

Clinical Image

Subcutaneous Nodules in a Child with Ectodermal Dysplasia

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Our patient is a 5 year old male with ectodermal dysplasia and no past history of infectious complications, who presented with 5 weeks of daily fevers and bilateral knee pain. His absence of sweat glands resulting in frequent episodes of hyperthermia had contributed to the delayed diagnosis of fevers. His presentation had been preceded by two episodes of sore throat, which had been treated with two 10 day courses of Amoxicillin. Subsequently he developed an intermittent rash appearing on his arms, trunk, and abdomen, bilateral knee swelling, and multiple palpable lesions on his occiput, lower back, elbows, and hands.

Patient's physical exam was notable for findings of ED such as sparse hair and abnormal dentition. Multiple subcutaneous nodules were present over the bony prominences (Figure 1-4). They were about 1cm in size, firm, non-tender, non-mobile, and located on the occiput, flexor aspect of right wrist, left styloid process, left olecranon, and overlying the thoracolumbar spinous processes. There was bilateral painful swelling of wrists, elbows



Figure 1



Figure 2

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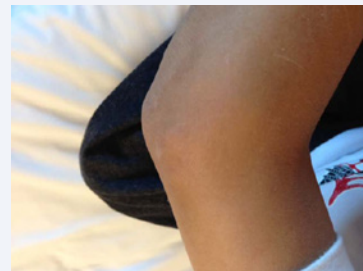
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Figure 3



Figure 4

and knees. Auscultation revealed a II/VI soft systolic ejection murmur, an echocardiogram was found to be normal. Laboratory findings showed an elevated ESR 25 mm/hr, CRP 5.8 mg/L, ASOT 697 units/mL. Throat culture grew Group C beta-hemolytic Streptococci, universally susceptible to penicillin, ampicillin, amoxicillin, cefotaxime, ceftriaxone, and meropenem.

The patient was diagnosed with Rheumatic Fever, based on multiple Jones criteria: arthritis (major), subcutaneous nodules (major), possibly erythema marginatum (major), arthralgia (minor), fever (minor), elevated ESR and CRP (minor). He was treated with Penicillin G 600,000 units IM x1, and following an

uncomplicated recovery, will continue monthly Penicillin G for prophylaxis of recurrence. Streptococcal carriage may be high in ED [1], but rheumatic fever has not yet been described in this population. Rheumatic fever following Group C Strep has only been described in the Aboriginal Australian population [2]. Nodules are considered a rare physical finding in rheumatic fever, and when present are commonly associated with carditis, which was lacking in our patient.

REFERENCES

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