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# ASYMMETRIC PERIFLEXURAL EXANTHEM OF CHILDHOOD. A CLINICO - EPIDEMIOLOGICAL PROSPECTIVE STUDY OF SEVENTY-FIVE CASES

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### **ABSTRACT**

Introduction: Asymmetric periflexural exanthem of childhood is classified as a rare self-limited and spontaneously resolving exanthem with unknown etiology that occurs mainly in children. **Objective:** To study the prevalence and clinical features of asymmetric periflexural exanthem of childhood in Iraqi patients. **Patients and Methods:** patients presenting with features of asymmetric periflexural exanthem of childhood were studied during the period from 2012 to 2015 in the department of dermatology and venereology of Al-Ramadi Teaching Hospital, a detailed history was taken and careful physical examination of patients were carried out, regarding skin and other systems. **Results:** Seventy-five patients with asymmetric periflexural exanthem of childhood, 40 males and 35 females were studied, with male to female ratio 1.1/1, their ages ranged from 17 days to 5 years with an average of 1.8 years, most patients[54%] were diagnosed between May and July with a peak in May[34.6%]. The prodromal symptoms were noted in 56% of cases, mostly of upper respiratory tract infections, the eruption consisted of erythematous papules which coalesced into eczematous plaques begin unilaterally mainly around knee region in 60%, Pruritus was noted in 65% of patients, lymphadenopathy present in [40%] and were mainly inguinal lymph nodes on the side affected initially. **Conclusion:** Asymmetric periflexural exanthem of childhood is a common disease affecting children mainly with a self-limited course, the lower extremities were the main affected sites in our locality.

KEYWORDS: Exanthem, Childhood, Fever.

## INTRODUCTION

In 1962, Brunner et al. described an eruption as lichen miliaris, which had unilateral, periflexural involvement, in 1992, the term Unilateral Laterothoracic Exanthem (ULE), was proposed by Bodemer and De prost, in 1993, Taieb et al. reported 21 cases with a similar eruption, and because the eruption did not always remain unilateral and could involve the lower extremities, they suggested the name Asymmetric Periflexural Exanthem of Childhood (APEC) instead. [1]

Asymmetric Periflexural Exanthem of Childhood(APEC) is of unknown etiology, but the patient's history (age at presentation and multiple affected children in a family), lack of efficacy of broad spectrum antibiotics treatment, the tendency for seasonal presentation during spring and winter and the presence of prodromal symptoms raise the possibility of viral etiology. [1,2,3,4] It's occurrence in association with immunodeficiency, such as during chemotherapy for leukemia and in immunocompromised hosts are also suggestive of infectious process. [5,6] The viruses that have been implicated are parainfluenza 2 and 3, adenovirus and parvovirus B19. [7,8] APEC is a clinical

syndrome that tend to be seasonal, occurs primarily in the late winter and early spring and appear to be most common in Europe. [9] It affects girls more often than boys with a female to male ratio about 2:1, occurs in children at age of eight month to ten years, mostly between two and three years. [10] Multiple cases have been reported in adults. [11,12,13]

About 60% of affected children had a preceding prodromal symptoms of mild upper respiratory system or gastrointestinal system including rhinitis, pharyngitis, otitis media, fever, vomiting and diarrhea and often with low-grade fever. [4,8,10]

Clinically the lesions are usually discrete about 1-4mm erythematous papules with pale halo that may be noted around some large lesions, these papules usually coalesce to form a poorly marginated plaques, or sometimes the disease present with morbilliform, scarlatiniform or eczematous eruptions. [2.8,12] The lesions begin unilaterally close to a flexural area, usually the axilla (75%) of cases, less often they begin in the groin

region and sometimes appear on an extremity, not on the trunk.  $^{[10,14,15]}$ 

Spread of the lesions is centrifugal with new lesions appearing on the adjacent trunk and proximal extremity, normal skin may intervenes between lesions, the contralateral side is involved in 70% of cases after 5-15 days but the asymmetrical nature is maintained throughout the illness and there is no right or left predominance. [7,8,14]

Pruritus is present in about 50-65% of cases, it is mild and there is no lichenification. [8,14] There are Lymphadenopathy usually confined to the areas of initially affected side in about 50-70% of cases and the general health of the children are not affected. [4,10,14] In contrast to other exanthems, APEC rarely involves the face, and sparing of the palms, soles and mucous membranes is typical. [7,8] The syndrome last 2-6 weeks on average, but may last more than two months and resolve spontaneously without sequels, [10,14,16] and the treatment is symptomatic only. [17,18]

APEC is commonly misdiagnosed as allergic contact dermatitis, non-specific viral exanthems,drug-related eruptions, miliaria, superficial fungal infections, scabies, lichen striatus, Gianotti-Crosti syndrome and atypical pityriasis rosea. [17] Its diagnosis is primarily clinical, and laboratory investigations are generally not required. [17,18] Histopathological changes of the lesion are generally non-specific, although, perisudoral lymphocytic infiltrates have been reported to be a fairly specific features for APEC. [5] There may be an accompanying interface dermatitis of the upper eccrine duct and adjacent epidermis. [10]

## PATIENTS AND METHODS

After obtaining a clearance for the study from the institutional ethics committee and with written informed consent of each patient's parents, all patients who attended the department of dermatology and venereology of Al-Ramadi Teaching Hospital during the three years study period from January 1,2012 to January 1, 2015 were carefully evaluated. preset proforma was used to collect data: regarding age, sex, time of the year, presence of any prodromal symptoms [ear, nose, throat, digestive or others ], family history of the same illness. Characteristic of the rash [date of onset, initial topography, description of the elementary lesion, pruritus, pattern of spread and duration of the eruption]. Presence of fever, enanthem, lymphadenopathy, splenomegaly and hepatomegaly were evaluated.

## RESULTS

During the study period, 75 patients who attended the department of dermatology and venereology of Al-Ramadi Teaching Hospital were diagnosed as having APEC, of these, 40 were males and 35 were females with male to female ratio 1.1/1, the ages of the patients ranged

from 17 days to 5 years with an average of 1.8 years, most patients [41/75:54%] were diagnosed between May and July with a peak in May [34.6% of cases], no concomitant or subsequent case of APEC or other rash or infectious disease was noted among relatives or contacts.

The prodromal symptoms were noted in 56% of cases, most often upper respiratory tract infections in form of rhinitis, otitis media, pharyngitis, cough, bronchitis with fever.

Most often the eruption consisted of erythematous micropapules which coalesced into poorly marginated eczematous plaques begin unilaterally close to a flexural area mainly around knee region in 60% of cases "Fig. 1:A and B" and" Fig. 2".



Figure 1 (A): Shows multiple micropapules affecting right knee region, both thigh and lower legs but right side was more affected in a child with APEC.



Figure 1 (B): Shows multiple micropapules affecting right knee region of the same child with APEC in Figure 1 (A).



Figure 2: Other affected patient with APEC with involvement of the right buttock and lower thigh by multiple papules.

Wrist was affected in 12%, elbow and ankle, each affected in 8%, axilla 5.4%, groin in 4% and others in 2.6% of cases "Fig. 3".

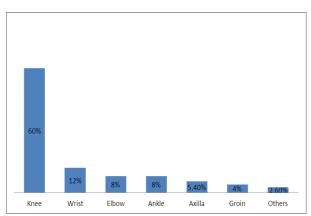


Figure 3: Shows areas of the body involved by APEC.

Eruption spread centrifugally and although both sides of the body were eventually involved, almost all had unilateral predominance, the lesions begins on the right side in 60% of cases while the left side first affected in 40% of cases. "Fig. 4".

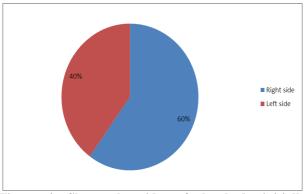


Figure 4: Shows the sides of the body initially involved by the eruption of APEC.

Pruritus was noted in 65% of patients mostly with mild intensity without lichenification, there is no involvement of face, palm, sole and mucus membrane.

Lymphadenopathy were noted in [40%] and were mainly inguinal lymph nodes on the side affected initially, they were firm and non-tender, and there was no splenomegaly or hepatomegaly.

## **DISCUSSION**

Viral exanthems are mostly associated with self-limited diseases, however, in some cases diagnosis of an exanthem may be crucial to patients and their contacts. APEC is classified as a rare disease that occur mainly in children and mostly in Europe, since 75 cases of APEC were collected during three years period, we consider it a common disease in our area, although the etiology of APEC is unknown, but the ages of patients at presentation, seasonal tendency and the presence of prodromal symptoms raise the possibility of viral etiology. [1.2,3,4]

In additions to these features in evaluated patients in whom males and females were nearly equally affected in favor of viral infections which affect both sex equally in contrast to previous studies in which female to male ratio was 2:1.<sup>[10]</sup>

About 60% of affected children had a preceding prodromal symptoms of upper respiratory and gastrointestinal infections, [8] these symptoms were complained in 56% of cases which also in favor of viral etiology.

Clinically the lesions present as discrete, erythematous papules usually coalesce to form poorly marginated plaques which begins unilaterally close to a flexural area usually the axilla in 75% of cases <sup>[14,15]</sup>,but in the evaluated patients, the knee region was the main affected site (60%) of cases while the axilla was involved in (5.4%) of cases, this may be due to geographical distribution of the disease and seasonal variation where most previous cases were reported from Europe, <sup>[9]</sup> while in the present study, the evaluated patients from Asia.

The eruptions spread centrifugally with both side affected, but almost all had unilateral predominance, the right side involved in 60% while the left side affected in 40% of cases, the cause for this preferential unilateral and asymmetrical affection by the rash is unexplained, but the new concept that an early post zygotic mutation has rendered the skin on one side of the body more reactive to infective agents, the subsequent bilateral involvement would reflect the less sever reactivity of the contralateral keratinocytes. <sup>[14,16]</sup>

Lymph nodes involvement of the initially affected side [50-70%], mainly axillary lymph nodes<sup>[10,14]</sup> while in the evaluated patients, the inguinal lymph nodes involved [40%] because the most affected side was the knee region [60%].

### CONCLUSIONS

APEC is a common disease affecting children mainly with a self-limited course, the lower extremities were the main affected sites in our locality. The importance of early recognition of this exanthem is that it would prevent the exhaustive investigations work up that is routinely performed to establish the diagnosis in a typical rash and to avoid overuse of medications especially steroids in spontaneously resolving disease.

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