Basal Cell Carcinoma in Children

Report of 3 Cases

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Background: The peak incidence of basal cell carcinoma occurs in the seventh decade of life and is rare in children. When found in the pediatric age group, basal cell carcinoma is usually associated with a genetic defect, such as basal cell nevus syndrome, xeroderma pigmentosum, or nevus sebaceus. In areas of intense UV radiation exposure, such as the southwestern United States, children may be at increased risk of developing this malignancy de novo.

Observations: Three children (2 boys, aged 8 and 16 years, and an 11-year-old girl) from Tucson, Ariz, with isolated basal cell carcinoma unassociated with any other disease or syndrome are described.

Conclusions: Basal cell carcinoma in children is probably the result of a combination of UV radiation exposure and genetic background. Early recognition in children can prevent extensive tissue destruction and excess scarring after excision. A higher index of suspicion for basal cell carcinoma may also aid in prompt diagnosis of a possible genetic disorder, such as basal cell nevus syndrome.


REPORT OF CASES

CASE 1

A 16-year-old Hispanic boy presented with a solitary heme-crusted nodule that had a raised ringlike border below the left orbit (Figure 1). He had first noticed the lesion 2½ years earlier, when it was a much smaller papule. Since that time, the lesion had enlarged considerably. No family history of skin cancer was noted. The differential diagnosis included BCC, discoid lupus erythematosus, and deep fungal infection. A diagnostic biopsy specimen was obtained and sent for histologic evaluation. The specimen showed BCC. A dermatologic surgeon excised the lesion using the Mohs micrographic surgery technique. There were no recurrences or new primary skin cancers during 4 years of follow-up.

CASE 2

An 11-year-old white girl with skin type III was seen for the evaluation and treatment of nonresponsive acne vulgaris. During the initial examination, a 3-mm dome-shaped, slightly translucent papule was noted on the left nasal labial fold. The patient's family history was negative for skin cancer. The differential diagnosis included epithelial cyst, fibrous papule, and BCC. Histologic examination of a diagnostic biopsy specimen showed nodular nests of basal neoplastic cells invading the dermis, consistent with nodular BCC. The lesion was excised using the Mohs micrographic surgery technique. Follow-up 4 years later revealed no recurrences and no new primary tumors.

CASE 3

An 8-year-old boy with a history of several severe sunburns presented with a
growing, spontaneously bleeding, 3-mm papule under the left eye (Figure 2). The lesion was umbilicated and had overlying telangiectasias. The patient had no family history of skin cancer. The differential diagnosis included BCC and molluscum contagiosum. A diagnostic biopsy specimen revealed nests of basal neoplastic cells in the dermis, consistent with nodular BCC. The patient and his parents elected to have the lesion removed by electrodesiccation and curettage. Two years after treatment, the patient had no recurrences and no new skin cancers.

COMMENT

Nonmelanoma skin cancers are the most common malignant neoplasms in the United States, representing one third of all cancers diagnosed every year.5,6 Basal cell carcinoma represents 75% of nonmelanoma skin cancers and has an estimated annual incidence of more than 700,000 cases nationally.7,8 The US average annual incidence of BCC in whites is currently 191 per 100,000 and is increasing at a rate of 3% to 7% per year.7,9

Ultraviolet radiation exposure is partly responsible for both BCC and squamous cell carcinoma, as evidenced by their increased prevalence after chronic exposure to sunlight and the preponderance of these lesions on sun-damaged skin. Although squamous cell carcinoma is associated with cumulative sun exposure, BCC in younger patients does not show this association.10,11 D’Errico et al10 report that BCC arising before the age of 40 years corresponds with childhood or recreational sun exposure but does not correlate directly with cumulative sun damage. Thus, in areas of the world where the UV radiation is most intense, such as the Sunbelt in the United States, childhood sun exposure is at a maximum and younger patients are at a higher risk of developing BCC.

Other factors besides sunlight are reported to influence the development of BCC. Gailani et al11 note a strong association between BCC and the inactivation of a gene at chromosome 9q22, which is thought to be a tumor suppressor. Inactivation of this gene was found in tumor tissue in 68% of BCCs examined and did not correlate directly with sun exposure or age. The cause of this mutation is unknown, but possible factors may include ionizing radiation, arsenicals, and polyaromatic hydrocarbons. Basal cell nevus syndrome and xeroderma pigmentosum represent inherited genetic mutations that predispose those affected to BCC. Patients with basal cell nevus syndrome are found to have a germline mutation on chromosome 9.12

The peak incidence of BCC occurs in the seventh decade of life.13 In the pediatric age group, BCC usually occurs in the setting of a known genetic defect (Table). Although uncommon, isolated BCC in children without these conditions has been reported.16-20 Price et al14 described a 17-year-old boy with a solitary BCC of the nose. The patient had a history of sunburns 1 or 2 times per

**Table: Genetic Defects Predisposing Children to Basal Cell Carcinoma (BCC)**

<table>
<thead>
<tr>
<th>Genetic Defect</th>
<th>Age at First BCC</th>
<th>Inheritance</th>
<th>Clinical Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Basal cell nevus syndrome</td>
<td>Childhood or adolescence</td>
<td>Autosomal dominant</td>
<td>Multiple BCCs, pits of palms and soles, cysts of jaw, ectopic calcification of cranial membranes</td>
</tr>
<tr>
<td>Xeroderma pigmentosum</td>
<td>Around 6 y</td>
<td>Autosomal recessive</td>
<td>BCC, squamous cell carcinoma, and melanoma, increased freckling, pigmentation, and dryness of sun-exposed skin, congenital lesion of scalp or face</td>
</tr>
<tr>
<td>Nevus sebaceus</td>
<td>Rare before puberty</td>
<td>No known inheritance</td>
<td>Lesions contain excess numbers of sebaceous glands, abortive hair follicles, and ectopic apocrine glands</td>
</tr>
</tbody>
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year since the age of 9 years. His mother had a BCC removed at the age of 44 years. Histologically, the tumor was described as superficial BCC. Scobie and Preston\(^{17}\) described a 4-year-old boy with a BCC of the scalp. The patient presented with a small "cyst" on the occipital region of the scalp and a family history of skin cancer. The lesion, described histologically as well defined, recurred 8 months after excision. Excision was repeated without recurrence of tumor, based on follow-up 1 year later.\(^{17}\) A 12-year-old boy living in Arizona was described by Comstock et al\(^{18}\) with a BCC on the nose. The lesion was described as superficial BCC. Scobie and Preston\(^{17}\) described a 4-year-old boy with a BCC of the scalp. The patient presented with a small "cyst" on the occipital region of the scalp and a family history of skin cancer. The lesion, described histologically as well defined, recurred after a 4-month delay in diagnosis.

It is debatable whether BCC is more aggressive in children. Leffell et al\(^{30}\) defined aggressive-growth BCC as sclerosing, morpheaform, infiltrative, or invasive into nerves. Their retrospective review showed an increased occurrence of aggressive-growth BCC in patients younger than 35 years old compared with older patients. In contrast, Betti et al\(^{13}\) and Dinehart et al\(^{10}\) found no increase in the frequency of the morpheaform pattern in younger patients. All 3 of our patients had histologically less aggressive forms of BCC.

As total incidence rates of BCC continue to rise, childhood cases may become more common. This increase in pediatric BCC may be especially true in areas of high-level UV radiation exposure. The percentage of sunny days during the year, higher altitude, and location closer to the equator may place children in these areas at increased risk. Early recognition can prevent extensive tissue destruction and scarring after excision and aid in prompt diagnosis of a possible genetic syndrome. We recommend that clinicians have a higher index of suspicion for BCC when evaluating questionable lesions in children.

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REFERENCES