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Research Article

EOSINOPHILIC GRANULOMA OF FEMORAL SHAFT MASQUERADING AS EWING SARCOMA: A CASE REPORT AND REVIEW OF THE LITERATURE

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A B S T R A C T

Introduction: Eosinophilic granuloma (EG) of bone is a rare tumor comprising less than 1% of all bone tumors. The nonspecific clinical and radiological pictures often make the diagnosis difficult. We report a case of EG of Femoral diaphysis that masquerades as Ewing sarcoma.

Case presentation: A 7-year-old child presented with right thigh pain and swelling for 2 months. Radiograph of thigh showed a lytic mid-diaphyseal lesion with endosteal cortical erosion and diffuse circumscribed laminated periosetal reaction with neobone formation (onion skin appearance). Magnetic resonance images revealed hypointense T1W and hyperintense T2W medullary lesion with circumscribed periosteal reaction and neobone formation. Considering the clinical and radiologic features, the provisional diagnosis of Ewing sarcoma was made. But biopsy of the lesion proved it to be an Eosinophilic granuloma of bone. The child was also investigated for other sites to evaluate associated lesions. There was no evidence of any other organ involvement. The solitary bony Eosinophilic granuloma was managed conservatively with regular evaluation of the lesion. At 15-months follow up, the lesion has completely healed and the child is asymptomatic.

Conclusion: Isolated EG of bone in children often arises a radiological confusion with subacute osteomyelitis and Ewing sarcoma. Conclusive diagnosis is established after histological examination of the lesion. The lesion may show spontaneous resolution, and hence a regular clinical and radiological follow up is needed to look for the behaviour of the lesion.

INTRODUCTION

Eosinophilic granuloma (EG) of bone is a rare tumor comprising less than 1% of all bone tumors. It was first identified by Lichtenstein and Jaffe (1940) as a solitary benign bone lesion [1]. Subsequently many researchers reported multiple lesions with similar histological features [2]. In 1953, Lichtenstein used the term histiocytosis-X to describe 3 types of reticulosis i.e; Eosinophilic granuloma for unifocal bone lesion, Hand-Schuller Christian disease for chronic systemic involvement and Lettere-siwe's disease for acute systemic involvement [2,3].

EG is usually seen in children below 10 years of age with male predominance. The skull, pelvis and diaphysis of long bone (15%) are common sites of involvement [3-8]. Because of its nonspecific clinical and radiologic features, the diagnosis of long bone EG always poses a challenge. In such circumstances biopsy has immense role in confirmation of the diagnosis [4-8]. There is no definite consensus on its treatment as well. Multiple modalities of treatment have been described including observation, local intralesional steroid injection, curettage and bone grafting, cryosurgery, radiotherapy and chemotherapy [3-10]. In such a scenario it is difficult to decide the best treatment for the patient.

We report a case of solitary EG of femoral diaphysis in a 7 year old child that was simulating Ewing sarcoma. However in due course of time the lesion healed completely without any intervention. The parents were informed that the data concerning the child will be used for publication, and they consented.

Case presentation

A 7-year old male child presented to our outpatient department with history of pain and swelling of right thigh for last 2 months. The pain was dull aching type, intermittent in nature, increasing with activity and was relieved to some extent with analgesic. The swelling progressed slowly over the last 2 months. He had no history of trauma, fever, weight loss or loss of appetite. On examination, there was mild tenderness on deep palpation of the bone. There was no rise in temperature or redness. Radiograph of the femur showed a mid-diaphyseal lytic lesion with endosteal cortical erosion. Surrounding the lesion, there was diffuse circumscribed laminated periosetal reaction with neobone formation (onion skin appearance) [Fig. 1]. These clinical and radiographic features raised the possibility of subacute osteomyelitis or

Ewing sarcoma. Haematological investigations except erythrocyte sedimentation (ESR 40 mm/hour) were within normal range.

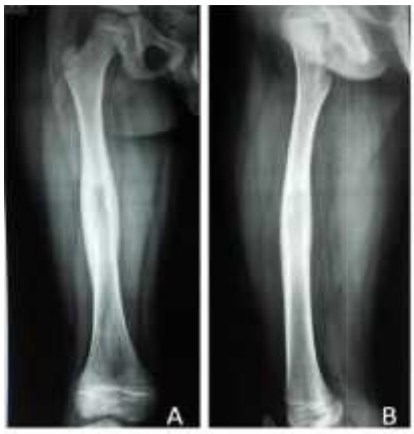

Figure 1 Antero-posterior (A) and lateral (B) radiograph of femur showing a mid-diaphyseal lesion with endosteal scalloping and widened medullary cavity with surrounding laminated periosteal reaction (onion peel appearance)

Magnetic resonance images showed hyperintense T2W and low to intermediate T1W signal. The lesion was surrounded by neobone with periosetal elevation. A small rim of soft tissue involvement was also noted [Fig. 2]. These findings were consistent with Ewing sarcoma. But biopsy of the lesion was not supportive of malignancy. There were numerous histicytic cells with bland looking ovoid nuclei having grooved or clefted nuclei. Many multinucleated giant cells were seen in a background of clusters of eosinophils and lymphocytes [Fig. 3]. These histological features were consistent with Eosinophilic granuloma (Langerhans cells histiocytosis) of bone. The child was further investigated with chest x-ray, skull x-ray and high resolution computed tomographic scan of chest. But all these investigations were normal, and hence the final diagnosis was a solitary Eosinophilic granuloma of bone.

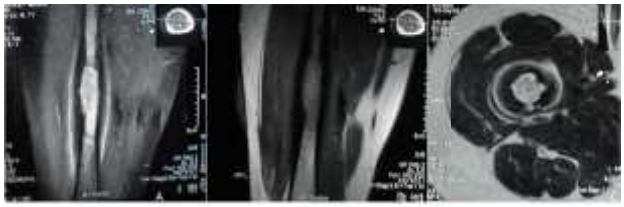

Figure 2 Magnetic resonance image showing T2W hyperintense andT1W hypointense lesion.

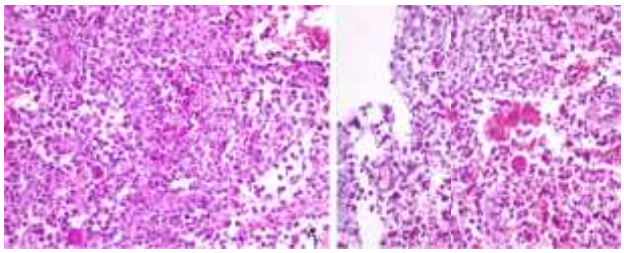

Figure 3 Picture (10X, 20X) demonstrating the histological picture of the biopsy specimen. There are numerous histiocytic cells with bland looking ovoid nuclei and grooved or clefted nuclei. Many multinucleated giant cells were seen in a background of clusters of eosinophils and lymphocytes.

The child was managed conservatively with regular follow up at 3 months interval. At each visit the lesion was evaluated radiographically. The size of the lesion was gradually receding in size and it showed complete healing at the end of twelve months. At 15 months follow up, the child was completely asymptomatic with normal radiograph of the femur.

DISCUSSION

In this case report, the diagnostic challenge and treatment of a rare bone tumor has been presented. Solitary EG of bone is a rare benign tumorous condition and there are handful of cases in literature [2-10]. Clinically most of the children present with pain and swelling and very uncommonly as pathological fracture. Plain radiography provides a clue about the bony lesion, but a specific diagnosis cannot be made without biopsy [3-10]. A wide variety of bone lesions may mimic eosinophilic granuloma; these include infections, traumatic lesions, and neoplasms [3-10].

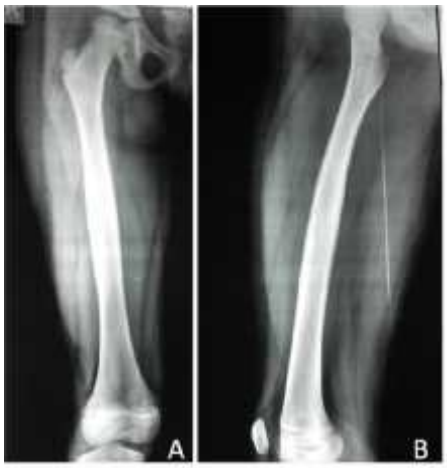

Figure 4 Six months later, the lesion has reduced in size.

EG is rarely seen in long bones below the knee and elbow. The solitary diaphyseal lesion is usually lytic, round or oval, and expansile, with ill-defined or sclerotic margins. There may be expanded medullary cavity with cortical thinning, intracortical tunnelling, or erosion of the cortex, and sometimes may be an associated adjacent soft-tissue mass. Laminated periosteal new bone formation is common around the involved segment of bone. These radiographic features mimic subacute osteomyelitis, brodie abscess and Ewing sarcoma [6,7]. The role of computed tomography and magnetic resonance imaging is also limited; these diagnostic tools only indicate the extent of the lesion. Hence a confirmatory diagnosis is not possible without biopsy [6,7]. The index case had similar presentation and we could not arrive at a final diagnosis without resorting to biopsy. Further confirmation is possible by using IHC staining such as S100, CD 1, monoclonal antibody OKT6 or electron microscopy [6,7].

The treatment of Eosinophilic granuloma is controversial with different modes of treatment claiming effectiveness [3-10]. Solitary EG of bones has shown spontaneous remission [4-6,8]. The effectiveness of intralesional steroid, curettage with bone grafting, chemotherapy and radiotherapy have also been proven by many researchers [3-10]. Plasschaert et al. reported the largest study on isolated EG of bone (32 patients) and noticed complete healing of the lesion in skeletally immature patients (17 patients). Six patients were treated with biopsy alone where as remaining 11 patients were treated with curettage (with or without bone grafting). All the lesions healed without any recurrences. They concluded that EG of

bone in immature skeleton usually heals by itself where as in skeletally mature patients it needs aggressive surgical treatment [8]. The prognosis of isolated solitary bony EG is remarkable. Mc Cullough in his series of 43 patients (36 solitary and 7 multiple) observed that 31 out of 36 solitary lesions healed and 5 became polyostotic [6].

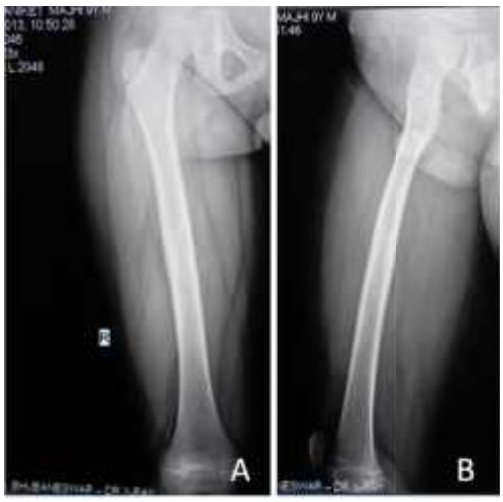

Figure 5 After 15 months, the lesion has completely healed.

In an interesting article by Han et al., no significant difference in radiological healing and functional recovery was observed between children (33 children with isolated bone EG) treated with chemotherapy/ excisional surgery and indomethacin. They noted more number of complications in children treated with anticancer drugs but indomethacin was well tolerated [9]. Munn *et al.* also observed complete healing and pain relief after indomethacin treatment in six patients who had isolated bone involvement [10]. The proposed mechanism of action of indomethacin in treating skeletal EG is that it inhibits prostaglandin E2 production by the LCH cells.

Some authors have reported satisfactory outcome following intralesional steroid injections. How local steroid works in EG of bone has not been elucidated but most probably it might be working similar to indomethacin. Radiotherapy has also controversial role and few authors claim successful outcome with this treatment [5,6].

From the above discussion, it is very difficult to decide the best treatment for a child with isolated bone EG. However the following step wise approach seems wise and practical. All isolated bone lesion should undergo biopsy (may be percutaneous) to confirm the diagnosis. Regular clinical and radiological follow up should be carried out to look for the behaviour of the lesion. As indomethacin is a cost effective non-invasive treatment, it may be administered. We prescribed various analgesics (Ibuprufen/ Dicofenac/Indomethacin) to our patient and that may have helped in healing of the lesion. Intralesional steroid may also be tried if the lesion progresses in size. Aggressive surgical approach, chemotherapy and radiotherapy should not be instituted unless the above treatment fails. A large osetolytic lesion with impending fracture should be stabilized/ splinted.

CONCLUSION

Isolated EG of bone in children often arises a radiological confusion with subacute osteomyelitis and Ewing sarcoma. Conclusive diagnosis without a biopsy is impractical. The lesion may have spontaneous resolution, and hence a regular clinical and radiological follow up may be carried out to look for the behaviour of the lesion.

Clinical message

Thigh pain with radiological picture showing 'onion-pill appearance' in the femoral diaphysis does not necessarily indicate towards Ewing sarcoma and may be other rare benign bone conditions. Because of the nonspecific clinical and radiological pictures in Eosinophilic granuloma and being a rare condition of bone, clinical diagnosis is often not precise. The diagnosis solely relies on histopathological findings. The lesion may have spontaneous resolution and hence regular observation for a period of time may be needed to look for the behaviour of the disease when there is no evidence of impending fracture.

Competing interests: None
Patient consent: Obtained

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