Atypical Early Onset Juvenile Sarcoidosis Presenting with Osteolytic Lesion

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Abstract
Sarcoidosis is a potentially fatal systemic inflammatory granulomatous disease, can occur in adult and pediatric patients, but it is relatively rare in children. Juvenile sarcoidosis has a diverse clinical course depending on the age of onset. Bone involvement is rarely noted in children with sarcoidosis and usually seen late in the course of the disease and is rarely the initial manifestation. Here we report a case of early onset juvenile sarcoidosis revealed by cutaneous and osseous lytic lesions of the phalanges.

Keywords: Atypical, Early onset, Juvenile, Sarcoidosis, Osteolytic lesions, Osseous sarcoidosis, Phalanges.

Case Report:
A 3-year-old girl, medically free, presented with a 9 months history of skin lesions at different body areas, persistent and progressed. The parents sought medical advice in many dermatology clinics, topical antibiotics and steroids were prescribed with no improvement. She had also swelling of both hands and feet with difficulty of movement early morning for few minutes. History of fever once at the beginning of her illness diagnosed with tonsillitis and treated with antibiotic. Good activity and appetite and no history of weight loss. Other systemic reviews were unremarkable. Family history was significant for multiple myeloma in her father. On examination, generally she looks well, normal growth parameters and vital signs. She had multiple scaly, erythematous to brown, indurated, discrete papules and plaques over the face, upper and lower extremities.

Also, she had 3*2 cm, erythematous, scaly nodule over the right thigh (Figure 1). There was a tender swelling of the phalanges of the fingers and toes bilaterally (Figure 2). The passive and active range of motion of all joints was normal. Cardiovascular, pulmonary, and abdominal examinations were normal. No lymphadenopathy was found.

Several laboratory investigations were performed, complete blood count showed picture of microcytic hypochromic anemia, mild elevation of sedimentation rate, and suboptimal vitamin D level.

Renal profile, liver profile, bone panel, C reactive protein, ANA, and rheumatoid factor all were normal. Chest radiography was normal and tuberculin skin test was negative. There was no elevation of the serum Angiotensin Converting Enzyme (ACE).

X-ray of the hands showed multiple bilateral expansile lytic lesions noted in the phalanges and both radiuses, some of them show aggressive feature such as periosteal reaction (Figure 3). Skin punch biopsy from the nodule over the right thigh showed intradermal well-circumscribed, noncaseating epitheloid granuloma with typical multinucleated giant cells and sparse peripheral lymphocytic infiltrates. Special stains and cultures are negative for pathogenic organism (Figure 4).

So, the diagnosis based on histologic evidence of noncaseating granulomas in the effected tissue and exclusion of other granulomatous diseases is cutaneous & osseous sarcoidosis.

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Bone survey revealed bilateral lytic lazy expansile lesions involving the short bone of [the phalanges, mid-carpal, distal radius and ulna and metatarsal bones with extensive periosteal reaction (Figure 5).

The patient started on prednisolone 15 mg PO once daily for 4 weeks. At follow up visit, there was decrease in the size of the subcutaneous nodule and decrease in the size of the osseous lesions clinically and radiologically (Figure 6).

Patient shifted to prednisolone tapered dose but unfortunately, she developed swelling of the phalanges again. Consultation with a rheumatologist suggested up-titration of the prednisolone and addition of methotrexate 7.5 mg PO once weekly with folic acid supplementation.

The patient was followed for 1 year. Her skin and bone lesions gradually decreased. There was no progression or involvement of other bones or new skin lesions. There has been no evidence of the systemic progression of disease.

**Figure 1:** Scaly, erythematous to brown, discrete plaques over the extremities. Also, she had 3*2 cm, erythematous, scaly nodule over the right thigh.

**Figure 2:** Swelling of the phalanges of the fingers and toes bilaterally.

**Figure 3:** X-ray of the hands showed multiple bilateral expansile lytic lesions noted in the phalanges and both radiuses, some of them show aggressive feature such as periosteal reaction.
Figure 4: Intradermal well-circumscribed, noncaseating epitheloid granuloma with typical multinucleated giant cells and sparse peripheral lymphocytic infiltrates. Special stains and cultures are negative for pathogenic organism.

Figure 5: Bone survey showed bilateral lytic expansile lesions involving the short bone of the phalanges, mid-carpal, distal radius and ulna and metatarsal bones with extensive periosteal reaction. These bone lesions are highly characteristic for sarcoidosis bone involvement.

Figure 6: After treatment with prednisolone there was decrease in the size of the osseous lesions clinically and radiologically. X-ray of the hands: before treatment (left side) and after treatment (right side).
**Discussion:**

Sarcoidosis is a potentially fatal systemic inflammatory granulomatous disease, can occur in adult and pediatric patients, but it is relatively rare in children. Cutaneous involvement may be more common in the pediatric population than in adults. The skin may be involved in about 24-40% of older children with sarcoidosis and in 77% of young children with sarcoidosis (1).

The most common specific eruptions are papular, consisting of red to yellow-brown or violaceous flat-topped papules, usually on the face. However, the entire spectrum of clinical cutaneous sarcoidosis has been described in children.

Two distinct forms of childhood sarcoidosis are noted. Older children usually present with a multisystem disease similar to the adult manifestation, with frequent lymphadenopathy and pulmonary involvement, as well as generalized signs and symptoms, such as fever and malaise. In contrast, early onset childhood sarcoidosis is a unique form of the disease characterized by the triad of rash, uveitis, and arthritis in presenting patients who are younger than age 4 years (2).

Osseous sarcoidosis has been reported in 3-13% of the adult cases. It occurs most often in patients known to have the chronic form of sarcoidosis with mult visceral manifestations (3). However, it can be the initial presentation of sarcoidosis in less than 1% of the cases. Bone involvement is rarely noted in children with sarcoidosis and no true percentage because of the rarity and the small number of reported cases.

Most frequent locations are the phalanges of the fingers and toes. Sarcoid lesions have also been reported to occur in the long bones, spine, and skull. Osteolytic bone lesions or erosive changes are rarely observed in association with musculoskeletal manifestations of childhood sarcoidosis and often associated with cutaneous and long-standing chronic multisystem disease (4).

Our patient is a 3-year-old girl presented with skin rash, subcutaneous nodule, and lytic and cystic bone lesions. There were no pulmonary clinical findings, no joint, eyes, or cardiac involvement. Although the characteristic triad of early onset juvenile sarcoidosis not present in this case, the diagnosis of cutaneous sarcoidosis was confirmed based on the histologic evidence of noncaseating granulomas in the affected skin and exclusion of other granulomatous diseases.

The lack of the classical triad of early onset juvenile sarcoidosis, which delayed the diagnosis several months, and the presence of osseous sarcoidosis early in the course of the disease contribute to the rarity of the case.

**Conclusion:**

Sarcoidosis in childhood is recognized as a systemic disease affecting various organs and having diverse clinical course depending on the age of onset. Early onset juvenile sarcoidosis presents usually with the classical triad of rash, uveitis, and arthritis. Although it is rare, osteolytic bone lesions must be considered as a manifestation of early onset juvenile sarcoidosis.

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**References:**