**Case 7**

A 60-year-old Thai female from Surin

**Chief complaint:** Multiple papules on her face for 40 years

**Present illness:** The patient presented with multiple papules on her face since age of 20. The lesions had persisted for several years with gradually increased in size during the past year. She noticed some lesions on her palms during the same period. She experienced multiple episodes of seizure since age of 50 and had to continue antiepileptic drug since then. 3 years ago, she gradually developed right side weakness. CT brain was done and showed solitary brain tumor at left parietal area.

**Past history:** as above

**Family history:** Her daughter had similar skin lesions on palms and soles.

**Physical examination:**

HEENT: not pale, no icteric sclerae

LN: not palpable

Abdomen: no hepatosplenomegaly

NS: Motor gr IV (Rt) gr V (Lt), sensory intact, cranial nerve grossly intact, DTR 2+ all

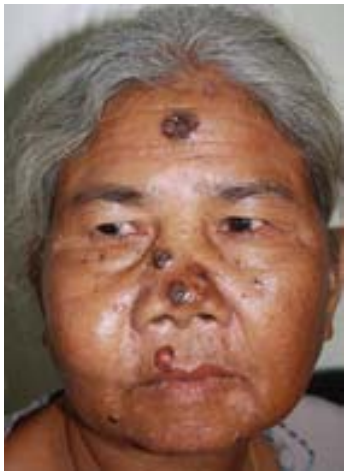
**Skin examination:**

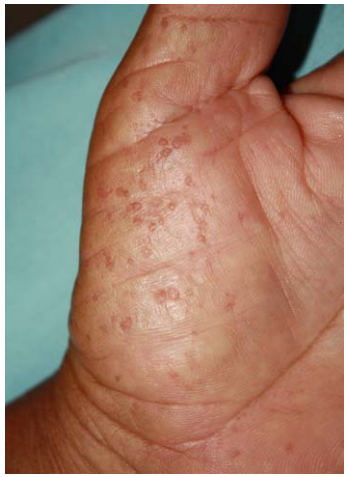
**-** Multiple brownish shiny papules and nodules with rolled border and central ulceration on face

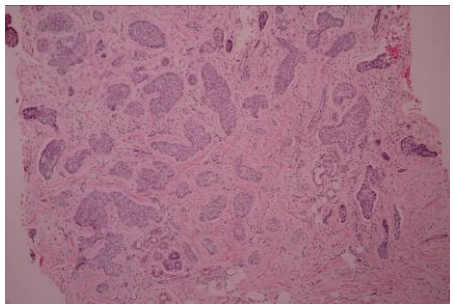
- Multiple palmoplantar pits

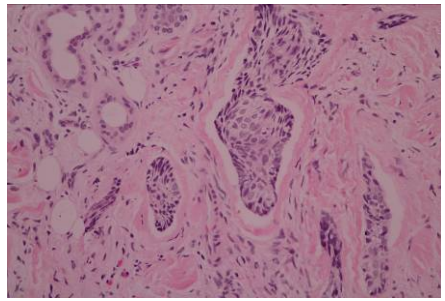
**Histopathology:**

Multiple aggregrates of atypical basaloid cells, some surrounded by clefts, in the upper dermis.









**Investigation:**

**X-ray skull series**: calcified falx cerebri, no jaw cyst

**CT brain:** single brain tumor with calcification at left parietal area

**Diagnosis:** Basal cell nevus syndrome

**Presenter:** Wikanda Panmanee
**Consultant:** Natta Rajatanavin

**Discussion:**

Basal cell carcinoma is a slow-growing malignant neoplasm derived from nonkeratinizing cells that originate in the basal layer of the epidermis. It is the most common skin cancer. Over 1 million new cases occur each year in United States.[1]

The pathogenesis of BCC involves exposure to UVL, particularly the UVB spectrum. Germline mutations in PTCH are observed in hereditary basal cell nevus syndrome but mutations in PTCH are also seen in sporadic BCCs at a rate of 30–40% with 41% of sporadic BCCs demonstrating UV signature mutations.[2] The development of BCCs in most individuals is multifactorial. The established risk factors are UV radiation, indoor tanning exposure, smoking, ionizing radiation. However, BCC is not directly correlated with cumulative UV exposure like SCC.

BCC is typically occur in middle-aged to elderly patients but less commonly in younger ones. In one previous study, most BCCs seen in patients younger than 40 years are found in women who are more likely to have a past or current history of cigarette smoking and blistering sunburns.[3]

Furthermore, BCCs arising in young adult also found in heritable condition called basal cell nevus syndrome which is an autosomal dominant condition characterized by five major components, including multiple nevoid basal cell carcinomas, jaw cysts, congenital skeletal abnormalities, ectopic calcifications, and plantar or palmar pits.

BCC usually develops on sun-exposed areas of head and neck but can occur anywhere on the body. The highest percentage of lesions occurred on the nose. Characteristics may vary for different clinical subtypes, which include nodular, superficial, morpheaform, pigmented BCC and fibroepithelioma of Pinkus. They tend to invade locally rather than metastasis.

Aggressive BCC include morpheic, infiltrative and micronodular pattern according to the accepted definition. Among this group, morpheaform basal cell carcinoma is special consideration from its presentation that may resemble a scar or a small lesion of morphea. Thus, the appearance of scar tissue in the absence of trauma or atypical-appearing scar at the site of previously treated lesion should be alert and consider for biopsy.

Neurotropic BCC is uncommon except in aggressive or recurrent lesions. Different reports show a perineural invasion incidence of 0.18-3%.[4, 5] Male sex is predominate. The most common sites involved were the nose, cheek, maxilla and forehead. Large tumors with significant subclinical extension are features that linked to this aggressive type. 5-year recurrence rate was 7.7% after Mohs micrographic surgery.[5]

To select the best treatment method for a specific BCC, a thorough knowledge of the modality, its complications and cosmetic results, and the recurrence rates are required. Most commonly used treatment options for BCC divided into surgical options such as Mohs micrographic surgery, surgical excision, electrodessication and curettage and cryosurgery and non-surgical options such as radiotherapy, topical imiquimod, topical fluorouracil and photodynamic therapy.

Overall recurrence rates of facial BCC, aggressive or not, reach high values up to 26% in many studies.[6-8] However, the prognosis for patients with recurrent BCC is favorable. For the metastatic disease, prognosis is poor with a mean survival of 8 to

10 months from the time of diagnosis.

Recurrent BCC following ablative laser procedures was more likely to be ‘recurrent BCC’ that was incompletely removed by the previous ablation than malignant transformation.[9]

## References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