Munchmeyer’s Disease
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ISSN: 1563-4086
Section: Musculoskeletal system
Area of Interest: Musculoskeletal system
Musculoskeletal bone Head and neck Thorax
Musculoskeletal soft tissue
Procedure: Diagnostic procedure
Procedure: Computer Applications-3D
Imaging Technique: Conventional radiography
Imaging Technique: CT-Quantitative
Imaging Technique: CT
Special Focus: Calcifications / Calculi Connective tissue disorders Obstruction / Occlusion Case Type: Clinical Cases
Authors: Dr. Akhilanand Chaurasia, Dr. Shailesh Kumar Singh, Dr. Vaibhav Mishra
Patient: 10 years, female

Clinical History:

A 10-year-old female presented with the chief complaint of progressive inability to open the mouth since the past year and multiple firm round swellings on the vertebral region, right scapula and right occipital region of the skull. She had laterally deviated short first toes of both feet.

Imaging Findings:

1. PA chest showing extensive focal and cord-like ossification of muscles and soft tissue of back, chest, abdomen and visualised right arm giving pattern of branching tree.
2. AP radiograph of both feet showing microdactyly of the great toes and bilateral hallux valgus.
3. Lateral view of thorax and abdomen showing cord-like ossifications of muscles of back
4. Axial CT images of thorax showing ossification of back, chest muscles.
5. 3D Coronal CT image showing posterior view of trunk and neck exhibiting excellent demonstration of soft tissue and muscle ossifications mimicking ‘tree branching pattern’.
6. 3D Coronal CT image showing anterior view of trunk and neck exhibiting excellent demonstration of soft tissue and muscle ossifications mimicking ‘tree branching pattern’.

Discussion:

Munchmeyer’s disease or Fibrodysplasia ossificans progressive (FOP) is a rare autosomal dominant disease [1]. It is also known as myositis ossificans progressive or stone man disease [2]. It was first described by Patin [3]. The worldwide prevalence of FOP is approximately 1 in 2,000,000 [4]. It is most commonly seen between birth and 10 years of age. The mean age of occurrence is three years. Pathologically it is characterized by progressive replacement of muscles, tendons, ligaments, fascia and aponeurosis by bone, leading to progressive stiffness of the adjacent joints. The progressive replacement of the chest wall may lead to pneumonia and death [5]. FOP is characterized by two cardinal features: a) heterotopic progressive osteogenesis, b) congenital abnormalities of the great toes [2]. However it is particularly disabling in children. The most characteristic deformity is microdactyly of both halluces due to a single phalanx in valgus position [6, 7]. This characteristic feature is found in our case also.
The patients affected by FOP are usually confined to bed by the age of 30. FOP primarily involves the neck (50%), dorsal paraspinal region (30%), head (10%) or limbs (10%). The 70% of FOP patients develop temperomandibular joint ankylosis [8]. The ectopic ossification is a hallmark sign of FOP. It can occur during the lifespan of a patient but most commonly it is found between 3 to 5 years of age [8, 9]. Radiologically, myossitis ossificans progressiva can be identified approximately two to four weeks after the onset of the process. In FOP, computed tomography can be used to delineate the central radiolucency encompassed by peripheral density [10]. The radiographs may aid in documenting minor osseous dysmorphism. Bone scintigraphy with 99mTc-MDP demonstrates early heterotopic ossification and helps in assessment of the extent and progression of the FOP [7]. The effective treatment for FOP is still unknown. FOP is managed conservatively by avoiding conditions potentially provocative of abnormal ossification. However administration of calcium chelators such as sodium etidronate and corticosteroids reduces the progress of the disease [10].

**Differential Diagnosis List:** Munchmeyer’s disease or Fibrodysplasia ossificans progressive, Albright hereditary osteodystrophy, Pseudomalignant heterotopic ossification, Progressive osseous heteroplasia

**Final Diagnosis:** Munchmeyer's disease or Fibrodysplasia ossificans progressive

**References:**

Brooke MH (1986) A clinicians view of neuromuscular diseases. 2Ed. Baltimore: Williams & Wilkins 239-242
Patin G (1692) Lettres choixis de feu. Cologne P. du Laurens Tome 1, vol. 5, p. 28
Description: Axial CT image of thorax showing ossification of back and chest muscles. Origin: Chaurasia A, Department of oral medicine and radiology, King George Medical University, Lucknow, INDIA
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Figure 6

Description: Panoramic radiograph showing reduced joint space in left temporomandibular joint

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Description: PA Chest showing extensive focal and cord-like ossification of muscles and soft tissue of back, chest and abdomen mimicking branching tree pattern. Origin: Chaurasia A, Department of oral medicine and radiology, King George Medical university, Lucknow, INDIA.
Description: AP (Antero-posterior) radiograph of both feet showing microdactyly of the great toes and bilateral hallux valgus Origin: Chaurasia A, Department of oral medicine and radiology, King George Medical university, Lucknow, INDIA
Description: Lateral view of thorax and abdomen showing cord-like ossifications of back muscles
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