Case 9

History
Shoulder pain.

Diagnosis?

Diagnosis
Osteopoikilosis.

Findings
Multiple, small round or ovoid areas of increased bone density, widely distributed around the shoulder joint are noted as an incidental finding.

Discussion
Osteopoikilosis ("spotted bones") is a radiologic curiosity inherited as a highly penetrant autosomal dominant trait. The bony lesions are asymptomatic. Incorrect diagnosis may lead to confusion with serious conditions, including metastatic disease. Some patients have connective tissue nevi called dermatofibrosis lenticularis disseminata, i.e., Buschke-Ollendorff Syndrome.

Patients are asymptomatic, and laboratory exams are normal. Osteosarcoma has been reported to occur in affected bone. Associated conditions include: dermatofibrosis lenticularis disseminata, scleroderma, syndactyly, dwarfism, endocrine abnormalities, melorheostosis, and cleft palate. Most patients have no other findings.

Radiographically, lesions are seen in the metaphyseal and epiphyseal areas of affected bones. The multiple, sclerotic, roundish lesions vary in size from a few millimeters to several centimeters. They are scattered within the cancellous bone and concentrated near the joint surfaces. The lesions cluster at the end of long bones, around the acetabulum of the pelvis, around the glenoid of the scapula, and in the small bones and cuboid bones of the hands and feet. Other than the pelvis, the axial skeleton is rarely involved. Lesions may increase or decrease in size as well as number.

Bone scan activity is normal. Pathologically, the lesions resemble a bone island. Differential diagnosis includes: sclerotic metastatic cancer, mastocytosis, tuberous sclerosis.

Reference