Childhood Sarcoidosis: An Atypical Case Report

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Sir,

Sarcoidosis is a multisystem granulomatous disorder of unknown etiology, mostly encountered in adults aged 20–40 years.[1] Childhood sarcoidosis is a rare entity with diverse clinical presentations and most reports in children aged 13–15 years.[2] The case described by us assumes clinical significance because of presentation in childhood and atypical clinical presentation, hence necessitating a high index of suspicion to arrive at the diagnosis.

A 12-year-old male patient, known case of cerebral palsy, presented with brown- to red-colored asymptomatic raised skin lesions on dorsa of both hands and ankles for the last 4 months [Figures 1-3]. The parents gave a history of recurrent eye disease with pain, blurring of vision, and photophobia since the age of 6 years. Case records revealed that the child was a known case of therapy resistant and recurrent anterior uveitis. There was no associated history of joint pain, dyspnea, chest pain, fever, or any other constitutional features.

Examination revealed two well-defined erythematous brown-colored slightly atrophic plaques (3.2 cm × 2.1 cm and 2.4 cm × 1.5 cm) without any induration or tenderness on dorsa of both hands. Medial aspect of bilateral ankle region revealed multiple ill-defined discrete violaceous papules coalescing to form a plaque with few atrophic areas in adjoining skin. Slit lamp ophthalmic examination revealed subacute granulomatous uveitis with interstitial keratopathy.

Tuberculin test was weakly reactive, erythrocyte sedimentation rate (ESR) (48 mm/h), and serum angiotensin-converting enzyme (ACE) levels were significantly raised (140 µL; normal range 8–65 µL).

Hematoxylin and eosin-stained section from lesion on right hand revealed mild perivascular lymphomononuclear infiltrate in the upper dermis with mild atrophy of epidermis. Deep dermis revealed naked epithelioid cell granulomas and absence of Langerhans giant cells with minimal areas of necrosis [Figure 4], which confirmed the diagnosis of sarcoidosis.

Childhood sarcoidosis presents in two distinct patterns, children younger than 5 years of age tend to have skin, ophthalmic and joint disease without significant pulmonary involvement, whereas older children have...
a multisystem disease pattern similar to adults with predominant lung and hilar lymph node involvement in addition to eyes, bones, and skin.\[2\]

Cutaneous sarcoidosis may present in any of the myriad specific forms; papular, plaque, nodular, macular, scar, angiopuloid or the rarer ones such as ichthyosiform, atrophic, lichenoid, erythodermic, verrucous, psoriasiform, or morpheaform.\[1\] Nonspecific lesions such as erythema nodosum, sweet’s disease, or pyoderma gangrenosum may occur alone or coexist with specific lesions of sarcoidosis.

Conjunctival or choroidal granulomas and peripheral multifocal choroiditis are considered as specific markers of ocular sarcoidosis; however, it is anterior uveitis or iritis which occurs more frequently and leads to considerable morbidity. Pulmonary affliction, though seen in approximately 80% of adult patients, is not frequently encountered in pediatric sarcoidosis. Bilateral asymptomatic hilar lymphadenopathy may be the only incidental finding. Joint involvement in childhood sarcoidosis presents as tenosynovitis with painless effusion or painful arthritis with limitation of movement.\[2\] Cardiac, renal, and central nervous system involvement are considerably rare in pediatric age group.\[2\] Sarcoïdosis in children <5 years may present as Blau syndrome (a triad of uveitis, arthritis, and asymptomatic papules).\[3\] The diagnosis of sarcoïdosis rests primarily on demonstration of noncaseating epithelioid granulomas with or without limited amount of fibrinoid necrosis on histopathological examination of skin or other affected organs.\[4\]

Elevated serum ACE levels and ESR are useful indicators of disease activity. Chest radiograph and electrocardiogram must be done in all cases along with ophthalmic slit lamp examination.\[4\] The prognosis and natural history of childhood sarcoïdosis are not clearly described due to rarity of disease and paucity of literature. Hence, we recommend that childhood sarcoïdosis be considered among the differential diagnosis of atypical cutaneous lesions even if morphology is not otherwise suggestive of sarcoïdosis. Not surprisingly, it has been said in context of sarcoïdosis that “one of its most singular details is the frequency of its clinical silence.”\[5\]

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**Conflicts of interest**
There are no conflicts of interest.

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