Multiple Brownish to Black Papules in a 13-year-old Boy

Yi- Ming Huang Woan-Ruoh Lee

CASE REPORT

A 13-year-old boy was found to have multiple brownish to black papules over neck, shoulder, abdomen and back since 1995. He was diagnosed with cerebellar medulloblastoma in 1991.Total tumor excision was done. Six courses of adjuvant chemotherapy with vinblastine, VP-16 and cisplatin were administered later. A huge recurrent tumor over the right temporal lobe with mass effect was noted in 1994. The patient underwent total recurrent tumor removal. Adjuvant craniospinal irradiation was given in the subsequent 2 months. No more recurrent tumor was noted during the follow up period. However, multiple brownish to black skin lesions were noted since 1995 which was one year after the completion of radiation. The skin lesions increased in size and number over the next year.

In the physical examination, numerous flesh-colored and brwonish papules were found in the area of the shoulders, abdomen (Fig. 1A), back (Fig. 1B) and neck where the patient had previous irradiation for medulloblastoma. The papules measured about 0.5-1mm in size, some appeared pearly, whereas others were pedunculated in shape similar to skin tags. Multiple tiny pits with hyper- pigmentation were found on the palms. Skeletal anomalies including frontal bossing and mildly increased interorbital distance (Fig. 1A), kyphoscoliosis (Fig. 1B) were also found. Incisional biopsies were done to 2 representative skin lesions on the posterior neck and lower back (Fig 2). Lateral mandible X-ray (Fig. 3) showed a large cyst over the left mandible ramus.

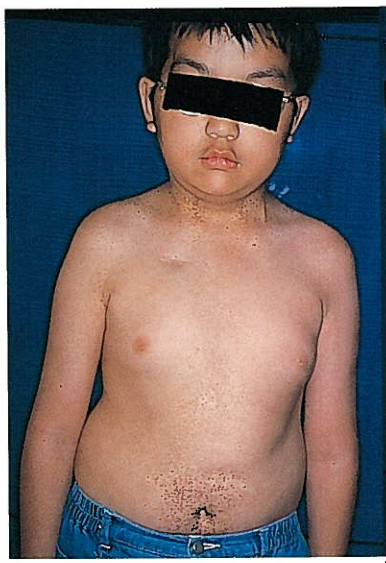

Fig. 1A

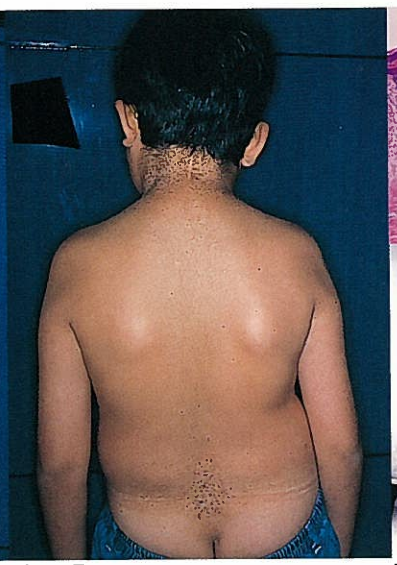

Fig. 1B

Fig. 2

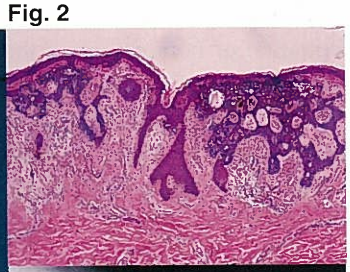

Fig. 3

From the Department of Dermatology, Taipei Medical University Hospital
Accepted for publication: October 29, 2001
Reprint requests: Woan-Ruoh Lee, Department of Dermatology, Taipei Medical University Hospital, 252 Wu-Shing Street, Taipei, Taiwan, R.O.C.
TEL: 886-2-23787139 FAX: 886-2-27372181

DIAGNOSIS: *Nevoid Basal Cell Carcinoma Syndrome*

DISCUSSION

The nevoid basal cell carcinoma syndrome,[1] also called Gorlin syndrome (NBCCS), is an autosomal dominant disorder characterized by both cancer predisposition and a variety of malformation.

Miscellaneous clinical features are noted in NBCCS syndrome.[1] Multiple basal cell carcinomas appearing as pearly to flesh- colored to reddish brown papules are found on the face, neck and upper trunk. Asymmetric palmar and/or plantar pits are also found. Jaw cysts, or odontogenic keratocysts occur after 7 years of age. Skeletal abnormalities include rib anomalies, and kyphoscoliosis. Central nervous system presentations include medulloblastoma, and meningioma. Other findings including cleft lip/palate and ovarian fibroma were also reported.

Alteration to the human homologue (*PTCH*) of *Drosophila* segment polarity gene patched (*ptch*) were identified in NBCCS patients as well as tumor tissue found in this syndrome.[2] Loss or inactivation of both copies of the normal allele causes predisposition to cancer formation.[3]

In our case, multiple basal cell carcinomas occurred over the area that was irradiated for medulloblastoma within one year after the completion of craniospinal radiotherapy. Strong[4] reported numerous basal cell carcinomas develop in the irradiated area, usually after 6 months to 3 years following radiotherapy. By the above finding, we can speculate that radiation energy can induce somatic mutation on the second allele in the tumor cells.

Any patient who presents with medulloblastoma at an unusually young age should be evaluated for NBCCS. In the past years, the medulloblastoma was treated with surgical excision and irradiations. However, increasing cases have showed that basal cell carcinomas developed after exposure to the ionizing radiation. Treatment with chemotherapy alone or reduced dose radiation therapy with chemotherapy, instead of standard-dose radiation therapy would be more beneficial to the patient.[5]

Treatment for multiple basal cell carcinomas is still challenging on account of multiple lesions with widespread distribution. Large numbers of the smaller ones can be electrodessicated and curetted under general anesthesia. Cryotherapy and topical 5-fluorouracial agents have been used with success. Periodic 3 to 6 months follow-up visits are warranted, at which total body cutaneous examination should be performed.

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