Multiple eruptive pilomatricomas in a young woman with glioblastoma multiforme

Received: February, 2020
Accepted: July, 2020
Published: February 2021

DOI:
10.25259/IJDVL_135_20

PMID:
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Sir,

Pilomatricoma is a benign tumor arising from the hair follicle cortex cells. Although they usually occur as a solitary tumor, about 5% of cases can present as multiple lesions. Multiple lesions usually have familial inheritance and can have syndromic associations such as myotonic dystrophy, Curschmann-Steinert

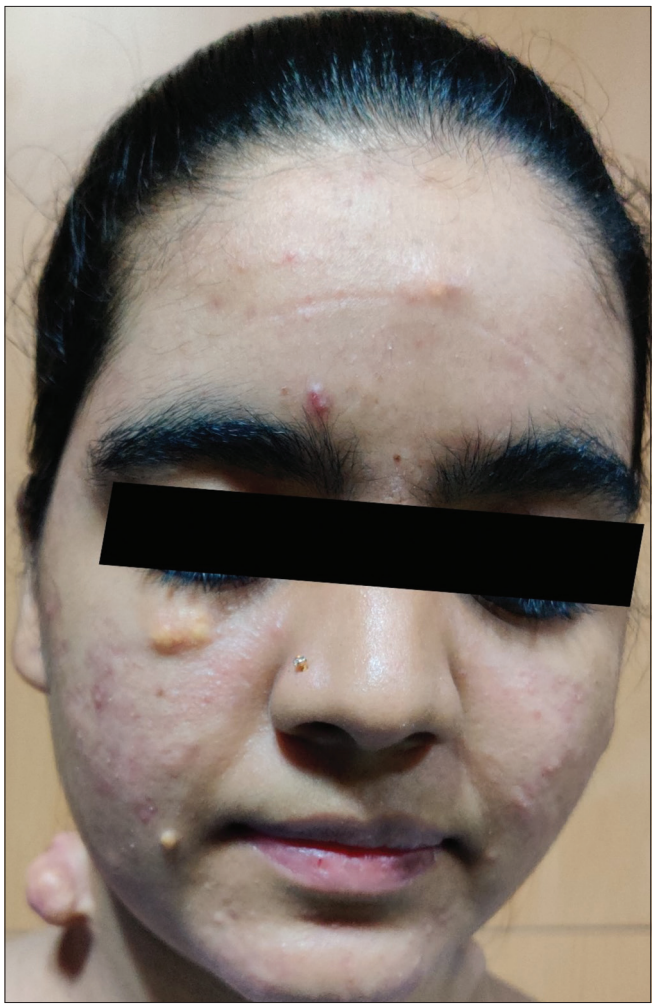

Figure 1a: Multiple eruptive pilomatricomas of varying size on face

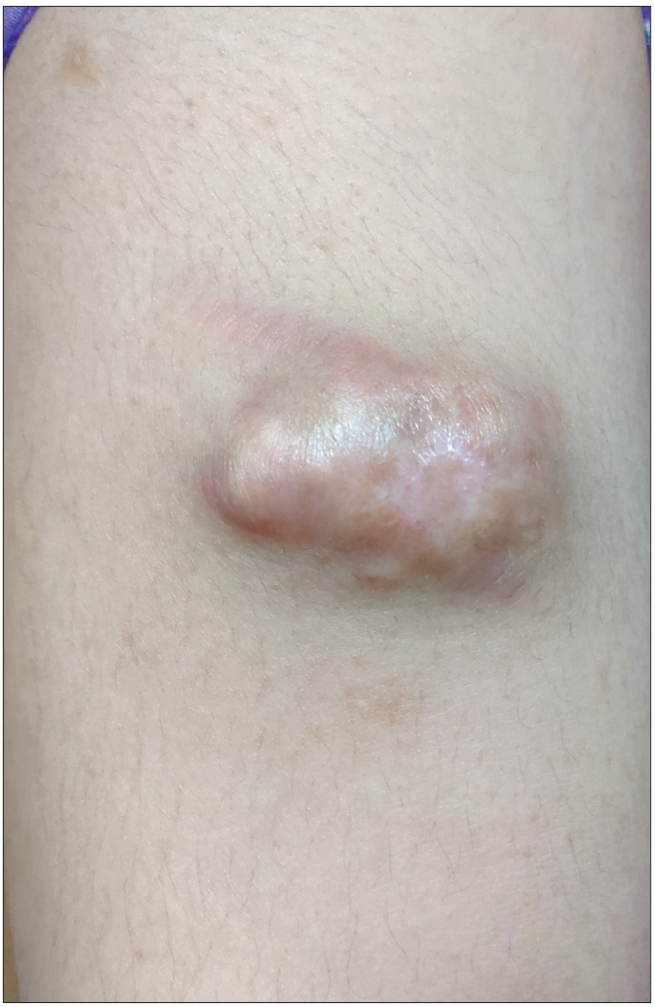

Figure 1b: Well-defined erythematous plaque with central areas of calcification on the right arm

How to cite this article: Harshatha S, Sivayogana R, Manuel A, Murugan S. Multiple eruptive pilomatricomas in a young woman with glioblastoma multiforme. Indian J Dermatol Venereol Leprol 2021;87:86-115.

syndrome, Gardner syndrome, Rubinstein-Taybi syndrome, Turner's syndrome, Sotos syndrome and gliomatosis cerebri.[1] Herein, we present a rare association of multiple eruptive pilomatricomas in a 20-year-old woman with glioblastoma multiforme. We found only a single case report of a similar association published so far in the literature.

A 20-year-old woman presented to our dermatology department with a sudden onset of widespread, multiple asymptomatic raised skin lesions for 2 months. Initially, the lesions were pea-sized but later progressed to attain the present size as depicted in Figures 1a-c. She was diagnosed with glioblastoma multiforme of the left temporal lobe 2 years back. She underwent surgical excision and completed radiotherapy and chemotherapy with temozolomide and phenytoin for seizures. Following this, magnetic resonance imaging was performed 6 months later, which showed complete remission of the tumor. There was no significant family history. The systemic examination was normal. Dermatological examination revealed multiple erythematous to skin-colored nodules on the face, trunk, upper and lower limbs. Lesions were non-tender, firm to hard in the consistency of size ranging from 0.5 × 0.5 cm to 5 × 3 cm. A few lesions showed bluish discoloration in the center and a few other lesions had central white hard areas indicating calcium deposition. There were no ocular, oral, or nail lesions. The patient did not exhibit any features of the abovementioned syndromes. Differential diagnoses considered were pilomatricoma, steatocystoma, bone tumors, epidermoid cyst and sebaceous carcinoma.

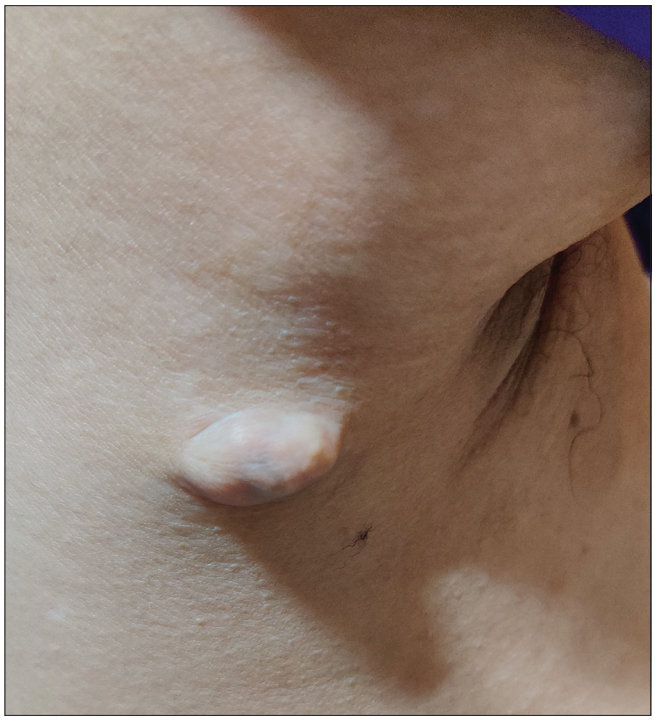

Figure 1c: Well-defined plaque with bluish discoloration in the center near the right axillary region

All baseline investigations were normal. Skin biopsy from one of the representative lesions, showed a partially circumscribed lesion composed of keratin, ghost cells and intermediate cells, with areas of calcification suggestive of pilomatricoma. There was no evidence of granuloma or malignancy [Figure 2a]. Glioblastoma tissue showed multiple fragments of neural tissue with adjacent cellular lesion with moderate atypia, vascular endothelial proliferation, hemorrhage and atypical mitotic figures [Figure 2b]. She underwent excision of cosmetically disfiguring lesions. Repeat magnetic resonance imaging was done 1 year later. It revealed gliotic changes in the left posterior temporal lobe suggestive of recurrence [Figure 3]. For this, she is being carefully monitored and managed conservatively under neurosurgeons.

Glioblastoma multiforme is a rare, rapidly growing, lethal brain tumor that belongs to the World Health Organization grade IV classification with a median survival period of only 14–15 months from the period of diagnosis.[2] Pilomatricoma occurring in the setting of glioblastoma, is extremely rare as only a single case has been reported worldwide.[3] Nuclear localization of the β catenin gene in basophilic cells is responsible for cellular proliferation in pilomatricoma and it is shown to be associated with a mutation in the β-catenin gene (most common being mutation in exon-3). Mutation in the β-catenin gene further

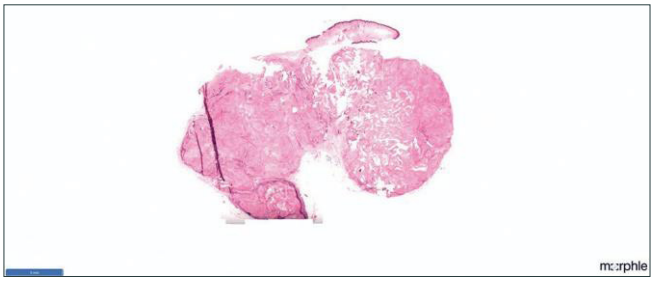

Figure 2a: HPE of pilomatricoma in scanner view

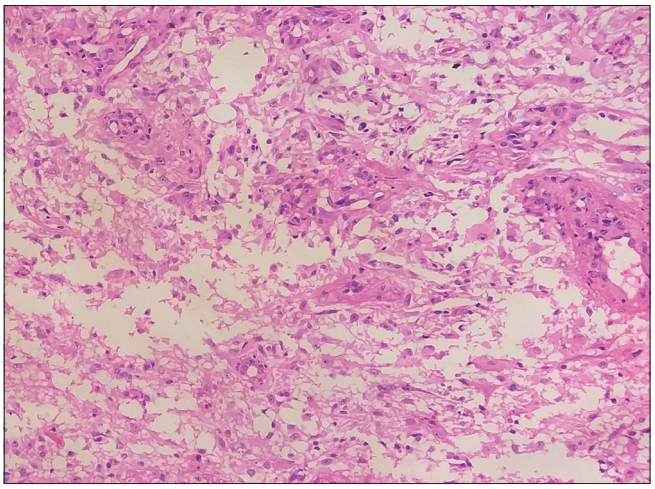

Figure 2b: Glioblastoma multiforme showing moderate atypia, vascular endothelial proliferation, hemorrhage and atypical mitotic figures (H and E, ×400)

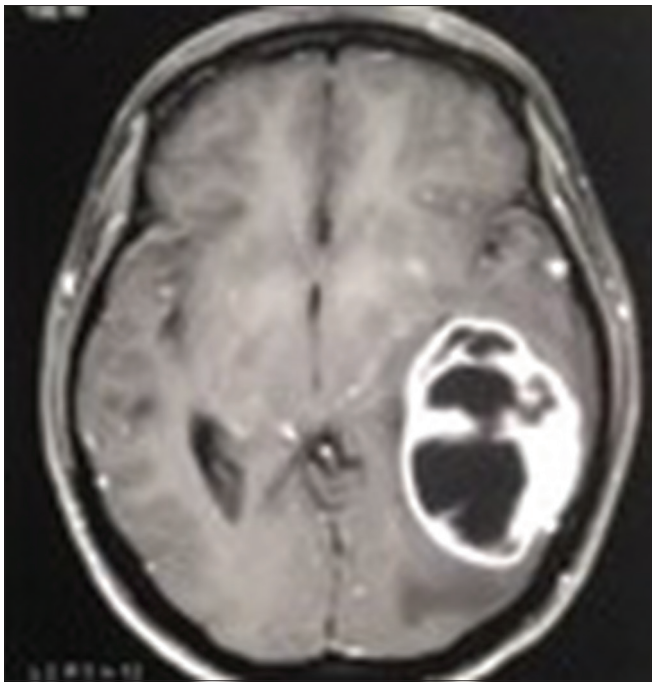

Figure 3: Magnetic resonance imaging-brain – glioblastoma multiforme tumor in posterior temporal lobe before excision

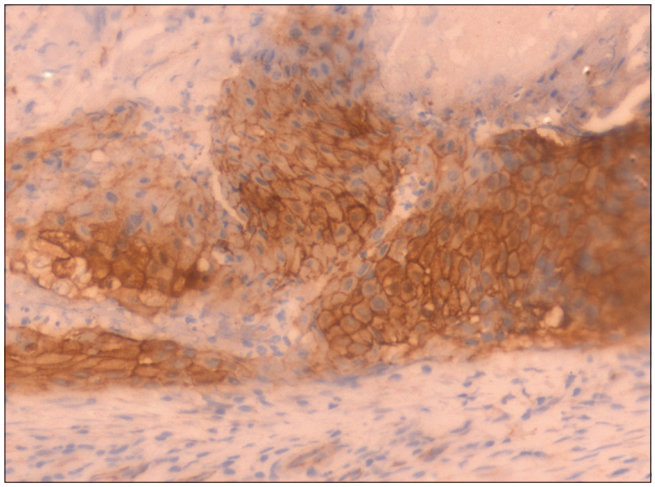

Figure 4a: Pilomatricoma tissue showing strong uptake of β-catenin (Immunohistochemistry, ×400)

causes Wnt signaling pathway activation.[4] Recent exploration has also established strong upregulation of the β-catenin gene in glioblastoma multiforme. This leads to the aggressive nature of the tumor, signifying a poor prognostic sign.[5] We suspected that similar β-catenin mutation was responsible for the development of pilomatricoma and recurrence of glioblastoma multiforme in our patient. Moreover, in the present case, there were no features of muscle dystrophy, muscle weakness, epidermoid cysts and osteomas, hypertrophic scars and keloid, cutis laxa and pterygium colli. By this, we have ruled out other genetic causes of tumorigenesis like myotonic dystrophy (dystrophia myotonica protein kinase gene), Curschmann-Steinert

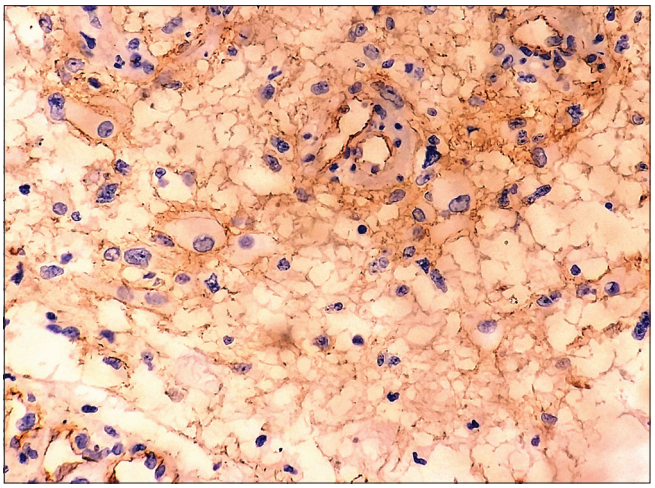

Figure 4b: Strong β-catenin staining of glioblastoma tissue (Immunohistochemistry, ×400)

syndrome (dystrophia myotonica protein kinase gene), Gardner syndrome (adenomatous polyposis coli gene), Rubinstein-Taybi syndrome (cAMP-response element-binding protein) and Turner's syndrome (chromosome 45, X0), respectively.[6] Since there was a proven association between pilomatricoma and glioblastoma from a previous case report,[3] immunohistochemical staining for β-catenin was carried on the pilomatricoma and the brain tissue and this was strongly positive. It showed the staining of the membrane, cytoplasmic and nuclear compartments of both the lesions [Figure 4a and b]. Unfortunately, we were unable to confirm it by genetic analysis, as our patient did not consent for it. Apart from the association of pilomatricoma with glioblastoma multiforme, Wachter has reported an association between multiple pilomatricomas and gliomatosis cerebri. In this report also, the role of β-catenin mutation in tumorigenesis has been stressed.[6]

We report this case to highlight the new rare association of glioblastoma multiforme and pilomatricoma which is not a mere coincidence, but has a strong genetic link concerning β-catenin and Wnt signaling pathway activation. Therefore, we hypothesize that the development of multiple eruptive pilomatricomas could be a cutaneous marker of recurrent and aggressive glioblastoma multiforme in the future.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal the identity but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest
There are no conflicts of interest.

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Dermoscopic features in a case of chondroid syringoma

Received: February, 2020
Accepted: June, 2020
Published: February 2021

DOI:
10.4103/ijdvl.IJDVL_167_20

PMID:
32969355

Sir,

A 42-year-old man with skin type IV presented with a one and half year history of a slow-growing, asymptomatic swelling on the right side of the upper lip. He denied any history of spontaneous ulceration or discharge from the lesion. Cutaneous examination revealed a solitary 1 × 1 cm firm, non-tender skin-colored to erythematous nodule on the right side of the upper lip [Figure 1]. The surface of the nodule was uneven and showed areas of brown pigmentation. There was no lymphadenopathy; other general and systemic examinations were within normal limits. Dermoscopic examination under polarized mode (Dermlite DL4, 10 × magnification) revealed white structureless area, milia-like cyst, irregular brown blotches with a few white lines, an erythematous rim at the periphery and linear curved vessels [Figure 2]. Differential diagnoses of nodular hidradenoma, solitary trichoepithelioma and nodular basal cell carcinoma were considered. Histopathology showed proliferation of epithelial cells arranged in nests, irregular tubulo-alveolar and ductal structures in a myxoid to fibromyxoid matrix. Apocrine decapitation secretion, cystic dilatation, clear cell change, mature adipocytes and keratocysts were evident at places [Figure 3a and b]. There was no evidence of any infiltrative growth pattern, pleomorphism, excessive/abnormal mitotic activity or tumor necrosis suggestive of malignant change. On immunohistochemistry, the outer epithelial and stromal cells were positive for vimentin and the inner epithelial cells for carcinoembryonic antigen and epithelial membrane antigen [Figure 3c and d]. A diagnosis of chondroid syringoma was made.

Cutaneous chondroid syringoma is a rare, benign, cutaneous sweat gland neoplasm that commonly occurs in the head and neck region of middle-aged men.[1] It usually presents as a solitary, well-circumscribed, asymptomatic, firm to hard, slow-growing, lobulated nodule of size varying from 0.5 cm to 3 cm. The color of the lesion varies from skin-colored to erythematous to bluish.[1] A consistent feature of chondroid syringoma is the lack of ulceration. Clinical diagnosis of chondroid syringoma is relatively difficult as it mimics other cutaneous tumors and cysts like basal cell carcinoma, hidradenoma, pilomatrixoma, sebaceous cyst and steatocystoma simplex.[1]

How to cite this article: Palit A, Sethy M, Nayak AK, Ayyanar P, Behera B. Dermoscopic features in a case of chondroid syringoma. Indian J Dermatol Venereol Leprol 2021;87:86-5.