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Subcutaneous Granuloma Annulare of the Scalp in Childhood: A Case Report and Review of the Literature

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ABSTRACT

Granuloma annulare is a benign inflammatory skin lesion of unknown etiology that is usually seen in adults and children and subtypes of it includes localized granuloma annulare, generalized granuloma annulare, subcutaneous granuloma annulare and arcuate dermal erythema. Etiology and pathogenesis of granuloma annulare are obscure, although there is much evidence for an immunologic mechanism. Precipitating factors are insect bites, sunburn, photochemotherapy, drugs, physical trauma, acute phlebitis and sepsis after surgery. Some investigators were suggested a relationship of granuloma annulare to a latent or clinically manifest diabetes or rheumatoid arthritis. In contrast, an association of subcutaneous granuloma annulare with these diseases in childhood has not been reported in the literature. Subcutaneous granuloma annulare of the scalp is rare lesion in childhood and nodules on the scalp are usually non-, or slightly mobile, whereas lesions on the extremities are freely mobile. For definitive diagnosis, a biopsy should be performed but wide surgical intervention or medical treatment is not indicated. In case of recurrence, no additional diagnostic studies are necessary.

KEY WORDS: Childhood, Scalp, Subcutaneous granuloma.

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INTRODUCTION

Granuloma annulare (GA) is a common, self-limited, benign inflammatory skin lesion of unknown etiology that is usually seen in both adults and children. Subtypes with distinct clinical features include localized granuloma annulare (LGA), generalized granuloma annulare (GGA), subcutaneous granuloma annulare (SGA) and arcuate dermal erythema (3,4). Different authors have referred to the same condition as benign rheumatoid nodule, pseudorheumatoid nodule, isolated subcutaneous nodule, subcutaneous palisading granuloma and palisading granuloma nodosum (2). Although SGA is seen exclusively in children, most cases are reported in surgical, pathology or dermatologic journals with little information in the general pediatric literature (3).

CASE REPORT

A 2-year-old girl was admitted to our hospital with three subcutaneous scalp masses. Two of these were in the right occipitoparietal area and 6 and 5 mm in diameter respectively and one was in the occipital area and 4 mm in diameter. In history, we learned that these masses were appeared 15 days before without any trauma and grew up in days. The lesions were immobile, nontender and no

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other dermatologic lesion was observed over the skin. No abnormality was determined during the neurological examination and the remainder of the physical examination. Laboratory tests revealed a normal complete blood count, antistreptolysin O, erythrocyte sedimentation rate, rheumatoid factor, blood glucose level and anti-nuclear antibody level. Cranial computed tomography with intravenous administration of contrast substance showed 2 right occipitoparietal and one occipital subcutaneous masses without any bone destruction or hyperosteosis (Figure 1). The masses were totally excised to prevent an increase in size. They were hard, involved the galea, tight on the periosteum and immobile. Histologically, the lesion was composed of palisading granulomas in the subcutaneous tissue. The granulomas were characterized by a well-demarcated zone of disintegrating extracellular material mixed with cell debris surrounded by a cuff of radially oriented fibroblasts varying with lymphocytes and histiocytes (Figure 2). The centrally located material had the appearance of collagen and elastic fibers in trichrome staining (Figures 3). Mucin was present in the areas of altered collagen. Occasional foreign body giant cells are present. Immunohistochemically these histiocytes were characterized by Mac 387 and CD68 expression. A diagnosis of palisading granuloma was confirmed. No recurrences or new masses were determined in the scalp subcutaneous tissue of this patient during two-year follow up.

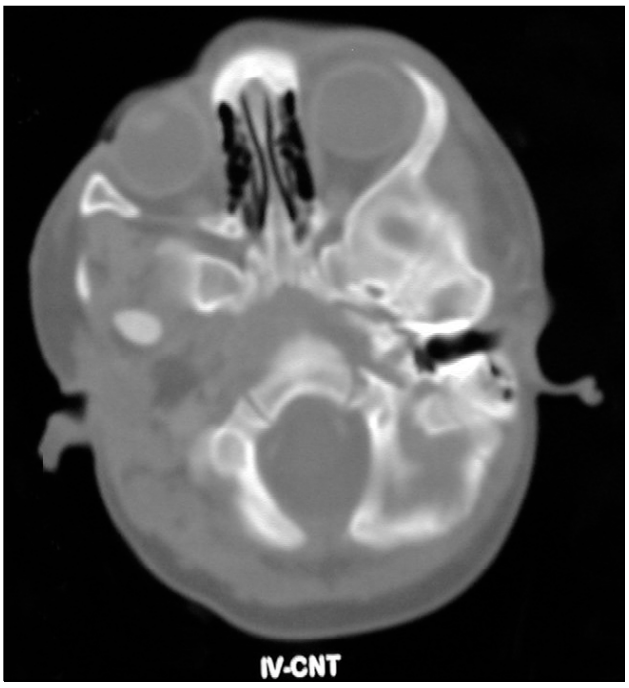

Figure 1: On cranial computed tomography with intravenous 1 administraton of cnotrast substance, right occipitoparietal and one occipital subcutaneous masses were seen without any bone destruction and hyperosteosis.

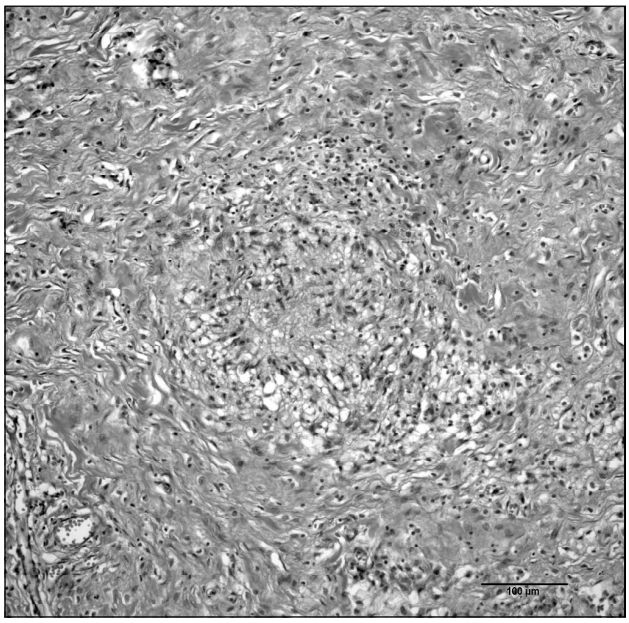

Figure 2: Typical granuloma with palisading histiocytes around necrobiotic collagen center (HE, x 20).

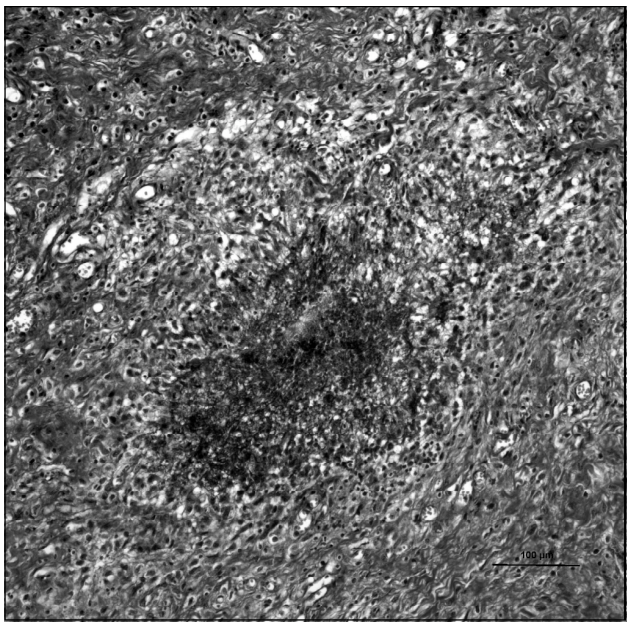

Figure 3: Typical granuloma with palisading histiocytes around necrobiotic collagen center (Trichrome staining, x 20).

DISCUSSION

GA was first described by Fox in 1895. Ziegler, in 1941, was the first to note the histological similarity of the isolated nodule seen in GA to the nodules seen in rheumatoid arthritis (3). The etiology and pathogenesis of GA are obscure, although there is much evidence that an immunologic mechanism, probably a delayed-type hypersensitivity, is involved (10). Immunologic studies of lymphocytes subsets and serum monocyte-modulating factors support the hypothesis that GA is the result of an aberrant cell-mediated immune response (2). More recently, vasculitis and cellular immunity have been hypothesized to play a role in GA.

Regarding the etiology, there is no good evidence for putative causes of SGA including tuberculosis, trauma, streptococcal infection, thyroid disease, Epstein-Barr virus, herpes zoster/varicella or collagen disorder. The majority of cases were reported in the first three decades of life (89% in Muhlemann and Williams' study, 79% in Andersen and Verdich's study, 62% in Studer's study) (10) . Reported precipitating factors of GA are insect bites, sunburn, Psoralen UV-A (PUVA), drugs, physical trauma, acute phlebitis and sepsis after surgery (1,10). Although there is no relationship between emotional stress and SGA, Studer mentioned that 15% of their patients reported stress situations as a trigger of recurrences.

The study and treatment of patients with SGA of all types generates controversy and confusion because widely variant clinical syndromes can be seen, the histology mimics other disorders and effective therapy remains to be undetermined (3). Adults presenting with rheumatoid nodules, histologically identical to SGA, are at significant risk for the development of rheumatoid arthritis or other connective tissue disease (2). Because of this association, efforts have been made to determine whether children with these lesions are at similar risk. Draheim et al (4) collected 54 cases at the Armed Forces Institute of Pathology in 1959; in no case did connective tissue disease develop after 1 to 14 years follow-up. Subsequent reports with smaller numbers of cases have also concluded that there is no increased risk for systemic rheumatologic disease in the children (3).

Although the association of SGA with diabetes mellitus is not yet established (4), several investigators have attempted to demonstrate a relationship of SGA with other systemic diseases, most notably with diabetes mellitus (4,7,9,10). The reason for searching for a relationship between diabetes mellitus and SGA is similar histopathological features with necrobiosis lipoidica diabeticorum, a well-recognized dermatologic complication of diabetes mellitus. In contrast, an association of SGA with diabetes mellitus in childhood has not been published in the medical literature (5).

Various diseases have been reported to involve the subcutaneous portion of the scalp in childhood. These include osteoma, eosinophilic granuloma, cranial fasciitis, palisading granuloma, sarcoidosis, fibrous dysplasia, dermoid tumor, traumatic granuloma, abscess, hemangioma, sinus pericranii and primary sarcoma (5). The differential diagnosis of subcutaneous nodules of the scalp and/or extremities is extensive and includes trauma, infection, tumor, metabolic bone or skin disease and inflammatory/autoimmune disease. All of the above can be ruled out with a proper history, focusing on issues regarding diet, weight loss and/or fever (3).

SGA most often manifests as a large, asymptomatic soft tissue mass. Although nodules are usually stable for months, they may rapidly enlarge over the course of weeks. The typical lesions of SGA are single or multiple, small, pinkish, nonulcerated nodules in the deep subcutaneous tissue. As shown in current studies, the most common lesion location is lower extremity, especially the pretibial area, followed by the hands (4). The buttocks, forehead and scalp are less commonly affected. Deep dermal or subcutaneous nodules on the extremities are attached to fascia and are often therefore mobile, whereas lesions on the scalp are attached to underlying periosteum and are therefore fixed or only slightly mobile. SGA of the scalp is a rare lesion in childhood and presents with immobile, nontender masses that are multiple and tend to vary in size over time (5). On histological evaluation, these nodules are similar to the nodules seen in adults with rheumatoid arthritis and to the lesion recognized in adult diabetic patients as necrobiosis lipoidica diabeticorum (4). The typical histology shows that the focus of the lesion has a central core of incomplete and mostly reversible change (necrobiosis) of connective tissue, surrounded by a wall of palisaded histiocytes. The necrobiotic centers are usually oval in outline,

slightly basophilic, devoid of nuclei and marked by a loss of outline of collagenous and elastic tissue fibers. Characteristically, the necrobiotic focus is not so large or deeply situated as those of rheumatoid nodules or as broad and diffuse as those of necrobiosis lipoidica (diabeticorum). If an accurate history is not available, simple laboratory tests including a complete blood count (CBC), plain radiograph and erythrocyte sedimentation rate (ESR) may be obtained for laboratory investigation. Further serologic studies such as antinuclear antibody, rheumatoid factor, complement and C-reactive protein are uniformly negative and probably not warranted (2). In case of benign history and normal examination except the nodules, no additional work-up is necessary (3). The presence or absence of calvarial invasion is an important consideration in the diagnosis: Plain radiographs show a soft tissue mass without bony involvement, calcification, or ossification. Cranial computerized tomography is useful for this assessment and helps rule out calvarial invasion. The possible value of magnetic resonance imaging findings in differentiating between types of subcutaneous masses has not been sufficiently clarified.

Although the clinical profile is characteristic, the definite diagnosis is based upon biopsy study and excisional biopsy should provide an adequate specimen in all cases. Therapy for these lesions has also varied. Most have been treated by excisional biopsy. Unfortunately, some patients have unnecessarily been treated with wide local excision that has required subsequent skin grafting (7). Since multiple lesions develop in many of these children, there has been a tendency to surgically excise them as they recur. However, the knowledge in the literature supports the fact that these lesions will regress over time if left alone. Medical therapy in the form of steroid injections, antibiotics, potassium iodine, dapsone, niacinamide, chlorambucil, isoretionin, retinoids (vitamin A derivates), PUVA and aspirin has also been attempted and found to be of no value (3,8).

The prognosis for children with these lesions is excellent. Although subsequent additional lesions will develop in some, they are self-limiting. A fine needle aspiration of a new lesion may serve to reassure the physician that the lesion is indeed benign. In a child with a negative history, otherwise normal physical examination and normal ESR, these appears to be no increased risk for the development of any systemic collagen vascular disease over that of the general population (8). Spontaneous resolution of SGA has occurred within 2 years in 50% of cases, although lesions may last weeks to decades. Recurrence, often at the same site, is noted 40% of cases.

In summary, no additional studies are necessary if the history and physical examination correlate with SGA. A biopsy should be performed for a definite diagnosis but wide surgical excision or medical treatment is not indicated. Recurrences locally or at other sites are common but usually do not need additional biopsy procedures (11).

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