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Dermatologic diseases presenting with pigmentation disorders in children: a single center experience
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Abstract:
Background: To determine the incidence and demographic characteristics of skin diseases presenting with hyperpigmentation in children applying to the dermatology department.
Methods: A total of 2815 children between the ages of 0 to 16 who applied to the dermatology clinic with the complaints of hyperpigmentation disorders were evaluated. The age, gender, socioeconomic status, place of residence and demographic characteristics of children with abnormal pigmentation skin lesions were investigated.
Results: A total of 2815 children applied to the dermatology clinic during the study period. Of these patients 1491 were female (53%) and 1324 were male (47%). Of these 266 (9.4%) were diagnosed with skin disorders presenting with hyperpigmentation. The causes of hyperpigmentation in these patients were as follows; pityriasis alba (2.6%), melanocytic nevus (2.1%), vitiligo (1.8%) postinflammatory hyperpigmentation (0.5%), and postinflammatory hypopigmentation (0.5%). According to the gender, p.alba, vitiligo, Becker nevus, acanthosis nigricans, tuberosclerosis and albinism were seen more in boys and nevus depigmentosus ephelis, postinflammatory hyperpigmentation/hypopigmentation and melanocytic nevus were seen more in the girls. Pityriasis versicolor was seen equally in both genders. According to age groups, melanocytic nevus were found to be more frequent between the ages of 0-2 and 12-16, whereas pityriasis versicolor was more frequent in ages 12-16 and P. alba in the 3-11 age group.
Conclusions: There are quite a substantial number of pigmentation diseases occurring in children. Early diagnosis and treatment are important because although these diseases mainly cause cosmetic problems, they can decrease the quality of life. Here, we attempted to define the demographic characteristics of diseases presenting with disorders in pigmentation in children.
Keywords: Melanocytic lesions, pigmentation disorder, pediatric dermatological diseases, demographic characteristics
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Introduction

Pigmentation disorders hold an important place in dermatological diseases and their frequency varies according to the population. They can cause both visual and psychological problems, especially in children [1]. Pigmentation disorders can either be due to various congenital or acquired diseases. Familial frequency, the onset
and course of the lesions, drug use and exposure to chemicals should be questioned and any presence of systemic findings should be investigated. The main diseases that present with disorders of pigmentation are pityriasis alba, melanocytic nevus, vitiligo, pityriasis versicolor, postinflammatory hyperpigmentation/hypopigmentation, nevus depigmentosus, ephelis, café au lait macules, Becker nevus, acanthosis nigricans, tuberosclerosis and albinism [2-4].

There are a limited number of researches dealing with these disorders in the pediatric age group. In this study, we aimed to evaluate the frequency, distribution and demographic characteristics of pigmentation diseases in children who had applied to a single center.

Material and Methods

2815 children between the ages of 0 and 16 who had applied to the Ankara Pediatric Health and Diseases Hematology Oncology Training and Research Hospital were evaluated retrospectively. Permission was obtained from the ethic committee. The relatives of the children were informed about the study and signed consent forms were obtained. All patients were examined by two specialist dermatologists. To compare the distribution according to the demographic data and diagnoses, patients were investigated in 4 categories: age 0-2 (infantile period), age 3-5 (pre-school period), age 6-11 (school period) and age 12-16 (adolescent period). The diagnosed skin diseases were evaluated according to age, gender, socioeconomic status, place of residence, family history and lesion location. The socioeconomic status variables questioned included family income, health insurance, living conditions, educational status of parents and number of siblings.

The SPSS version 20 program was used for statistical analysis and the Fisher test was used for the comparison of categorical variables. For the determination of statistical significance the Chi-Square test was used. The limit of statistical significance was determined as p <0.05. With suspicious diagnoses, diagnostic tools including the Wood light exam, dermatoscope evaluation, mycotic search and skin biopsy were utilized.

Results

Of the 2815 patients, 266 (9.4%) were determined to have skin diseases presenting with pigmentation disorders. The distribution of diseases according to gender is shown in the Table 1.

When the diseases were examined according to gender, P. alba, vitiligo, Becker nevus, tuberous sclerosis, albinism and acanthosis nigricans were seen more seen in boys, and melanocytic nevus, nevus depigmentosus, ephelis, and postinflammatory hyperpigmentation and hypopigmentation were seen more in girls (Table 1). The increase in incidence of P. alba with increased age and decreased socioeconomic status in boys was found to be statistically significant (p<0.05). An increased incidence of vitiligo in older children was found to be statistically significant (p<0.05), but the gender of this condition was not found to be significant. The increase in incidence of melanocytic nevus with increases age in girls was found to be statistically significant (p<0.05). On evaluation according to age group, melanocytic nevus and vitiligo were more frequent in the age groups 0-2 and 12-16, pityriasis versicolor in ages 12-16 and p.alba in ages 3-11 (Table 2).

The pigmentation disorders seen most frequently in decreasing order are as follows; p. alba (2.6%), melanocytic nevus (2.1%), vitiligo (1.8%), pityriasis versicolor (0.8%), postinflammatory hyperpigmentation (0.5%), postinflammatory hypopigmentation (0.05%), nevus depigmentosus (0.3%), ephelis (0.2%), Becker nevus (0.2%) and café au lait (0.07%),
acanthosis nigricans (0.07%), tuberous sclerosis (0.07%) and albinism (0.03%). Vitiligo was found to be more common in middle class patients living in urban environments. While the incidence rate in first degree relatives was 11.5%, it was found to be 5.7% in second degree relatives. P.alba was seen in 6.8% of first degree relatives. P.alba was seen more in patients with lower socioeconomic status. Melanocytic nevi were seen more in low income urban residing children and this increase in condition with decreased socioeconomic status was found to be statistically significant (p<0.05). Postinflammatory hypopigmentation was seen more in the middle class living in low income districts. Postinflammatory hyperpigmentation, ephelis and café au lait macules were more common in the middle class living in an urban environment.

Table 1. Distribution of pigmentation disorders according to gender.

<table>
<thead>
<tr>
<th>Patient numbers</th>
<th>Disease</th>
<th>Female</th>
<th>%</th>
<th>Male</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>73</td>
<td>P.alba</td>
<td>21</td>
<td>28</td>
<td>52</td>
<td>72</td>
</tr>
<tr>
<td>58</td>
<td>Melanosis nevus</td>
<td>31</td>
<td>53</td>
<td>27</td>
<td>46</td>
</tr>
<tr>
<td>52</td>
<td>Vitiligo</td>
<td>25</td>
<td>48</td>
<td>27</td>
<td>52</td>
</tr>
<tr>
<td>22</td>
<td>Pytriasis versicolor</td>
<td>11</td>
<td>50</td>
<td>11</td>
<td>50</td>
</tr>
<tr>
<td>15</td>
<td>P.hiperpigmentation</td>
<td>9</td>
<td>60</td>
<td>6</td>
<td>40</td>
</tr>
<tr>
<td>15</td>
<td>P.hipopigmentation</td>
<td>9</td>
<td>60</td>
<td>6</td>
<td>40</td>
</tr>
<tr>
<td>11</td>
<td>N.depigmentosus</td>
<td>6</td>
<td>54</td>
<td>5</td>
<td>45</td>
</tr>
<tr>
<td>6</td>
<td>Becker nevus</td>
<td>2</td>
<td>33</td>
<td>4</td>
<td>67</td>
</tr>
<tr>
<td>7</td>
<td>Ephelid</td>
<td>5</td>
<td>71</td>
<td>2</td>
<td>29</td>
</tr>
<tr>
<td>2</td>
<td>Cafe au lait</td>
<td>1</td>
<td>50</td>
<td>1</td>
<td>50</td>
</tr>
<tr>
<td>2</td>
<td>Tuberosclerosus</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>100</td>
</tr>
<tr>
<td>2</td>
<td>Akantosis nigricans</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>100</td>
</tr>
<tr>
<td>1</td>
<td>Albinism</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>100</td>
</tr>
<tr>
<td>Total 266</td>
<td></td>
<td>120</td>
<td>100</td>
<td>146</td>
<td>100</td>
</tr>
</tbody>
</table>

Discussion

Pigmentation disorders have an important place in dermatological diseases. The prevalence of these disorders varies from country to country [1-4]. There are very few studies on this subject in our country. In the studies that have been done, rates have been found to range between 3.6% and 9.9%. In our retrospective study, we found the rate of pigmentation disorders in dermatological diseases to be 9.4% of the 2815 children that applied to our clinic.

P.alba is a hypomelanosis most commonly seen at equal gender rates in atopic children and teens. It is believed to be an eczematous dermatitis and a postinflammatory hypomelanosis [5-7]. It commonly affects the face but can also be seen on the neck, trunk, back, extremities and scrotum. It shows a tendency to recover with increased age. It can be widespread and of long duration in atopic cases.8 In our country, it is seen at a rate of varying from 1.6% and 4.9% [7,9]. Inanır and friends reported the incidence of
p.alba as being 12% in a school screening of 785 school children in a low socioeconomic district[7]. It has been reported at an incidence of 5.2% in Kuwait, 2.9%-3.7% in Switzerland, 1% in Hong-Kong and 5.8% in India. In our study, the incidence of P.alba was found to be 2.6%, and was seen more in boys. The reasons for this low rate may be because there are no screening programs in nurseries and schools, our population does not pay too much attention to this disorder or the fact that this diagnosis is made at level 1 and 2 hospitals. P.alba has been found to be more common in cases with low socioeconomic status.

Table 2. Pigmentation disorder distribution according to age.

<table>
<thead>
<tr>
<th>Patient</th>
<th>General (%)</th>
<th>Within Group (%)</th>
<th>Age 0-2 n=670</th>
<th>Age 3-5 n=510</th>
<th>Age 6-11 n=913</th>
<th>Age12-16 n=722</th>
</tr>
</thead>
<tbody>
<tr>
<td>P. Alba</td>
<td>73</td>
<td>27.4</td>
<td>4 11.1</td>
<td>18 35</td>
<td>39 39.7</td>
<td>12 14.2</td>
</tr>
<tr>
<td>M. nevus</td>
<td>58</td>
<td>21.8</td>
<td>9 25</td>
<td>12 23</td>
<td>18 18.3</td>
<td>19 23.4</td>
</tr>
<tr>
<td>Vitiligo</td>
<td>52</td>
<td>19.5</td>
<td>8 22</td>
<td>10 19.6</td>
<td>16 18</td>
<td>18 22.2</td>
</tr>
<tr>
<td>P. versicolor</td>
<td>22</td>
<td>8.3</td>
<td>1 2.7</td>
<td>3 5.8</td>
<td>4 4</td>
<td>14 17.2</td>
</tr>
<tr>
<td>P. hiperpigment</td>
<td>15</td>
<td>5.6</td>
<td>3 8.3</td>
<td>2 3.9</td>
<td>5 5</td>
<td>5 6.1</td>
</tr>
<tr>
<td>P. hipopigment</td>
<td>15</td>
<td>5.6</td>
<td>4 11.1</td>
<td>2 3.9</td>
<td>7 7.1</td>
<td>2 2.4</td>
</tr>
<tr>
<td>N. depigmentosus</td>
<td>11</td>
<td>4.1</td>
<td>4 11.1</td>
<td>3 5.8</td>
<td>3 3</td>
<td>1 1.2</td>
</tr>
<tr>
<td>Ephelid</td>
<td>7</td>
<td>2.6</td>
<td>0 0</td>
<td>0 0</td>
<td>3 3</td>
<td>4 4.9</td>
</tr>
<tr>
<td>Becker Nevus</td>
<td>6</td>
<td>2.3</td>
<td>1 2.7</td>
<td>0 0</td>
<td>1 1</td>
<td>4 4.9</td>
</tr>
<tr>
<td>Café au lait</td>
<td>2</td>
<td>0.8</td>
<td>1 2.7</td>
<td>0 0</td>
<td>1 1</td>
<td>0 0</td>
</tr>
<tr>
<td>Tuberousclerosis</td>
<td>2</td>
<td>0.8</td>
<td>1 2.7</td>
<td>1 1.9</td>
<td>0 0</td>
<td>0 0</td>
</tr>
<tr>
<td>A. nigricans</td>
<td>2</td>
<td>0.8</td>
<td>0 0</td>
<td>0 0</td>
<td>1 1</td>
<td>1 1.2</td>
</tr>
<tr>
<td>Albinism</td>
<td>1</td>
<td>0.4</td>
<td>0 0</td>
<td>0 0</td>
<td>0 0</td>
<td>1 1.2</td>
</tr>
</tbody>
</table>

Benign melanocytic nevi, which originate from melanocytes are commonly seen in infants. They display an increase in puberty. Benign nevi are seen at a rate of 6-12%, whereas atypical nevi are seen at a rate of 1% [10-14]. We did not come across any atypical nevi during this study. The prevalence of pigmented nevi has been reported as 2-14.4% in Turkey[7,9,11-16] 13.6% in Sweden, 9.1% in Switzerland, 0.05% in Hong Kong and 0.17% in Kuwait[1-4]. Prevalence varies with country. In our study, we found this rate to be 2.1% with most cases (25%) being in the 0-2 age group where lesions were mostly located on the head and trunk. Since families
worry more about nevi, they seek medical opinion more. Having other cases of nevi in the family was not found to be statistically significant.

Vitiligo is an autoimmune disease of the skin and mucosa which is characterized by depigmentation. The prevalence of vitiligo is 0.5-2%[17]. There are very few studies about the epidemiological properties of vitiligo in children in Turkey. Previous studies in our country have reported the prevalence to be between 1.4-8%[9,10,15,16,18]. The prevalence of vitiligo was found to be 0.4% in Egypt, 0.8% in Switzerland, 0.15% in Kuwait and 1.28% in India[1,2,13,19]. We determined this rate to be 1.8% in our study. In 50% of patients onset occurs before the age of 20, mostly between 8-12 years of age [20]. Handa et al, in a study of 625 cases, reported the average age of onset as being 6.2 and found that it more commonly affected girls[20]. In our study, 60% of cases were between the ages of 6-16. It has been reported that vitiligo is seen more in females than in males, but according to studies with large sample groups the difference was not found to be significant[21]. We found these rates to be 48% in girls and %52 in boys, and this difference was not found to be statistically significant. Vitiligo cases have a 10-27% of family story and in these cases the onset of vitiligo is early, usually before the age of 30. This supports genetic factors in its pathogenesis[20]. Aksoy and friends determined a family story in 16% of first degree relatives in a 63 patient group[21]. As in the study by Aksoy et al[21], we also found the prevalence of vitiligo in first degree relatives to be 11% and 5% in second degree relatives. Vitiligo is more commonly seen in the middle class that live in urban environments.

Nevus depigmentosus (achronic nevus) is a rarely seen cutaneous leucoderma most commonly seen before 3 years of age. It is common on the trunk and proximal extremities. They are one sided and do not pass the midline. They are found in equal frequencies in males and females [3,23]. In our study, the rate of nevus pigmentosus was found to be 0.5%, with most cases being between the ages of 0-2 and affecting the trunk most. In a comprehensive study by Nanda et al, the prevalence of depigmentosus nevi was found to be 0.73% [1].

Postinflammatory hypopigmentation is seen in patients with diseases such as psoriasis, eczema and pityriasis versicolor, seen as well defined grayish white lesions due to obstruction in melanosome transfer at the lesion site, [24] whereas post inflammatory hyperpigmentation is due to the accumulation of melanin phagocytized by macrophages after epidermal injury. It is a scar of previous disorders such as burns, lichen, pemphigus, eczema, light dermatosis, porphyria and xeroderma pigmentosum. Nanda reported postinflammatory hypopigmentation at a rate of 0.4% and postinflammatory hyperpigmentation as at a rate of 0.6% [1]. In Turkey, this rate is between 0.02% and 1% [9,11]. The most comprehensive retrospective study was conducted by Sardana and friends in India which included 30.078 children between the ages 0-12.
They found the prevalence of postinflammatory pigment disorder to be 1.68% [13]. In our study, we found this rate to be 0.5%. Although both were seen more in the middle class, hypopigmentation was found to be more common in low income districts and hyperpigmentation more in higher income districts.

Becker nevus which usually occurs in boys after the age of 10 is seen commonly on seen as a light brown widening organoid lesion on the shoulders and upper arms. Arnold et al found its prevalence to be 0.5% [25]. In our study, we found its rate to be 0.02%, occurring more common on the shoulders of children between the ages of 12-16.

Ephelis are brown macules that occur in light skinned individuals from exposure to the sun [26]. It is seen at differing rates among communities. In Sweden, the prevalence of ephelis lesions was reported to be 18.4% [3]. We only observed 7 cases (0.2%) and most were female. This low rate may be due to families not paying too much attention to these lesions.

Café au lait marks are usually seen in 10-20% of births as round or oval shaped lesions. They can be associated with certain neurocutaneous diseases. Fung et al reported the prevalence of café au lait lesions as being 4.4%, Wenk et al as 8% and Karaca et al as 1.5% [2, 4, 15]. In our study, we only observed two cases with lesions on the trunk of the body with the rate of 0.07%. Tuberousclerosis is an autosomal dominant neurocutaneous disease characterized by skin lesions, epilepsy and mental retardation. 85% of cases have 1 to 100 leaf shaped hypopigmented skin lesions [25-27]. We had two male cases in this study.

Acanthosis nigricans is characterized by hyperpigmented hyperkeratosed verrucous plaques. It is frequently related to endocrine disorders such as insulin resistance, obesity and malignancies. It is seen between 0.5-74% [17,24]. In our study, we had two cases that were in the 5-12 age group being treated for obesity.

Albinism is hereditary disorder caused by deficiency of tyrosinase, which is needed for melanin synthesis, and is characterized by decreased pigment in the eyes, skin and hair. Its occurs one in 17000-20000[22]. We only had one boy case in our study.

In conclusion, vitiligo and nevi are pigmentation disorders that cause concern in families. Our study gives us an understanding of the dermatological diseases that present with pigmentation disorders in our country because it was conducted in a reference hospital in the second largest city in Turkey. We believe that to better determine the problems, more comprehensive epidemiological studies are needed.

References

7- Inanir I, Sahin MT, Gündüz K, Dinc G, Turel A, Oztürkcan S. Prevalence of skin conditions in primary school children in


