

Physiotherapeutic Approach in Polyostotic Fibrous Dysplasia: A Case Study

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Abstract

The present case study explains the rare condition of polyostotic fibrous dysplasia of a 35 year old female. The case could be stated as a McCune-Albright syndrome. The protocol of physiotherapy clinical evaluation and conservative physiotherapeutic treatment approach has significantly improved the pain status and physical functioning of a patient at 4 weeks of intervention. The physiotherapy treatment protocol will help to improve the health status in fibrous dysplasia cases.

Key words: Polyostotic fibrous dysplasia, McCune-Albright syndrome, Physiotherapeutic approach, TENS, Physiotherapy

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Introduction

Polyostotic fibrous dysplasia is a relatively rare benign skeletal disorder which has been caused by defective osteoblast differentiation, abnormal fibroblast proliferation and replacement of normal bone structures with fibrous connective tissue (1, 2). There is a failure in remodelling of immature bone into mature lamellar bone that leaving a mass of immature bone woven in dysplastic fibrous tissue (3). The McCune-Albright Syndrome is a complex disorder that is affecting the bone, skin and endocrine system. It consists of three conditions; the unilateral polyostotic fibrous dysplasia, ipsilateral cafe-au-lait spots on the skin and endocrine disturbance (4, 5). The case which has been represented over, has all the three conditions and it can categorised into McCune-Albright Syndrome.

Case report

A 35 year old female had come to the orthopaedic department with a primary chief complaint of upper back and chest pain. Pain was predominantly more on left side compared to right

side. Her secondary complaints were left arm pain and left knee pain. The pain was lasting since 5 to 6 months and it has gradually increased. Patient has reported the history of fall, twice in her childhood. The impact of fall was on left side of her body during the both episodes and there was a suspect of fracture on left side of femur. Since 5 to 6 months, she has started experiencing pain in her upper back and chest with a history of breathlessness in between. Pain was dull aching and continuous in frequency. Her pain was 7 out of 10 on Numerical Pain Rating Scale (NRS). The aggravating factors were the spine movements; especially forward and backward bending, and bed transfer activities. The side lying position was relieving for her. Patient had visited to Aurangabad Hospital and her investigations had revealed the rare condition of bone disorder. Her MRI of left shoulder had revealed the polyostotic fibrous dysplasia involving proximal metaphysis and diaphysis of left humerus and in scapula. Her CT scan of neck, abdomen-pelvis, and chest had revealed the polyostotic fibrous dysplasia involving the left femur, left humerus, left acromion process and left ischium, left inferior ischio-pubic ramus and left 5th rib on anterior aspect. The x-ray investigations revealed the extensive bone expansion of left humerus (Figure 1), left

Table 1. Physiotherapy treatment approach

Intervention	Duration
TENS	20 minutes, thrice a week for 4 weeks
General mobility exercises: Active ROM exercises	10 repetitions/session, 2 sessions/day for 4 weeks
Strengthening exercises: Limb and Core Muscle strengthening	10 repetitions/session, 2 sessions/day for 4 weeks
Cold pack	15 minutes, twice daily for 4 weeks



Figure 1. X-ray investigations revealed extensive bone expansion of left humerus

radius, and left femur with thinning of cortex of proximal tibia. (Figure 2) In endocrine test investigations, her parathyroid levels were high suggesting the hyperparathyroidism.

The female was prescribed pain medications earlier but they gave temporarily relief. And hence she had come to the MGM Hospital, Navi Mumbai for further detailed evaluation and appropriate treatment.

The positive findings of clinical evaluation based on physiotherapy protocol are summarized as:

On observation- The attitude of limb: The left arm was flexed, left lower leg was externally rotated and left foot was everted. The swelling was presented over the left thigh with a shiny appearance of skin. The cafe-au-lait spots were presented over the left side of her back. (Figure 3) The deformities were presented; the cubitus valgus of left arm of 25 degrees, flexion deformity of left knee of 15 degrees and limb shortening of left lower limb.

On palpation- There was a local rise in the temperature due to fever. Grade 1 tenderness was presented over left spine of



Figure 2. X-ray findings revealed extensive bone expansion of left femur with thinning of cortex of proximal tibia

scapula, medial and lateral border of scapula and on left upper trapezius region. Grade 4 swelling (Hard, Bony type) was presented over left distal one-third of thigh and proximal tibia.

On examination- In Range of Motion (ROM) assessment, her range of internal rotation of left shoulder was restricted. In muscle strength assessment based on Medical Research Council (MRC) grading, her shoulder, hip and knee muscles were grade 4. She was able to complete the range against the gravity on submaximal resistance. The limb-length discrepancy was significant. The true limb-length discrepancy showed 7 cm shortening of left lower limb compared to right side. There was a reduced chest expansion at T2 (Axilla) and T4 (Nipple) levels.

Physiotherapy treatment approach: (Table 1)

The goals of treatment were:

To reduce the pain

To optimize the physical function

The conservative physiotherapy treatment for pain relief consisted of an application of Transcutaneous Electrical Nerve Stimulation (TENS) over left arm and left knee for 20 minutes. The session was given for thrice a week for 4 weeks. The physical functioning was improved by performing general mobility



Figure 3. Presence of cafe-au-lait spots on left side of back

exercise and strengthening exercises for muscles of shoulder, hip and knee twice a day for 4 weeks. The cold pack was given to reduce the swelling around the thigh and knee for 4 weeks.

The patient showed a significant improvement in her pain status which has been reported as 3 out of 10 on NRS and improved functioning in her Basic Activities of Daily Living (BADL) after 4 weeks of session.

Discussion

The case which represents the rare condition of fibrous dysplasia is involving the multiple bone sites interpreting as a polyostotic fibrous dysplasia. The present case could be stated as a McCune-Albright syndrome as the all conditions of syndrome were positive in this case.

The conservative physiotherapeutic treatment approach showed a significant improvement in her condition at 4 weeks of intervention. One of the case study by Sagmeister *et al.*, have also stated that the patient was managed conservatively for 8 weeks of intensive physiotherapy and was showed a greater recovery and was returned to baseline at 3 months of post injury (6). Being a rare condition, the physiotherapy treatment approaches and protocol for such cases have not emerged in a greater extent in the field. Thus the present case study will provide some beneficent outcomes for treating patients with fibrous dysplasia.

Conclusion

The polyostotic fibrous dysplasia is a rare case. The physiotherapy treatment approaches are effective in managing the subsequent pain and physical functioning in patients with fibrous dysplasia.

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All authors made substantial contributions to conception, design, acquisition, analysis and interpretation of data.

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