic osteomyelitis by the following characteristics: sequestrum, clearly defined cloacae and periosteal reaction and metaphyseal site.

Table 1: Differential features of common bone tumours.[5]

TUMOUR	AGE (yrs)	COMMON SITES	LOCATION	CLINICAL FEATURES	RADIOLOGICAL FEATURES	D/D	PATHOLOGY
Osteosarcoma	15-25	Lower end of femur, upper end tibia	Metaphysis	Pain ++ Swelling++ Duration-weeks to months	Sunray appearance, Codman's triangle, Tumour new bone +	Ewing's tumour	Tumour cells with osteoid or bone formation
Ewing's tumour	5-15	Femur ,tibia Flat bones*, Multicentric*	Diaphysis	Pain++ Swelling+ Fever + Duration weeks-months	Onion- peel appearance	Osteosarcoma, osteomyelitis	Sheaths of round cells
Osteoclastoma	20-40	Lower femur Upper tibia Lower radius	Epiphysis region	Pain+ Swelling + Duration mths	Soap bubble appearance No tumour new bone	Aneurysmal bone cyst, fibrous dysplasia	Multinucleate giant cells in fibrous stroma

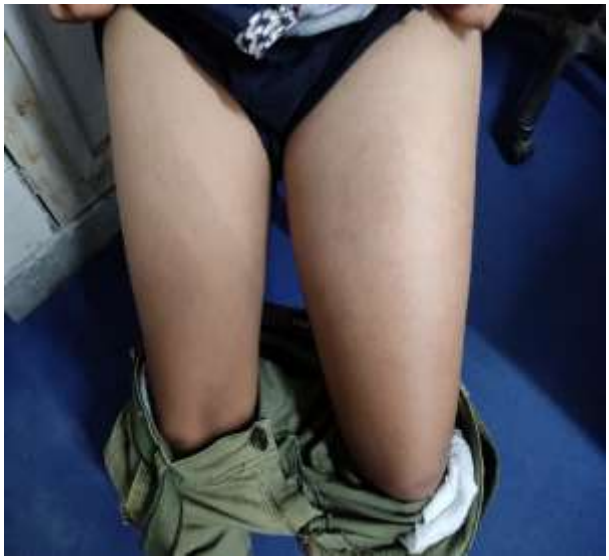

Fig. 1

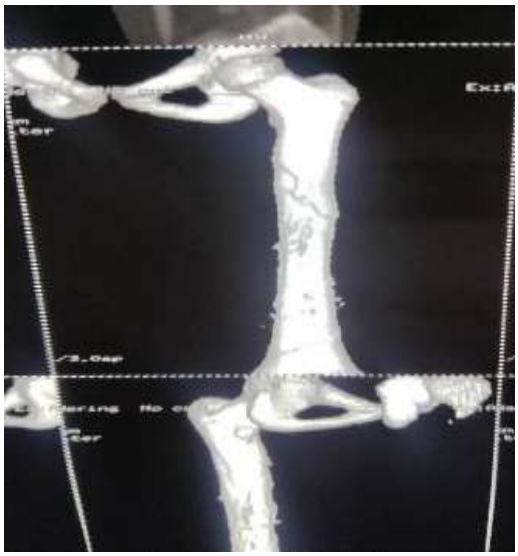

Fig. 2

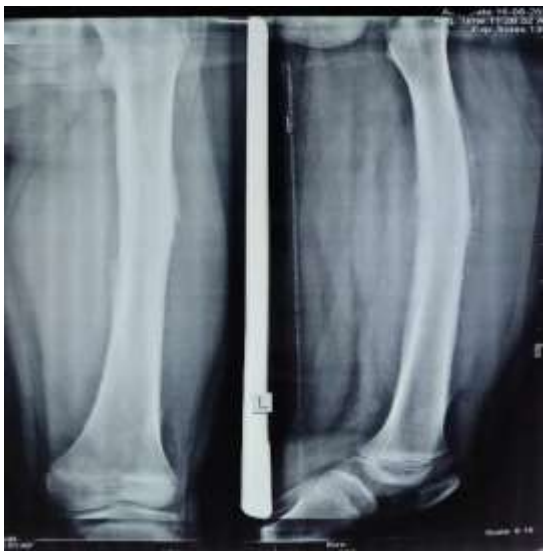

Fig.3

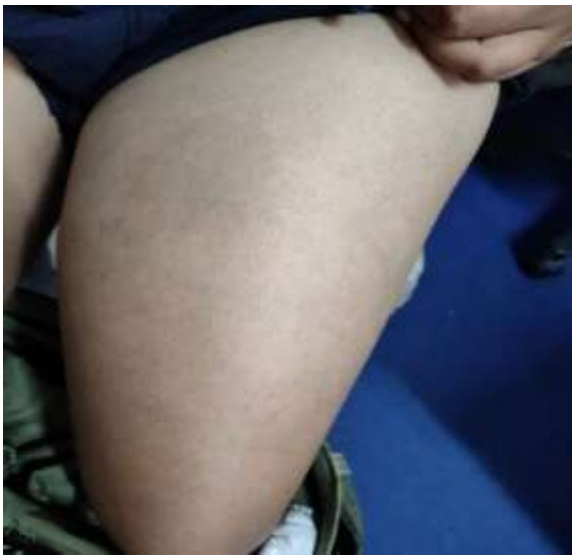

Fig.4

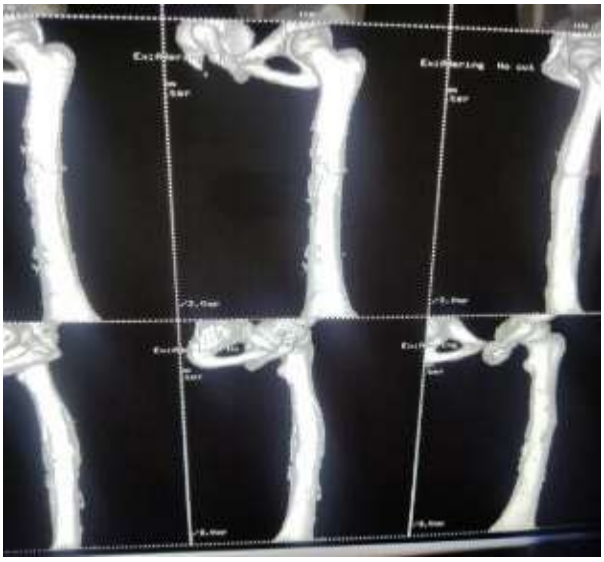

Fig. 5

Fig. 1 & 2: Shows Swelling in The left thigh of patient. Fig 3 shows xray finding of irregularity in cortical region of mid shaft left femur along with bowing deformity and periosteal reaction. Fig.4 & 5 shows CT finding of periosteal reaction along the mid shaft of the left femur with adjoining subperiosteal fluid collection.

CONCLUSION

The conclusion of the study is that any pediatric patient with idiopathic swelling in thigh region can be the case of Osteomyelitis, Osteosarcoma or Ewing's sarcoma. It should be differentiated with other bone tumours like osteoclastoma, chondrosarcoma as described earlier. Proper diagnosis is very important in such cases. Tumors like Ewing's sarcoma get easily ignored by the physicians if not examined properly. Correct diagnosis should be made with the help of higher investigations like CT and histopathological examination. In case of doubt one should not conclude the diagnosis without going through the higher investigations so that appropriate and effective treatment can be provided to the patient on time. Patient who came to us with this kind of swelling was diagnosed as osteomyelitis earlier on the basis of clinical examination, radiological examination and CT scan. Later on, after further investigations like MRI and histopathological examination eventually the diagnosis heads up towards Ewing's Sarcoma and the final diagnosis was made and treatment was started accordingly.

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