POLYOSTOTIC FIBROUS DYSPLASIA: BIZARRE AND A RARE CASE REPORT

Dinesh Rajandran[1], Goh Teik Chiang[1], Rubinderan Muthusamy[1], Hesootharan Govindarajan[1], Lai Min Chung[1], Jeeva Shanumugam[1]

[1]Hospital Lahad Datu

Introduction: Fibrous dysplasia (FD) is an uncommon benign condition which was initially described by Schottenstein in 1938 and Jaffe in 1942. It is classified into monostotic which is 70-80% of cases and polyostotic in 20-30% of cases. It accounts for about 2.5% of all bone tumors. It is associated with mutations encoded by GNAS1 on chromosome 20. Osteoblasts undergoes increase in proliferation and faulty differentiation leading to fibrotic bone matrix. Polyostotic FD (PFD) is more aggressive and progresses until skeletal maturity and then becomes quiescent.

Discussion: A 30-year-old female presented with limp and pain in her right thigh in clinic. Patient had previous history of right femur fracture 3 years ago which was treated conservatively. On x ray, there was multiple multilocular cystic lesions with ground glass diffuse rarefactions throughout the right femur. Similar lesion was seen over right humerus, base of skull showed hyperostotic bone formation. CT scan showed a radio dense mass involving right zygomatic, maxilla and mandible causing facial asymmetry. Bone biopsy showed characteristics of FD. The patient was managed with bisphosphonates and observed closely in our clinic.

Conclusion: FD results from defect in osteoblastic differentiation affecting final maturation of bone. It usually manifests before the 3rd decade of life. This case fell within the age group described. PFD more frequently involves skull and facial bones as presented in this case. PFD may occur alone or as part of McCune- Albright Syndrome (MAS). Two thirds of patient are symptomatic with initial symptoms of pain over involved limb usually associated with limping, pathological fractures or both. The characteristic sign of disease, 'shepherd-crook deformity' due to severe coxa vara deformity is well demonstrated in this case. Currently there are no clearly defined systemic therapies for PFD. Small, uncontrolled trials using second generation bisphosphonates (Pamidronate) suggest that bisphosphonates may be effective.