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Adenoid Basal Cell Carcinoma - A Rare Variant and a Mimic of Adenoid Cystic Carcinoma

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***Abstract**: Basal Cell Carcinoma is a common cutaneous malignancy, occurring primarily on the face . It has many histological variants. We present a case of a 71- year old farmer with a nodule near outer canthus of the right eye, since six months. Histopathological examination of the excised nodule revealed a tumour composed of basaloid cells with an adenoid pattern of growth. Based on the histopathological and immunohistochemical features, a diagnosis of adenoid type of basal cell carcinoma was made. The adenoid BCC is a rare type and closely resembles the Primary Cutaneous Adenoid Cystic Carcinoma. Differentiation of these two histologically similar lesions is important as their evolution and prognosis is different.*
***Keywords:** Adenoid variant, Basal cell carcinoma, Cutaneous Adenoid Cystic Carcinoma.*

I. Introduction

Basal Cell Carcinoma is the most common cutaneous malignancy, arising from basally located cells of epidermis and pilo-sebaceous unit. It occurs mainly in adults, in the head and neck area, primarily on the face. Few cases are reported in children, usually in association with genetic syndromes. It is most commonly seen in light-coloured skin with history of prolonged exposure to sunlight. The clinical presentation is usually in the form of a nodulo-ulcerative growth. It has many histological sub-types including solid, micronodular , infltrative and pigmented. The Adenoid type is an uncommon sub-type and resembles the Primary Cutaneous Adenoid Cystic Carcinoma (ACC) histologically. The exact histogenesis of ACC is uncertain, but it is believed to be of eccrine or apocrine origin [1]. As a rule, Basal cell carcinomas (BCC) do not metastasize. We report one case of adenoid BCC with histological resemblance to ACC.

II. Case Report

Clinical presentation A 71-year old male patient presented with a slow-growing nodule near the outer canthus of the right eye, since 6 months..The patient was a farmer with history of long hours of sun-exposure. The lesion was 1.5 cm. in diameter, greyish-black, in colour (Fig.1).Clinical examination did not reveal any enlargement of regional lymph nodes.

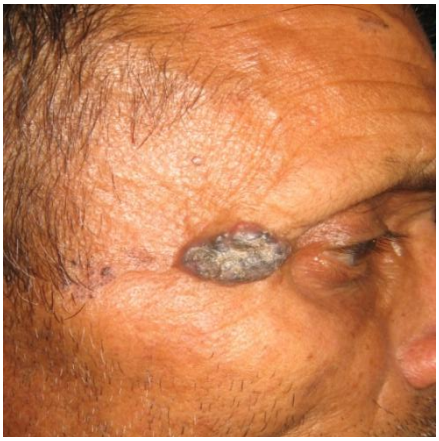

Fig 1. Clinical photograph of nodule near lateral canthus of right eye.

Radiological studies did not reveal any metastases. The clinical diagnoses offered were a Basal cell carcinoma or a Melanoma. An excision of the nodule was done.

Gross Morphology

A nodule measuring 1.5 cm was received for histopathological examination. The surface was greyish-black with focal ulceration. Cut surface was grey-black and firm.

Microscopy.

Histopathological examination of the H&E stained sections revealed a tumour composed of lobules of basaloid cells with a connection to the overlying epidermis(Fig 2a). The cells were arranged in an adenoid and lace-like pattern (Fig 2b). Retraction space was seen around few nests (Fig 2c). Palisading was noted at the periphery of an occasional nest (Fig 2d). Melanin pigment and melanophages were seen in many nests(Fig 2e) Some of the cells were large and showed abundant eosinophilic cytoplasm and nuclear enlargement with prominent nucleoli and mitosis (Fig 2f). The centre of many lobules showed amorphous eosinophilic material (Fig 2g). .Immunohistochemical studies for BerEP4 (Fig 3) showed diffuse staining throughout the lesion while staining for HMB 45 and EMA was negative, thus ruling out melanoma and cutaneous adenoid cystic carcinoma.

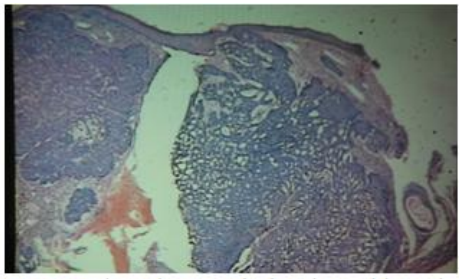

Fig 2a: Microphotograph showing epidermal attachment of tumour in upper dermis. (H&E, x40)

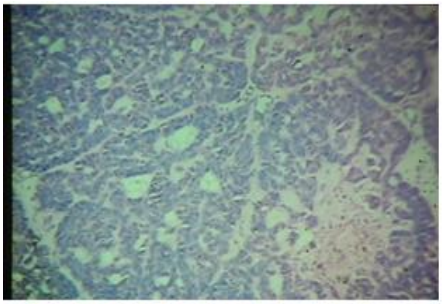

Fig 2b: Microphotograph showing adenoid Pattern of basaloid tumour cells (H&E,x100)

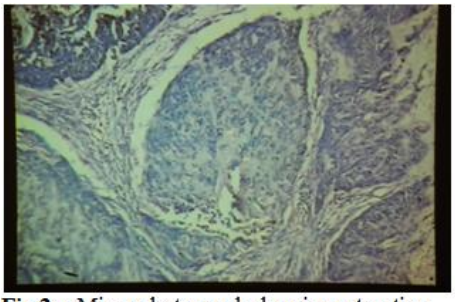

Fig 2c: Microphotograph showing retraction space around nests of tumour cells (H&E, x100)

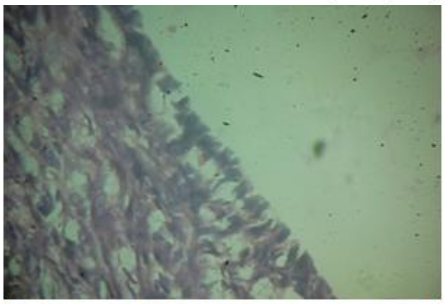

Fig 2d: Microphotograph showing peripheral palisading (H&E, x400)

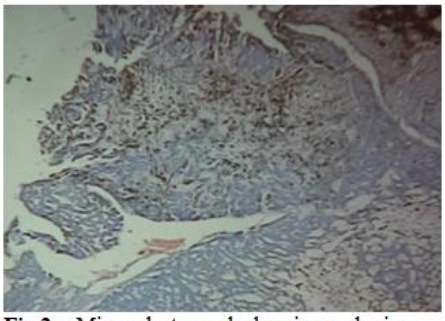

Fig 2e: Microphotograph showing melanin Pigment in tumour nests (H&E, x40)

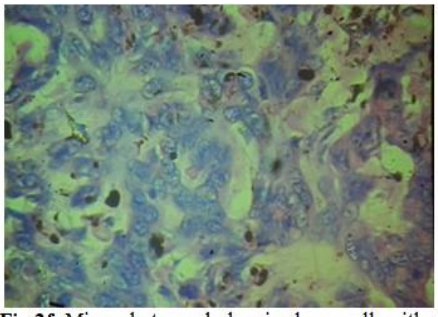

Fig 2f: Microphotograph showing large cells with eosinophilic cytoplasm & large nuclei (H&E,x400)

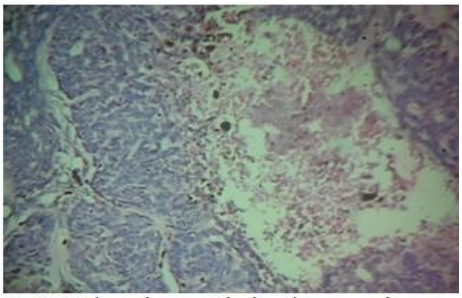

Fig 2g: Microphotograph showing amorphous granular material in the centre of tumour nest. (H&E, x100)

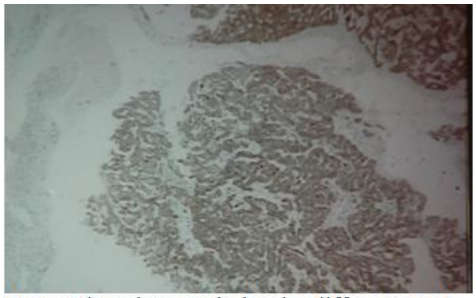

Fig 3: Microphotograph showing diffuse BerEP4 Stainnig in the tumour cells (Immunohistochemistry, x100)

III. Result

Based on the morphology, microscopic features of basaloid cells, connection with epidermis, retraction space and IHC expression for BerEP4 and absence of EMA staining, a diagnosis of Basal cell carcinoma of adenoid type was made.

IV. Discussion

Basal cell carcinoma was first described by Jacob in 1827 who called it "ulcus rodens". The present name was proposed by Krompecher in 1903[2]. They constitute a group of malignant cutaneous tumours characterized by the presence of lobules, columns, bands or cords of basaloid cells. They arise from basal epidermal cells or from pilo-sebaceous unit..Some researchers also refer to them as Trichoblastic carcinomas . Most of the basal cell carcinomas arise in the head and neck area, predominantly on the face. BCCs occur in adults from fourth decade onwards with a peak after sixty years of age and show a male preponderance. They are seen in children in association with genetic disorders like Basal cell nevus syndrome, Bazex syndrome, Xeroderma pigmentosum, etc. Besides being associated with prolonged exposure to sunlight as in rural areas or as part of a professional exposure such as in sports persons, other factors include arsenic exposure, radiotherapy and following burns and other scarring.

Clinically, basal cell carcinoma presents as pearly nodulo-ulcerative lesions, pigmented [melanoma-like] , morphea-like and fibroepithelioma-like lesions. Many histologic sub-types are well-known, the commonest being the solid nodular, infiltrative and superficial types [3]. All have in common, groups of basaloid cells in different arrangements, a connection with overlying epidermis, peripheral palisading of tumour cells and retraction or cleft in between tumour nest and surrounding stroma.

The adenoid variant is a rare histologic type which is characterized by basaloid cells in intertwining strands and lace-like pattern within cribriform or glandular spaces,, thus creating an adenoid pattern. Exact incidence of adenoid BCC is not known, but Bastiaens, et al. reported the incidence of 1.3% [3]. Adenoid BCC has been reported on un-exposed areas of skin and also at rare loctions like cervix and prostate. Tambe et al have reported an adenoid BCC at an unusual location in the lumbo-sacral area [4]. In a study of basal cell carcinomas of eyelids, Hussain et al reported an incidence of adenoid basal cell carcinoma at 6.67% in tumours of eyelids[5]. Recurrences are known to occur in BCC and are usually attributed to inadequate excision of histopathologically tumour-free margins. However, metastasis from BCC is rare and ranges from 0.0028%-0.55% ; less than 400 cases have been reported in the literature[6]. Adenoid BCC is regarded as a low grade malignancy compared to other more common subtypes like nodular and morpheic forms and treatment is similar to other BCCs. .

Cutaneous Adenoid Cystic carcinoma is an extremely rare tumour,first reported by Boggio in 1975[7] . It affects middle-aged and older persons and has a predilection for women. Adenoid cystic carcinoma is commonly seen in organs like salivary glands, breast, cervix ,larynx, ceruminous and lacrimal glands. When metastases from ACC of other organs is excluded, Primary Cutaneous Adenoid Cystic carcinoma (PCACC) is the most probable diagnosis. It is a slow-growing, tumour usually located on the scalp, chest and abdomen. It arises as a skin-coloured , firm nodule, measuring in size from 0.5-8cm and with an average duration of 9.8 years prior diagnosis[8].Ulceration and bleeding is variable.

PCACC is a tumour with disputed histogenesis, thought to be of salivary gland origin or from eccrine or apocrine glands. Histolologically, it shows a tumour located in mid and deep dermis, characterized by islands and cords of basaloid cells within glandular ,cystic, cribriform and tubular patterns. The glandular spaces show the presence of a mucus-like or hyaline membrane-like material. Absence of nuclear palisading and lack of connection with epidermis and retraction space help to differentiate it from BCC. Perineural invasion is often seen. It shows immunohistochemical staining for EMA and often for CEA. In one study, Wick et al have compared ACC of skin with ACC of salivary gland and adenoid BCC. They found that all four adenoid cystic carcinomas expressed posistivity for CEA and EMA while the adenoid basal cell carcinoma did not show positivity for both markers [9].

Cutaneos ACC has an indolent but progressive course with recurrence in more than 50% of patients. Kato, et al, have described a case of PCACC with lymph node metastasis[8]. Singh and Ramesh have described a case of PCACC with metastases to lungs[10]. In view of nodal and pulmonary metastases, surgical excision with wide tumour-free margin and follow-up radiation is recommended.

V. Conclusion

Adenoid basal cell carcinoma is a rare type of BCC. It shares the histologic pattern of Adenoid cystic carcinoma, and can be differentiated with the help of characteristic histologic features. Immunohistochemistry is a helpful adjunct, although not imperative, if histological differences are conclusive. Patients with metastasis in ACC have been reported while BCC rarely metastasizes. The distinction between these two tumours assumes great importance, since treatment modalities and prognosis of these two morphologically tumours is different.

Acknowledgements

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