Nat. Volatiles & Essent. Oils, 2021; 8(5): 712 - 717

NVEO
Natural Volatiles &
Essential Oils

A Rare Case Report on Management of Fibrodysplasia Ossificans Progressiva with Ventricular Septal Defect

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ABSTRACT

Background: Fibrodysplasia ossificans progressiva is a progressive fibrodysplasia. It's a rare genetic condition. This disorder is characterised by soft tissue ossification and congenital stigmata of the limbs. the case report of a male child who was tracked for 10 years, starting at the age of three years and nine months when the diagnosis was made. A newborn was born with a ventricular septal defect and bilateral hypoplasic hallux valgus. The transsternal method was used to fix this flaw. When the 32-month period was over, Following that, neck motion was restricted, and ectopic ossification developed. Oral prednisone or other anti-inflammatory medicines were used to treat the sickness of four crises aggravation. The medicine sodium etidronate (5 to 10 mg/kg/day) was prescribed for roughly six years, but treatment was stopped for a few days due to osteopenia. The course of disease has been relentless, with restrictions to severe movement.

Patient specific information: A male child was admitted in acharya Vinoba Bhave rural hospital on dated 10/07/2021 with the chief complaint of pain, fever, swelling, reduced mobility in joints, knees, wrists, shoulders, spine, and neck, ossification of short toe and thumb, progressive fusing of neck vertebrae, muscle and fibrous tissue , tumor like swelling in shoulder and other joints, abnormal lateral curvature of the spine etc.

Main symptoms or important clinical findings: A male child born to a non-consanguineous parents as their firstborn admitted to acharya Vinoba bhave rural hospital on dated 10/07/2021 with the chief complaint of pain, fever, swelling in tendons, the patient had undergone various investigation like careful history ,Complete blood count, X-ray, CT scan, MRI, and other physical examinations are all available.

Conclusion: Fibro dysplasia ossificans is a progressive fibro dysplasia. It has been characterized as a connective tissue disorder due to overexpression of a bone morphogenetic protein. There is no any curative therapy is available& their management is dependent on the child's deformity, with a focus on preventing aberrant ossification. As a result, clinician awareness of the disease is critical.

Keywords: Muscle Biopsy, Stiff Spine, Etidronate Treatment, Myositis Ossificans

INTRODUCTION

It is Genetic illness is a very rare occurrence This disorder is characterised by soft tissue ossification and congenital stigmata of the limbs. We provide a case study of a male kid who has been followed for 10 years, beginning when the diagnosis was made at the age of three years and nine months. A male infant was born with a ventricular septal defect, which was fixed when he was 32 months old using a transsternal technique. Ectopic ossification arose as a result of the restriction of neck motion. Oral prednisone or other antiinflammatory medicines were used to treat the four crises. It was given to patients on an as-needed basis for nearly six years at a dose of 5 to 10 mg/kg/day.[1]

The disease has progressed steadily, with considerable movement limits, including the chest wall. The information about the case report is displayed in a review of the case report.

We look at the case report criteria for diagnosis, as well as the essentials of disease management and treatment. Myositis ossificans is often replaced with fibrodysplasia oossifican progressiva. Ectopic osteogenesis takes place. Muscle fasciae, ligaments, tendons, and joint capsules, rather than the muscle fibres themselves, include connective tissue. These could imply non-specific or secondary pathogenic changes.[2]

Because of the overexpression of a bone morphogenetic protein, fibrodysplasia ossificans progressiva (FOP) was recently classified as a connective tissue disorder.

Therefore the Because no curative therapy is available, this disease is managed solely on the basis of the child's malformation and also depend on severity of symptoms and Preventing abnormal ossification is particularly important. As a result, it is critical that practitioners become more aware of the disease. [3]

A case report of a patient with FOP who was followed for ten years is presented. We look at the diagnostic criteria, therapy options, and management guidelines for FOP. Basic surgical techniques, such as muscle biopsy, have undesirable repercussions are highlighted, as they may promote ectopic ossification and are not a contributing disease for the diagnosis (FOP).

A male child was considered in good condition. An Due to a deformity of the great toes, an orthopedic consultation was needed. The infant was discharged after a simple roentgenogram of the feet.

Routine follow up of paediatric There was a heart murmur elsewhere, and the somatic development was retarded. A transsternal procedure was used to address a congenital interventricular septal defect. despite the fact that cyanosis was not a complaint.

The hemogram, erythrocyte sedimentation rate, serum calcium, phosphorus, alkaline phosphatase, creatine phosphokinase, alanine and aspartate transaminases, urinalysis, urine calcium and phosphorus, and creatinine clearance all came back normal. As the condition progressed, alkaline phosphatase levels fluctuated between normal and slightly high. There were areas of abnormal uptake in the soft tissue of the left lumbar paravertebral region, right posterior chest wall, and posterior side of the right arm on a recent 99mTc-MDP scan, with consistent tissue calcifications of varying degrees of uptake. Throughout four periods, heterotopic ossification was monitored in the dorsal thoracic area, inferior right abdomen, biceps, medial left arm, and forehead. Heterotopic progressive osteogenesis and congenital anomalies of the great toes are epifeatures. Neonatologists, paediatricians, neurologists, orthopaedic physicians and surgeons, rheumatologists, and other auxiliary personnel involved in the diagnosis and treatment of neuromuscular diseases should be aware of the condition.[4]

Myositis ossificans is prefered over fibrodysplasia ossificans progressiva (FOP). Nonspecific alterations may occur, as well as subsequent pathogenic changes. Because ectopic osteogenesis occurs in the connective tissue within muscles, fasciae, ligaments, tendons, and joint capsules rather than in the muscle fibres themselves, the term fibrodysplasia ossificans progressiva is preferred over myositis ossificans. It's possible that nonspecific, possibly harmful modifications are present.[5]

Due to overexpression of a bone morphogenetic protein, FOP has recently been defined as a connective tissue condition.; nevertheless, because no curative medication is available, management is focused on the patient's deformity. It's especially effective at preventing aberrant ossification. As a result, increased awareness of the disease among practitioners is critical.

Minor surgical procedures such as muscle biopsy, which may cause ectopic ossification and is a bone morphogenetic protein, are noted as having detrimental consequences. Because no curative therapy is available, patients are managed based on their malformation. It's especially effective at preventing aberrant ossification. As a result, there is a greater awareness of the disease.

PATIENT INFORMATION

Patient specific information:

A male child was admitted in Acharya Vinoba Bhave rural hospital on dated 10/07/2021 with the chief complaint of pain, fever, swelling Tendon ossification of muscle and fibrous tissue, progressive fusing of neck vertebrae joint, knee, wrist, shoulder, spine, and neck mobility restrictions short toe and thumb, tumor like swelling in shoulder and other joints, abnormal lateral curvature of the spine etc.

Because of a deformity of the great toes, an orthopedic consultation was requested. The infant was discharged after a simple roentgenogram of the feet. A heart murmur and impaired somatic development were discovered during routine paediatric follow-up elsewhere. Although cyanosis was not a complaint, a transdermal technique was used to treat a congenital interventricular septal defect.

Primary concerns and symptoms of the patient:

Present case visited acharya Vinoba Bhave rural hospital at pediatric ICU on dated 10/07/2021 with chief complaint of pain, fever, swelling in tendons, increasing Neck vertebrae fusion, ossification of muscle and fibrous tissue, reduced joint mobility, knee, wrist, shoulder, spine, and neck short toe and thumb, tumor-like swelling in shoulder and other joints, improper lateral curvature of the spine, and so on. The primary symptom that was noticed at the time of admission.

Medical family and psychosocial history:

Present case had medical history of ventricular septal defect and fibrodysplasia ossificans progressiva . He took a treatment for that. He belongs to nuclear family. Fibrodysplasia ossificans progressiva was a medical condition that her grandfather suffered. He was intellectually sound and aware. He had maintained positive relationships with doctors, nurses, and other patients.

Relevant past intervention with outcome:

Present case had admitted in Acharya Vinoba Bhave rural hospital with chief complaint of pain, fever, swelling Tendon ossification, ossification of muscle and fibrous tissue, progressive fusing of neck vertebrae joint, knee, wrist, shoulder, spine, and neck mobility restrictions. After plain roentgenogram of the feet is done and hemogram after investigation fibro dysplasia ossificans progressive was observed and his outcome was good .

Clinical findings

The patient was conscious his body built was moderate A male child was considered in good condition. Because of a deformity of the great toes, an orthopedic consultation was requested. The infant was discharged after a simple roentgenogram of the feet. A heart murmur and impaired somatic development were discovered during routine pediatric follow-up elsewhere. Despite the absence of cyanosis, a congenital interventricular septal defect was corrected using a transsternal method. The patient's vital signs were normal.

Timeline

Present case had no any history of medical problems. Patient consider in good condition. The infant was discharged after a simple roentgenogram of the feet. A heart murmur and impaired somatic development

were discovered during routine pediatric follow-up elsewhere. Despite the absence of cyanosis, a congenital interventricular septal defect was corrected through a transsternal method.

Diagnostic assessment:
Patient diagnosed with fibro dysplasia ossificans progressive based on patient history, physical examination, and additional investigations and other investigation are normal. In most circumstances, a patient's deformity of the big toe, as well as quickly changing swelling on the head, neck, or back, can be used to determine whether fibrodyspcan be used to determine whether fibrodysplasia ossificans progressiva is present (FOP). Other investigations were also carried out.

No challenges experience during diagnostic evaluation.
DIAGNOSTIC ASSESSMENT
DIAGNOSTIC CHALLENGING : - No any challenges during the diagnostic evaluation .
DIAGNOSIS:- After physical examination and investigation doctor diagnose a case of a fibrodysplasia ossificans progressive.

Prognosis
The prognosis of disease was good .

Therapeutic intervention:
Present case took the presently there is a no definitive medical management for fibrodysplasia ossificans progressive treatment and the only management strategies supportive the current management consideration are classified as class l , class ll and class lll medication along with few muscle relaxant and special medical consideration including injury prevention scalp nodules maintaining respiratory health and capacity, infection, precautions, perticularly during the flu season.
Before the advent of heterotrophic ossification and even before the appearance of soft tissue lesions, definite molecular diagnosis is now available for FOP. During the early phases of disease development, when diagnosis is most likely to occur, no adjustments in therapeutic intervention were made. He was took all supportive treatment and outcome was good .
No any change in restorative .

FOLLOW -UP AND OUTCOMES
CLINICAL AND PATIENT ASSESSMENT OUTCOMES : Patient condition was improved .
IMPORTANT CHECK OUT INVESTIGATION AND OTHER TEST RESULTS :- To preventing the progression of disease and trying to reserve any signs and symptom that has been appeared. Doctor advised follow up after blood investigation and other examination to know the further disease progression .
INTERVENTION ADHERENCE AND TOLERABILITY :-patient took all prescribed medication regularly but sometimes he was refuse to take medication .he also followed the dietitian advised . dietician was advised healthy food and rich in calcium and multivitamin supplementation . His intervention adherence was satisfactory .
Adverse and unanticipated events :- No any
Nursing perspectives:
Monitored the vital signs of male child per hourly, monitored the heart rate of male chlid .

DISCUSSION :
Present case was admitted in A.V.B.R . Hospital with the chief complaint of mobility of restrictions in neck , fatigue , weakness , fatigue, swelling in hand etc .after physical examination and investigation doctor diagnosed a case of fibrodysplasia ossificans progressive. calcium and multivitamin should be provide for

supplementation. FOP should be diagnosed as possible as and it also depends on patient history and radiological finding .In FOP the radiologic examination is needed and sub types of big toes can diagnosed with the second decade . So the early management of FOP is needed .The FOP related to the degree of dysmorphism and it is not visible in the absence of conditions and hand malformation commonly related with the condition .

The most common anomallies are first metacarpal and bracynesophalenge of fifth finger.[6] FOP occur continuously by trauma eg.intra muscular injection that involves vaccine local anesthesia near the muscle and vebepuncture . From clinical and radiograph FOP is diagnosed . (foot and hand stigmata). Biopsy can cause recurring ossification of the location, which can be as bad as the original lesion. Acute or persistent limb swelling is another clinical manifestation of FOP. Tissue turgor increase with multifactorial aetiology. circumference of limbs expand in one or more places. In people with FOP, calcemia and phosphatemia is a laboratory test for FOP which is frequently normal or changes in quantity. For early detection of disease the assessment and ossification is important for diagnose FOP the genograms of feet was done and development may be possible with bone scintigraphy-MDP.

Whole medical management of FOP is conservative also currently effective medical management not available it base on client need. That is, avoiding situations that could lead to aberrant ossification. The medicine was administered to our patient on an intermittent schedule during a five-year period to prevent osteopenia. However, nodules continued to form during treatment, and bone demineralization became a severe side effect. When a corticosteroid was administered to treat inflammation prior to ossification, the symptoms were often less severe, and the medicine is now advised in these cases. The oral Corticosteroid and intravenous injection given in same time for a good effect [7] and there is no effective treatment for FOP. Few of the related studies were reviewed[8-14].

CONCLUSION:

Because of excessive bone morphogenetic Protein the FOP is known as the Disorder of connective tissue. The management of FOP is depend on clients health condition for that curative treatment does not available. particularly at preventing abnormal ossification . Patient was support to all supportive treatment. The recovery was good.

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