Case Report

Solitary Eosinophilic Granuloma of the Radius: A Case Report

Junu Devi*[1], Sukanya Kalita[2]

[1]Department of Pathology, Gauhati Medical College, Guwahati, Assam. India
[2]Department of Pathology, Wintrobe Hospital, Guwahati. Assam. India

Keywords: *Eosinophilic granuloma, Bone tumor, IHC*

ABSTRACT

Eosinophilic granuloma (EG) is an uncommon tumor of bone and soft tissue usually presenting in children. The clinical and radiological features of EG are highly valuable but not sufficient for diagnosis. Histopathology and immunohistochemistry give confirmatory diagnosis. We present here a case of EG localized to the radial head which was diagnosed histopathologically. Immunohistochemistry was done for CD1a, which was positive in our case confirming the diagnosis.

***Corresponding author:**
Dr Junu Devi, C-3 Kanika Apartment. Noonmati Lastgate,Guwahati.Assam India-781020
Phone: +91- 9435144568
E-mail: drjdevipath@gmail.com

Introduction

Eosinophilic granuloma (EG) is a form of Langerhan cell histiocytosis (LCH) and was first described by Lichtenstein and Jaffer in 1940.[1] In 1953 Lichtenstein used the term Histiocytosis X for a group of diseases that included EG, Hand-Schuller-Christian disease and Letterer-Siwe disease.[1] All of them share common histogenesis and have similar morophological findings. It is considered a neoplasm of the mononuclear phagocytic system of unknown etiology. The disease is characterized by clonal proliferation of a special kind of histiocyte of the antigen- presenting dendritic type called the Langerhan cell (LC).[2]

LCH predominantly affect children and young adults, but it can be found in any age group. The estimated incidence is about 5 per million, with most cases occurring in childhood.[3] Solitary EG accounts for the majority of the LCH cases usually involving bone and less commonly the lymph node, skin and lung.[4] The pathogenesis of LCH is ill understood, whether it is a reactive process or neoplastic is still not defined. Spontaneous resolution occurs is 10-20% of patients and hence initial period of observation is often advisable. LCH is the preferred terminology over histiocytosis X. Among all three entities EG has got good prognosis; it may regress spontaneously and is extremely radiosensitive.

Case Report

A six year old girl presented with complaints of pain and swelling over the right forearms for one and half months. There was no history of trauma, systemic examination was unremarkable, local examination revealed an ill defined swelling 3X1cm in diameter in the distal part of right forearm which was firm immobile and slightly tender. The overlying skin appeared normal.

Routine hematological and biochemical investigations were within normal limit, X-ray radius showed a well demarcated 3x1cm oval osteolytic lesion in the head of the radius. In addition there was no soft tissue involvement outside the bone. Although clinical findings suggested diagnosis of tubercular osteomyelitis radiological findings did not favour such diagnosis. FNAC was done but smears were inadequate - scanty material showing mostly inflammatory cells predominantlty neutophils and eosinophils along with occassional histiocytic cells in a haemorrhagic background therefore repeat FNAC was advised. However biopsy material (curratage) was sent by the surgeon. Histopathological examination of the curetted material revealed proliferation of atypical Langerhan cells mixed with eosinophils and multinucleated histiocytic giant cells which was compatible with EG (Fig1). Immuno-histochemically the membranes of the Langerhan cells were positive for CD1a.(Fig2).She was treated by curettage and doing well on follow up.

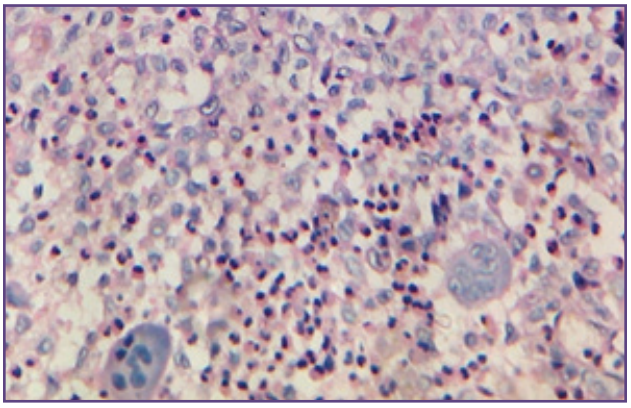

Fig. 1: Microscopic picture of Eosinophilic granuloma (H &E ;X400)

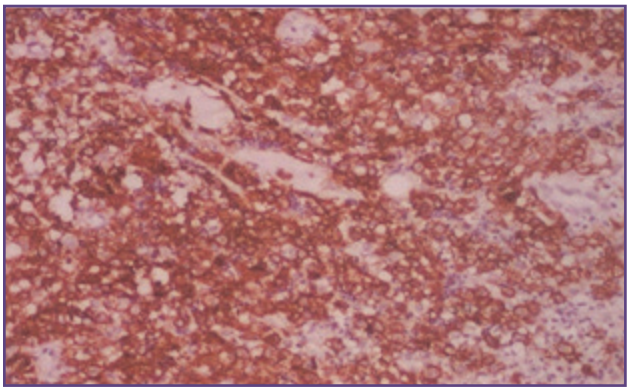

Fig. 2: Immunohistochemical stain: Positive for CD1a

Discussion

EG is a rare bone tumor representing less than 1% of all bone tumors.[5] It is the mildest and commonest form of LHC.[6] It commonly affects children and young adults, particularly males. Any bone can be involved; the more common sites are the skull, mandible, spine, ribs and the long bones. [7] It can be asymptomatic or can present as localized pain, tender swelling and fever.

Patient with EG fall into two groups - disease limited to either bone or soft tissue or combination of both. LCH can have a benign course or sometimes can present with a very diffuse disease.[8] LCH can be subdivided in to three clinico-pathological entities acute disseminated LCH (Letterer- Siwe disease) unifocal and multifocal unisystem LCH (EG), multisystem LCH (Hand Schuller Christian disease). It has a highly variable course with partial or complete healing of the lesion, recurrence after treatment, progression or spontaneous remission without treatment.[9]

The radiological depiction of LCH (EG) is necessary as to determine the activity and nature of the tumor, plain X-ray depicts the size and borders of the lesion. CT and MRI demonstrate the exact size and borders of the tumor as well as the situation of the surrounding tissues and a probable hematoma. The clinical and radiological findings are often not specific enough to determine diagnosis.[10] Cytology is very helpful in diagnosis of EG of bone.[4] The classic cytological features of EG include high cellularity composed of sheets and scattered Langerhan cells with characteristic grooved, folded, indented nuclei admixed with polymorphous population of numerous eosinophils, neutrophils, lymphocytes, plasma cells, multinucleated giant cells and macrophages.[4,11] Charcot Leyden crystal may also be observed in the background.[12] Diagnosis is confirmed by histopathological examination and immunohistochemistry.

Histopathologically EG is characterized by clonal proliferation of Langerhan type histiocytes, which are diagnostic.[13,14] These contain Birbeck granules (electron microscopic feature) whose role is yet unknown. Eosinophils, lymphocytes, fibroblasts, foam cells are not pathognomonic. Morphologically the key feature is the identification of Langerhans cells with characteristic grooved, folded or indented nuclei in the appropriate melieu that includes variable numbers of eosinophils & histiocytes including multinucleated forms.[4,11] An ultra-structural hallmark of LCH is the characteristics tennis racket-shaped Birbeck granules in the cytoplasm. [6]Immunohistochemically LCH is positive for CD1a and s100 protein, Langerin (CD207),MCH classII, PNA (peanut agglutinin).[18] CD1a is diagnostic.

Important differential diagnoses are osteomyelitis, lymphoma, chondroblastoma, Ewings sarcoma, metastatic carcinoma.[4]A possibility of osteomyelitis, especially of tuberculous nature, is the most important differential diagnosis particularly in our country where tuberculosis is so common. But eosinophilic infiltrate and characteristic nuclear grooves are important differentiating points. In case of lymphoma, presence of eosinophils and histiocytes can mimic Hodgkins lymphoma but identification of Reed-Sternberg (RS) cell should be mandatory for diagnosis of the disease. Again characteristic nuclear features of LH cells are absent in lymphoid cells. Immunohistochemical markers can also be helpful in differentiating the diseases.[4] Another differential diagnosis chondroblastoma also show nuclear groove and indentation and thereby resemble LC. However, LCH has a polymorphous population of cells including neutrophils, eosinophils and foamy histiocytes, rather than bimorphic population of chondroblasts and osteoclastic giant cells typical of chondroblastoma. LCH also present with less numerous osteoclast-like giant cells.[15] In Ewing sarcoma characteristic small round cell morphology makes it easily distinguishable from LCH. Metastatic carcinoma especially renal cell carcinoma overwhelmingly exhibit a clustered cell morphology and epithelial features.[4]

The etiology of LCH is unknown and there is continuing debate whether this condition is neoplastic or nonneoplastic. Studies of the x-linked androngen receptor gene have demonstrated a monoclonal proliferation of Langerhans cells in all major clinical syndromes.[6] Senechal et al[16] proposed that enhanced cell survival rather than uncontrolled LC proliferation as in neoplasia, is likely to play a major role in the maintenance and dissemination of these slow-growing tumors.

The clinical course is related to the number of organs affected at presentation, with an overall survival of 95% for patients with unifocal disease;10% of patients with unifocal disease eventually progress to multisystem disease.[6]With multifocal disease,60% have a chronic course,30%achieve remission, and mortality is up to 10%.[17]

Management of LCH (EG) is controversial because of unpredictability of outcome and possibility of spontaneous healing. Modalities vary from observation, curettage intralesional steroid, low dose radiation, high dose systemic corticosteroid and chemotherapy. Low dose radiation in 4-6 fractions may be used when the disease is extensive, inaccessible or if it threatens an important structure.[8] Solitary bone lesion (EG) may be amenable to excision or limited radiation, however systemic disease often requires chemo-therapy, use of systemic steroid is common, singly or as an adjunct to chemotherapy.

Conclusion

EG is an uncommon entity which poses difficulty in its diagnosis due to its variable clinical presentations. Biopsy and immunohistochemistry give confirmatory diagnosis.

Ackowledgement

None

Funding

None

Competing Interests

None declared

References

1. Char DH, Ablin A, Beck stead J. Histiocytic disorder of orbit. An Opthalmol 1984; 16:867-73
2. Jon A, vinay K. White cells, lymph nodes, spleen and thymus. In: Ramzi SC, Vinay K, Tucker C eds.

Robbins pathologic Basis of Disease 6th edn W.B. saunders Co. 1999:685.

3. Nicholson HS, Egeler RM, Nesbit ME. The epidemiology of Langerhen cell histiocytosis. Hematol oncol clin North Am 1998; 12: 379-84.
4. Jain A, Alam K, Maheshwari V, Jain V, Khan R. solitary Eosinophilic granuloma of the Ulna: Diagnosis on fine needle aspiration cytology. Journal of Cytol 2008; 25:153-6.
5. Ando A, Hatori M, Hosaka M, hagiwara Y, Kita A, Itoi E. eosinophilic granuloma arising from the pelvis in children: A report of three case, Upsala J med Sci 2008: 113: 209-16.
6. Weiss LM, Grogan TM, Muller hermelink stein H, Dura T, Favara B, paulli M, Fller AC. Langerhan cell histocytosis. In: Jaffe ES, Harris NL, stein H, Vardiman JW, editors, pathology and genetics of tumor of haematopoietic and lymphoid tissue. Lyon: IARC Press; 2001; P: 280-2.
7. Kaul R, Gupta N, Gupta S, Gupta M. Eosinophilic granuloma of skull bone. J Cytol 2009; 26: 156-7.
8. Kramer TR, Noecker RJ, Miller JM, Clark LC. Langerhan cell histiocytosis with orbital involvement. Am J opthalmol 1997; 124:814-24
9. Chacha PB, Khong BT. Eosinophilic granuloma of bone. A diagnostic problem. Clin orthop 1971; 80:79-88.
10. Kitsoulis PV, Paraskevas G, Vrettakos A, Marine A. A case of Eosinophilic granuloma of the skull in an adult man: a case report. Cases journal 2009;2:9144
11. Mukhopadhyay S, Mitra PK, Ghosh S. Touton like giant cell in lymph node in a case of langerhan cell histiocytosis. Journal of cytol 2007; 24: 191-2.
12. Kemer PV, Mousavi A, Karimi M, Bedayat GR. Fine needle aspiration of Langerhan cell histiocytosis of the lymph node. A report of six cases. Acta cytol 2002;46:753-6
13. Lichtenstein L. Histiocytosis X eosinophilic granuloma of bone, lettere-siwe disease and schuller-christian disease).Further observation of pathological and clinical importance. J Bone joint surg Am. 1964; 46:76-90.
14. Sai S, Fujii K, Masui F, Kida Y. Solitary eosinophilic granuloma of the sternum. J orthop sci 2005; 10: 108-111.
15. Fanning CV, Sneige NS, Carrasco CH, Ayala AG, Murrary JA, Raymond AK. Fine needle aspiration cytology of chondroblastoma of bone.Cancer 1990;65:1847-63
16. Senechal B, Elain G, Jeziorski E, Grondin V, Patey-Mariaud de serre N, Jaubert F, et al. Expansion of regulatory T- cells in patients with Langerhan cell histiocytosis. PLoS Med 2007;4:e253
17. Komp D, EI Mahdi A, Starling K, Easley J, Vietti T, Berry D, et al. Quality of survival in histiocytosis X: A Southwest Oncology Group study.med Pediatr Oncol 1980;8:35-40.
18. Kumar N, Sayed S, Vinayak S. Diagnosis of langerhan cell histiocytosis on fine needle aspiration cytology. A case report and review of the cytology literature. Pathology Res Int 2011;2011;439518.