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Case Report

Basaloid follicular hamartoma: an infrequent entity

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ABSTRACT

Basaloid follicular hamartoma is a rare, benign, skin adnexal tumor. Several clinical patterns have been reported, but they all share the same histopathological features. BFH may be hereditary or nonhereditary and can be accompanied by systemic diseases. Microscopic examination of BFH shows branching cords and anastomosing strands of basaloid cells in a loose, fibrous stroma. The most important pathological differential diagnosis is infundibulocystic basal cell carcinoma. These two lesions must be differentiated carefully based on clinical presentation and histopathological picture, and even molecular studies may be needed. We report a case of 60 year old female who came with a complaint of asymptomatic dark colored raised lesion over right side of face since 2 years. On evaluation lab investigations were found to be normal and histopathological examination showed interanastomosing cords and nests of basaloid cells, few abortive hair papillae and prominent melanophages. Based on these changes patient was diagnosed with basalloid follicular hamartoma and was advised for surgical excision.

Keywords: Basaloid follicular hamartoma, Benign tumor, Basaloid cells

INTRODUCTION

Basaloid follicular hamartoma (BFH) is a unique benign follicular hamartoma characterized by variable clinical presentations, identical histologic features and possible associations with numerous disorders. It was first described in 1969 by Brown et al in association with myasthenia gravis and diffuse alopecia (Brown-Crounse syndrome).[1] Characteristic histopathology of BFH is the presence of branching cords of undifferentiated anastomosing basaloid and squamoid proliferations connected to the overlying epidermis.[2] Basaloid follicular hamartoma may be acquired or hereditary. Hereditary types can be localized or generalized. Generalized forms are usually associated with systemic manifestations.[2,3] The acquired variants of BFH include localized and solitary forms. Localized BFH may present as a soft, hairless, irregularly elevated plaque in the retroauricular area or it may present as a localized plaque of scalp alopecia.[4] The solitary variant presents as a solitary, asymptomatic, smooth-surfaced, hyperpigmented papule or nodule that could arise anywhere particularly on the nose. We report a case of a 60-year-old female who presented with a single hyperpigmented atrophic skin lesion over right temple region of the face which clinically resembled basal cell carcinoma but was diagnosed as basaloid follicular hemartoma after clinico-pathological correlation.

CASE REPORT

A 60-year-old female came with a complaint of asymptomatic dark coloured raised lesion over the right side of face since 2 years. The lesion started initially as a small raised asymptomatic lesion which gradually progressed to attain the present size. The patient has not taken any treatment for this previously. No history of photosensitivity, any drug exposure, no history of any systemic symptoms suggestive of any connective tissue diseases, no history of any other systemic illness related to the present condition was present. General physical examination showed no abnormality. Past and family

history was not contributory. On dermatological examination well marginated plaque with central atrophy and raised margin was seen over right temple region of the face (Figure1).

Figure 1: Well marginated plaque with central atrophy and raised margin over right temple region of the face.

Figure 2: Three discrete hypopigmented blackish papules over anterior margin of the lesion.

Faint hyperpigmentation was seen over the mildly raised posterior border. Three discrete hypopigmented blackish papules were seen over anterior margin of the lesion (Figure 2). One papule showed central crater. No other skin lesion seen. Scalp, oral mucosa, hair, palms, soles and nails were normal. Based on the above clinical picture a differential diagnosis of superficial type of BCC, Discoid lupus erythematosus, porokeratosis, seborrheic keratosis (atrophic form) was considered. Complete laboratory workup showed normal hemogram and serum biochemistry profile. Viral screening for HIV, HBsAg and HCV did not show any abnormality. Imaging studies like chest xray and ultrasound abdomen were normal. Histopathology examination showed thin epidermis and interanastomosing cords and nests of basaloid cells in the superficial and deep dermis (Figure 3). Few abortive hair papillae (Figure 4) with papillary mesenchymal bodies are noted. In addition, the upper dermis showed prominent melanophages.

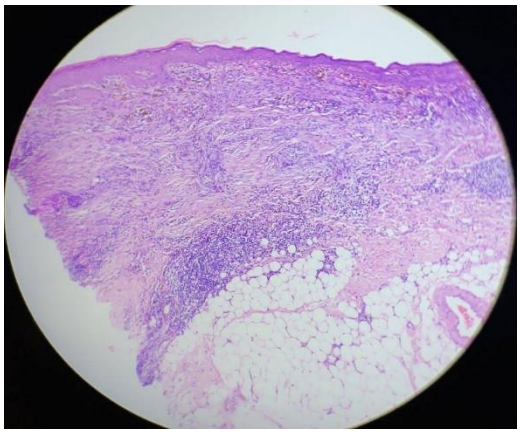

Figure 3: Interanastomosing cords and nests of basalloid cells.

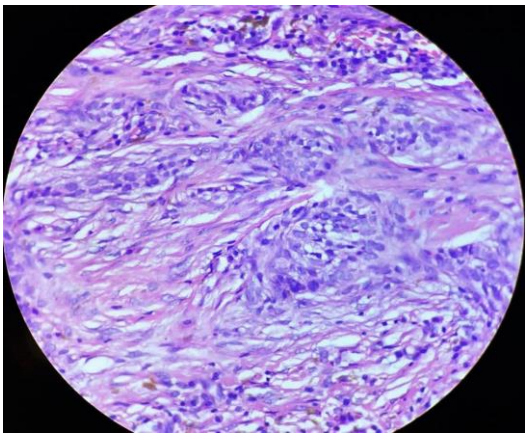

Figure 4: Abortive hair follicle.

There is mild perivascular and interstitial inflammatory infiltrate composed of lymphocytes, few plasma cells and occasional eosinophils. There is no evidence of atypia/significant mitotic activity. No features of peripheral palisading of basaloid cells, stromal retraction artefacts, stromal mucin were found. HPE findings are suggestive of Basaloid follicular hamartoma. Based on the clinical features and histopathology the case was diagnosed as basaloid follicular hemartoma and referred to plastic surgery for complete excision.

DISCUSSION

BFH is a rare, benign, superficial malformation of hair follicles first described in 1969 by Brown et al in association with myasthenia gravis and diffuse alopecia (Brown-Crounse syndrome. They can present as solitary lesions, or in a generalized form associated with systemic

disease. Basaloid follicular hamartoma occurs in two forms: hereditary and acquired. The hereditary types include generalized and unilateral (nevoid) forms, while the acquired types include localized and solitary forms.[5,6] Five clinical variants of BFH include solitary or multiple papules; a localized plaque with alopecia; a localized linear or unilateral papule or plaque; a generalized variant dominantly inherited familial type without associated disorders; and generalized papules associated with alopecia and myasthenia gravis. Individual lesions are small, skin-coloured to brown papules, nodules or plaques, with or without associated milia and/or comedones, commonly present over the face, scalp, and occasionally, the trunk.[5,7] In our case the patient presented with single hyperpigmented annular plaque over right temple region of the face. Histopathology revealing proliferation of basaloid cells in anastomosing strands and cords is a key marker in the diagnosis of BFH.[8] In our case histopathology examination showed thin epidermis and interanastomosing cords and nests of basaloid cells in the superficial and deep dermis with few abortive hair papillae with papillary mesenchymal bodies. There is no evidence of atypia or any features of peripheral palisading of basaloid cells. Basaloid follicular hamartoma and infundibulocystic BCC might actually represent the same disease.[9] Apart from the clinical and histopathological similarity, both entities have positive CK20 staining. Furthermore, dysregulation of the sonic hedgehog pathway has been implicated in the pathophysiology of both diseases. The gene aberration in BFH is a mutation in the patched (PTCH) gene on chromosome 9q23, which is the same gene implicated in nevoid basal cell carcinoma syndrome (Gorlin-Goltz syndrome).[9] However, the expression of the mutation in BFH is not as severe. There are no standardized treatments for BFH. Treatment options include surgery, cryotherapy, CO2 laser, and imiquimod. Vismodegib, an inhibitor of the hedgehog signalling pathway might play a role in the treatment of severe disease in the future.[10] The prognosis is usually excellent, unless associated with systemic disorders and/or BCC develop. In our case based on the solitary lesion and for practical reasons patient was referred to plastic surgery department for complete excision.

CONCLUSION

Our case highlights the infrequent entity of basaloid follicular hamartoma which clinically resembled superficial basal cell carcinoma and discoid lupus erythematosus but was diagnosed as basaloid follicular hamartoma after histopathological examination. In all cases of proliferative diseases detailed histopathological examination help in pinpointing the diagnosis and help in avoiding potential aggressive treatment.

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