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Post-Auricular malignant nodular hidradenoma in a young female – A Rare Case Report

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Abstract

Malignant nodular hidradenoma is a rare eccrine sweat gland tumour with the commonest site being the head and neck region followed by the extremities. It is seen mainly in the sixth decade of life with a slight male preponderance. The usual manifestation is that of a solitary, slow-growing painless papule or nodule. These tumours have been reported to have a high local recurrence rate.

We present a rare case of malignant nodular hidradenoma in a young female occurring in the post-auricular area.

Keywords: Malignant Nodular Hidradenoma, Young female, Post-auricular.

Introduction

Malignant nodular hidradenoma (MNH) is a rare eccrine sweat gland tumour arising from the secretory coil or the sweat duct. The most common site of occurrence is the head and neck region. It is seen commonly in the elderly with a slight male preponderance.(1) Usually, they are solitary tumours covered by intact skin; however, some tumours may show superficial ulcerations. In contrast to benign clear cell hidradenomas which are well demarcated, malignant ones show invasion into the surrounding tissue.(2,3) There may be angio-lymphatic invasion. Metastasis is early and usually leads to death.

Case Report

A 24 year old married female presented to the ENT department with a swelling behind the left ear for 6 months. Sometimes she had severe pain. On examination the outer surface was nodular, with haemorrhagic spots. The swelling measured 2.5 cm in diameter. The clinical impression was that of epidermal cyst or haemangioma. The mass was resected and sent for histopathological evaluation. The specimen measured 2.5x2.5x1 cm with the outer surface irregular and nodular. Cut sections showed solid and cystic areas along with haemorrhagic spots.

Histopathology showed sections covered by stratified squamous epithelium with underlying clusters and sheets of tumour cells not connected with the epidermis. The margins were irregular but pushing (Fig. 1a, b). The individual cells were round and showed features of anaplasia and mild pleomorphism. Nuclei were vesicular with coarse chromatin and had a small centrally placed prominent nucleolus. The cytoplasm was scanty and clear. Some of the cells had abundant clear cytoplasm. Mitoses was more than occasional, some of them atypical (Fig. 1c). Cells with clear cell change and focal glandular formation were evident (Fig. 1d). Stroma was pale, homogeneous and highly vascularised. Vascular invasion was evident. The neoplastic cells showed positivity with PAS stain (Fig. 2a). Immunohistochemical markers showed strong positivity with Cytokeratin (Fig. 2b) and negativity with CEA, ER, PR, CD34 and VEGF (Fig. 2c-g). Ki67 indicated a low proliferative index (Fig. 2h). A diagnosis of malignant nodular hidradenocarcinoma (MNH) was made. Post-operatively one year follow-up of the patient was uneventful.

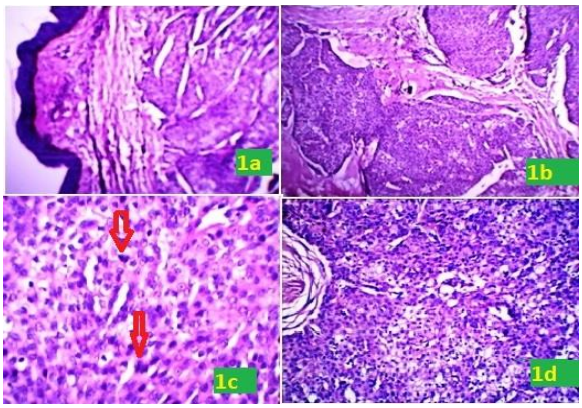

Fig. 1: (a) The tumour cells are lying away from the epidermis (H&E X 40); (b) Clusters and aggregates of neoplastic cells (H&E X 100); (c) Individual neoplastic cells with atypical mitosis (H&E X 400); (d) Cells showing clear cell changes with focal glandular formation (H&E X 100)

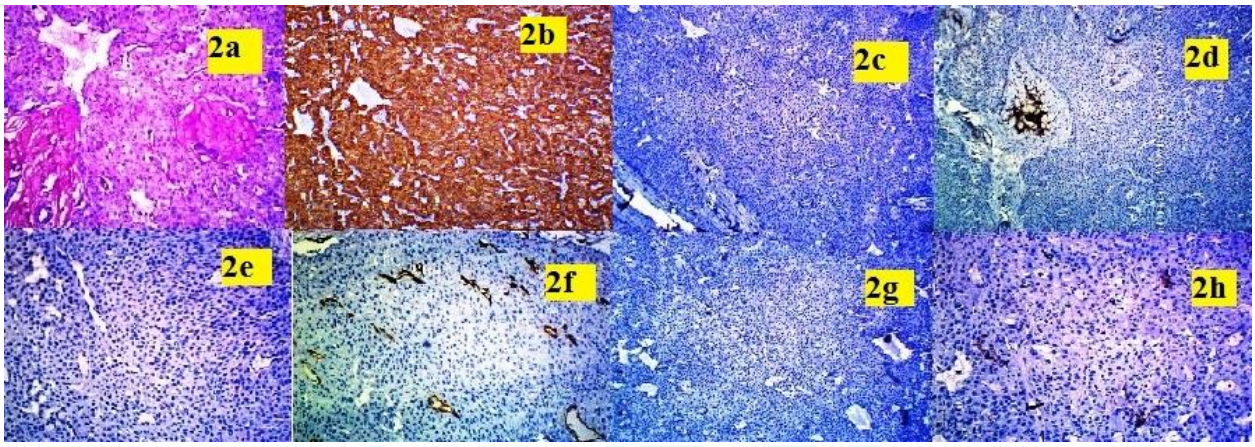

Fig. 2. Photomicrograph showing (a) PAS positivity; (b) Cytokeratin positivity; (c) CEA negativity; (d) ER negativity; (e) PR negativity; (f) CD 34 negativity; (g) VEGF negativity; (h) Low proliferative index on Ki-67

Discussion

MNH was first reported in 1954 by Keasbey and so far about only 70 cases have been documented in the English literature.[4] The overall incidence of all eccrine carcinomas is 6% which represent <1% of all skin neoplasms. Eccrine neoplasms (Poromas) tend to involve: 65% on the soles, 10% on the palms and 25% in other regions including extremities, face, neck and trunk. It usually manifests as a slow-growing painless papule or nodule on the head, neck or extremities. The peak incidence is in the sixth decade of life; with a slight male predominance.[5,6] MNH are mostly solitary lesions but if multiple then they are termed "poromatosis". These lesions occur in clusters and at times may appear hyperpigmented due to excessive melanin, or purplish due to proliferation of blood vessels. The diameter usually varies from 0.2 to 1.5cm.[6] The lesions may be ulcerated.

Adnexal neoplasms are derived from pleuripotent cell that have the ability to differentiate towards any of the lineage. Owing to a common origin many tumours may share their features while some may contain elements of two or more appendage in varying degree of maturation.[5] The incidence of adnexal neoplasms varies from 0.3-0.5%.[7-9] The morphological appearance is the presence of solid nests with or without cystic areas filled with mucinous material. The closely aggregated tumour cells display a round fusiform or polygonal biphasic cell population, one eosinophilic cell type and the other clear cell type.[10] Hidradenoma may exhibit both cystic and solid features and so needs to be differentiated from apocrine tumours. Co-existence of both eccrine and apocrine components is seen in Syringocystadenoma Papilliferum (SCAP) and Fox–Fordyce disease. Clear cell hidradenoma and hidradenocarcinoma may occasionally mimic metastatic clear cell carcinomas especially from thyroid, lung or kidney.[6] Malignant adnexal neoplasms, though rare have a poor outcome especially when not diagnosed early. Ackerman's diagnostic criterias to label a cutaneous tumour malignant include: irregular borders, asymmetry, horizontal orientation, epithelial cells arranged in irregular aggregates, large areas of necrosis, infiltrating of the dermis or subcutis, frequent mitoses with few of them atypical, stroma irregular and scanty, sometimes myxoid and presence of anisonucleosis.[7]

Immunohistochemitry reveals strong positivity for Epithelial Membrane Antigen (EMA), Estrogen (ER) and Progesterone Receptors (PR) and CEA. P63, CFDP-15 and C-erbB2 may be negative. The cells in the excretory coil express positivity for Low Molecular Weight Keratin, EMA and CEA, and S100 in the basal layer. The acrosyringeal cells (intra-epidermal portion) are stained with the High Molecular Weight Keratin (HMWK) and Cytokeratin (CK) 14. MNH show negativity for GCFDP-15. Ki67 has been reported positive in 40% of neoplastic cells.[6] Skin tumours with eccrine differentiation often express positivity for ER and PR, which has important clinical implications, since endocrinal therapy is possible in such cases.[9]

The general suggestion is to perform a wide excision. An inadequate excision of the primary lesion is a poor prognostic factor because even lesions labelled as benign have a tendency to recur locally if not completely excised. The 5-year postsurgical survival rate for MNH is reported to be less than 30%. Mohs micrographic surgery may prove superior to the conventional excision and manifests a lower recurrence rate. The choice of adjuvant therapy is controversial and is influenced by age and general condition of the patient. The tumour has a high local recurrence rate (50%), and tends to metastasise to the regional lymph nodes, bones or visceral organs in about 60% of cases.[1] A close follow-up for the initial 5 years is mandatory, during which there are maximum chances of recurrence and death. A radical surgery should be performed on the basis of histological diagnosis. Lymph node dissection is indicated only when there are signs of invasion; prophylactic dissection otherwise has not yet proved to increase the disease free interval.[1]

Conclusion

Malignant nodular hidradenomas are rare sweat gland tumours seen mainly in the head and neck region. Diagnosis is based on the peculiar histopathological features. Treatment is wide surgical excision and regular follow up.

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