Myositis Ossificans and Fibrodysplasia Ossifica Progressiva

Myositis Ossificans (MO) is a form of heterotopic ossification where bone tissue forms within muscle or other soft tissues. It exists in two distinct forms:

- **Myositis Ossificans Circumscripta (Nonhereditary)**
- **Myositis Ossificans Progressiva (Hereditary)**, also called **Fibrodysplasia Ossificans Progressiva (FOP)**

1. Myositis Ossificans Circumscripta (Nonhereditary Form)

Etiology and Risk Factors

- Typically occurs following **blunt muscle trauma** (e.g., contusion, strain, or hematoma)
- Common in **young adults and athletes**
- Risk increases with **premature return to activity** post-injury
- Most affected sites: **quadriceps femoris**, **brachialis**, and **deltoid**

Clinical Presentation

- Appears within **1–2 weeks post-trauma**
- Localized **pain, swelling, warmth**, and **restricted joint movement**
- May present with a **firm, tender mass** over the muscle
- ESR and alkaline phosphatase levels may be elevated
- Symptoms gradually improve, unlike **osteosarcoma** (pain worsens over time)

Imaging Findings

- **Plain Radiographs**: Normal early (first 1–2 weeks), then show **peripheral calcification** progressing **centripetally** by 2–6 weeks
- **CT Scan**: Demonstrates **well-defined peripheral ossification** with central lucency
- **Bone Scan**: Increased uptake in the lesion <3 weeks post-injury
- **Ultrasound**: May detect early soft-tissue changes
- **Key Differentiation**:
    - **MO**: Peripheral-to-central calcification
    - **Osteosarcoma**: Central-to-peripheral calcification

Histopathology

- **Early phase**: Proliferation of undifferentiated mesenchymal cells infiltrating muscle
- **2–3 weeks**: Peripheral **osteoid formation**, with immature fibroblasts centrally
- **Mature lesion**: Peripheral **lamellar/woven bone**, central **fibrous tissue**, occasional cartilage component

Management

- **Conservative management** is first-line:
  - **Rest and immobilization**
  - **NSAIDs** (e.g., indomethacin) for pain and to limit ossification
  - **Gentle active ROM** exercises once inflammation subsides
  - **Avoid passive stretching**—may exacerbate the condition
- **Surgical excision**:
  - Considered only after **9–12 months** (when lesion matures)
  - Indicated if function is impaired or pain persists

2. Myositis Ossificans Progressiva (Fibrodysplasia Ossificans Progressiva, FOP)

Etiology

- **Rare, autosomal dominant** genetic disorder
- Caused by mutations in the **ACVR1 gene** (activin A receptor type I)
- Results in progressive **heterotopic ossification** of connective tissue
- Triggered even by **minor trauma**, intramuscular injections, or viral illnesses

Clinical Features

- Onset in **early childhood**
- **Progressive restriction of movement** due to ossification
- Characteristic **malformed great toes** (hallux valgus or short great toes)
- Episodes of **painful soft tissue swellings** precede ossification
- Ossification follows a **predictable anatomical pattern** (cranial-to-caudal, axial to appendicular)

Complications

- Respiratory compromise (due to ossification of chest wall)
- Malnutrition and difficulty with oral intake (jaw fixation)
- Profound physical disability over time

Diagnosis

- **Clinical features + genetic testing (ACVR1)**
- **Imaging**: Extensive soft tissue ossification
- Avoid biopsy or trauma as it may exacerbate ossification

Management

- **No definitive cure**
- Prevent trauma, intramuscular injections, or surgical interventions
- **Short courses of corticosteroids** during flare-ups
- Use of **bisphosphonates**, NSAIDs, or experimental therapies (e.g., palovarotene) under investigation

Key Differentiating Features Between MO and Osteosarcoma

Feature	Myositis Ossificans	Osteosarcoma
Age	Teens/young adults	Teens/young adults
Trauma history	Common	Rare
Pain progression	Improves over time	Worsens over time
Calcification pattern	Peripheral ? central	Central ? peripheral
Location	Diaphysis, soft tissue	Metaphysis, bone
Imaging	Shell-like ossification	Sunburst pattern, Codman’s triangle
Histology	Zonal ossification, benign	Malignant osteoid, atypia