A 17 year-old boy presented with multiple nodular swelling in right upper limb and feet for 2 years, and proximal muscle weakness for 2 months. Two years ago, the patient noticed a small nodular swelling over the wrist, which later became hard. Over time, he developed multiple nodular swellings without any limitation of daily activities. For the last 2 months, he has developed difficulty in standing from a sitting position. Such types of illness didn’t run in his family, and the patient had no history of taking myopathic drugs.

General examination revealed multiple nodular swellings involving the ulnar surface of the right forearm, 2nd & 5th metacarpophalangeal joint, wrist, right foot, which were nontender, most of them were hard with few firm in consistency, fixed with underlying structure but free from discharging sinus, largest one is (2.2 cm). On CNS examination, only muscle power of the proximal group of the lower limb 4/5, feature of proximal myopathy. Investigation shows Hb 11.2g/dl, ESR 15 mm (1stHr), TC 15,000, CPK: 2881 U/L, S.Creatinine, RBS, S.uric acid, S.calcium, S.albumin, Thyroid function test, S.electrolytes and Urine R/M/E are normal. CRP is negative, SGPT: 110 U/L, SGOT: 223 U/L. ANA and Anti-Centromere Ab are negative. Muscle biopsy features consistent with dermatomyositis. Prednisolone 40mg was administered daily with symptomatic improvement. In a recent follow-up, patient muscle weakness was improved but no exacerbation or resolution of calcinosis was observed.
Discussion:
When calcification is processed any tissue other than bone and teeth is termed calcinosis and can occur in many condition including connective tissue disease, hyper-parathyroidism, renal failure and vitamin D intoxication. Calcinosi may be divided into four categories according to the pathogenesis as follows: dystrophic, metastatic, diopathic and iatrogenic. In connective tissue disease, calcinosis is mostly of dystrophic type and it seems to be localized process rather than an imbalance of calcium homeostasis. Calcinosi in connective tissue disease about 9% patient with seleroderma and 5% to 20% of adult and 40% to 74% of children with dermatomyositis. The existence of calcinosis is indicative of a good prognostic sign of survival but may also be incapacitating.

Dermatomyositis is an idiopathic inflammatory myopathy with characteristic cutaneous manifestation, including heliotroperash, gottronpapules, periungual telangiectasias, photo distribution erythema, poikiloderma and alopecia. Although heliotrope rash and gottron papules are specific feature, calcinosis may occur up to 40% of children or adolescent.

The laboratory hallmark are elevated creatine kinase, aldolase and transaminase, and a characteristic pattern of EMG-spotty muscle necrosis, regeneration, and inflammation are the pathological hallmark. Calcinosi can be a disability complication that may affect the skin, subcutaneous tissue. It occur most during the course of juvenile dermatomyositis. Calcinosi usually occurred two or three year after onset of dermatomyositis, after that the deposition remain stable and spontaneous resolution has been occasionally reported. The cause and mechanism of calcification are unknown. calcium deposition are often in those muscle that were most severely affected during acute phase of disease. Serum calcium, phosphate and urinary calcium values are within the normal range. The calcinosi can be demonstrable both clinically and radiologically. A whole body scan with 99m TC pyrophosphate and CT scan can also identify calcinosi. Aggressive treatment with high doses of prednisolone and physical therapy can decrease the incidence of calcinosi. The use of bisphosphonate in the treatment of soft tissue calcification has varing result. Two studies shows suppression of Gla synthesis by warfarin sodium may prevent deposition and allow for removal of existing calcinosi. Large and localized mass may be remove surgically.

Conclusion:
Calcinosi is often signals a improved prognosis. Spontaneous regression of calcification was reported up to 50% of the cases.

References:
5. Blane CE, White SJ, Braunstein EM, Blane CE. Pattern of calcification in Childhood dermatomyositis. AJR 1984;142;397-400