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Huge malignant hidradenoma of the scalp: A case report

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Abstract
Malignant nodular hidradenoma (MNH) is rare, highly malignant, primary skin tumor which may have both apocrine and eccrine variants. MNH has peak incidence in the sixth decade of life; with an equal male/female distribution. Scalp is a known but it is unusual site of occurrence. Wide local excision is the treatment of choice with clearances of 2 cm recommended. Selective lymph node dissection is often used. The value of adjuvant radiotherapy has not been confirmed. As early recurrence is common despite the wide excision close follow-up is mandatory.

Keywords: malignant nodular hidradenoma, scalp

Introduction
Malignant nodular hidradenoma (MNH) is an infrequent, highly malignant, primary skin tumor which may have both apocrine and eccrine variants [1]. It has exceedingly low incidence of 0.001% [2]. Several synonyms have been reported in the literature, such as malignant clear cell myoepithelioma, malignant acrospiroma, clear cell hidradenocarcinoma, clear cell eccrine carcinoma, clear cell hidradenoma, solid-cystic hidradenoma and eccrine acrospiroma. Scalp is a known but it is unusual site of occurrence [3]. Most reported cases are in elderly people over 50 years of age, though they may occur at any age. MNH has a very poor outcome, high recurrence and a high metastatic rate [4].

Case report
History: A 17 year old male patient presented with painless huge mass at the left partial region of the scalp since 4 years.
It was asymptomatic to begin with, but started producing intense itch, associated with purulent discharge since last 8th months. There was no history of any local trauma, fever, pain and bleeding from the lesion.
The personal and family medical history were insignificant especially for any malignancies, immunosuppression or similar condition.
General physical examination did not reveal any significant clinical abnormality or any lymphadenopathy local or distant. The local examination revealed solitary a dull erythematous, firm, non-tender, non-fluctuant, cauliflower-shaped growth of size 9 × 8 cm which was fixed to the deeper subcutaneous tissues at left parietal region of scalp. The overlying skin was tense, thin and eroded at the center with crust and purulent discharge.
Head CT scan showed a large irregular cranial heterogeneous solid cystic mass mainly solid in the left partial region it measure about 90X8.7X7.7 which shows strong enhancement of the solid component as well as multiple feeding vessels seen in the base of the mass, the underlying bone shows no bone erosion, osteolysis or sclerotic changes.
The clinical and radiological differential diagnosis are lymphoma, hemangioma, lymphangioma, infected hematoma and benign skin tumors.
Metastatic work up revealed normal (Neck ultrasound, Chest CT scan And Abdominal ultrasound)
FNAC (Fine Needle Aspiration Cytology) was done before the operation and reveled groups of oval to spindle cells mix with stromal fragment. This cells have nuclei with mild to moderate nuclear pleomorphism and with nuclear overlapping, consisting with spindle cell neoplasm (corresponding to C4). Excisional biopsy is recommended.
Our decision was a wide local excision with safety margin, which was performed under general anesthesia, the defect of the mass was reconstructed with flap advancement and a split-thickness skin graft, which healed slowly in about 4 weeks.
It is probable that malignancy originated within the mass of benign hidradenoma that had been present since many years.
At gross pathologic examination, a nodule measuring 9X7X5.2 cm (height). Serial slicing show a gray white nodular tumor with small cystic space. Microscopically show malignant scalp tumor formed of nodules and nests of tumor cells centered in the dermis and infiltrating the subcutaneous tissue, the tumor cells have round to ovoid nuclei and clear to esinophilic cytoplasm. Moderate mitotic activity is noted and there are foci of tumor necrosis.
All Surgical margins including the deep margin were free of tumor, the nearest margin was 2 cm. The Patient advised to follow up in the outpatient clinic every 3 months in the first 2 years as early recurrence is common despite the wide excision.

Discussion
Sweat gland tumors are mostly benign. Primary eccrine carcinomas are uncommon tumors [5]. The first reported case of sweat gland tumor dates back to 1865 [6]. The largest number of cases of malignant sweat gland tumors was

provided by Berg and McDivitt in 1968 with a total of 101 cases included and till today it remains the largest case series published [7]. Hidradenocarcinoma accounts for around 6% of malignant eccrine tumors and accounts for less than 0.001% of all tumors [5].

MNH has peak incidence in the sixth decade of life; with an equal male/female distribution; MNH has been reported most frequently in the head and neck and as in our case. Rarely on the lower and upper limb [7]. Mostly the tumor cells originates in the ductal or secretary part of the sweat gland [8]. MNH is slow-growing tumor but may undergo a rapid phase of growth in a short period of time. The patients are often asymptomatic with solitary skin lesion measuring around 1–5 cm. At an obscure point in time, the tumor transitions into an aggressive form with expansion to regional or distant metastatic sites, most ordinarily to lymph nodes [8]. Malignant lesions may arise from benign nodular hidradenomas [9]. Histologically, a numbers of cell types, for example polygonal cells, clear cells and spindle cells may be present [10]. The cells express the high molecular weight cytokeratins CK5/6 and CK7, as well as p63, androgen receptor, estrogen receptor and sometimes Her-2/neu. Ki-67 and p53 staining may be useful histologic parameters [11, 12].

A proper understanding of the natural history of MNH is essential for suitable treatment. The initial treatment of MNH is very important to achieve a good result. Wide local excision with negative margins is the most effective treatment. Wide surgical excision with at least 2–3 cm has been advocated in some reports. If wide margins cannot be achieved because of anatomical or functional conditions, Mohs micrographic surgery can be used [13]. Regional lymph node metastasis, mostly effected so prophylactic lymph node dissection has been reported [14]. Lymph node dissection has remained area of debate since a long-term benefit has not been clearly demonstrated. More studies are necessary to determine whether this procedure results in a survival benefit or local control. In our case no lymph node dissection was done. Due to poor prognosis and high local recurrence rate, adjuvant chemotherapy and radiotherapy have been reported in some cases. Radiotherapy can be used when surgery is not feasible or negative surgical margins cannot be achieved. Recently, reports have shown that radiation therapy may have a role in preventing local recurrence, while in others, radio-resistance has been reported [15].

Chemotherapy has been used sporadically for residual metastatic disease, but until now, the efficacy of adjuvant chemotherapy has not been demonstrated either alone or in combination with radiation therapy. Estrogen receptor analysis has also been recommended for the treatment of MNH, where patients with positive tests may undergo hormonal therapy. Trastuzumab, a monoclonal antibody against HER2 receptor, was well-tolerated and effective as adjuvant therapy for MNH patients with HER2/neu positivity in some cases [16]. In our study, a marginal resection was achieved, therefore, no additional wide resection or adjuvant therapy was performed. The local recurrence rate has been reported within the first 2 years from diagnosis to be as high as 50% depending on the surgical margin. Metastatic lesions have been reported in the regional lymph nodes, lung and bone (60% of the cases). Once the tumor recurs, it turns highly aggressive with a strong tendency for invasion of surrounding tissues and metastases to distant sites [7]. The prognosis for survival is generally poor with a 5-year disease-free survival rate reported to be < 30% [17]. Even after the complete excision MNH has a potential for local recurrence, tends to metastasize, and often cause death [18]. In a report of seven cases from Italy, six patients died within 15-45 months of diagnosis [19].

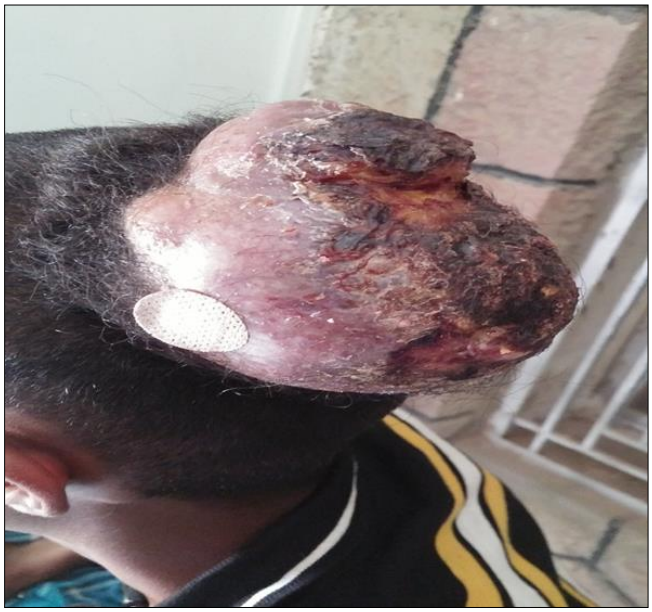

Fig 1

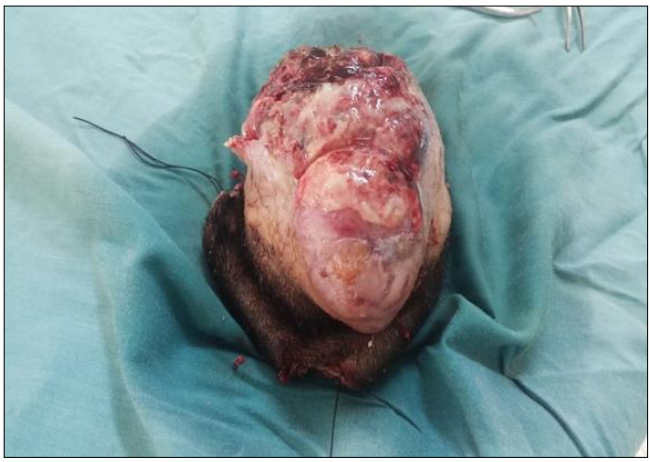

Fig 2

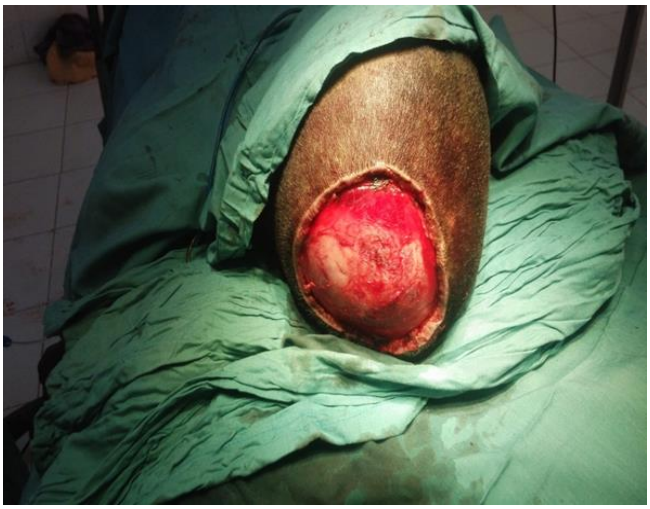

Fig 3

Fig 4

Fig 5

Conclusion

Malignant nodular hidradenoma is a rare aggressive cutaneous adnexal tumor. Early treatment should be contemplated in all. Wide surgical excision of the tumor with a least 2-3 cm of clear margins for both primary disease and local recurrences is the treatment of choice. Once the tumor recurs, it turns highly aggressive with a strong tendency for invasion of surrounding tissues and metastases to distant sites. The prognosis for survival is generally poor with a 5-year disease-free survival rate reported to be < 30%.

Conflict of interest

Authors have declared that there is no conflict of interests.

Reference

1. Kadir Balaban, Murat Şedele, Alper Sayiner, Malignant nodular hidradenoma of the scalp: A case report. J Clin Anal Med. 2017; 8(suppl 3):240-1.
2. Sinhasan SP, Harthimath BC, Sylvia MT, Bhat RV. "Metastatic malignant nodular hidradenoma": A rare case report with review of literature. Clin Cancer Investig J. 2015; 4:651-4.
3. Tanmoy M, Sampath S, Bhagavatula ID, Asha U, Dhaval S. Malignant nodular hidradenoma of scalp. J Neurosci Rural Pract. 2014; 5(4):423-5.
4. Weedon D. Tumors of cutaneous appendages. In: Weedon D, editor. Weedon's skin pathology. 3rd ed. Edinburgh: Churchill Livingstone Elsevier, 2010, p.758-807.
5. Abhishek S, Nupur B, Vivek K, Ashok KC. Current management approach to hidradenocarcinoma: a comprehensive review of the literature. Ecancer medical science, 2015; 9:517.
6. Giorgini E, Tugnoli G, Aprile S, Collina G, Villani S, Biscardi A. *et al*. Malignant nodular hidradenocarcinoma arising on the areola of a male patient: Case report of an "orphan disease" and review of the literature. J Carcinog Mutagen. 2012; 3:129-33.
7. Nair PS. A clinicopathologic study of skin appendageal tumors. Indian J Dermatol Venereol Leprol, 2008; 74:550.
8. Ohta M, Hiramoto M, Fujii M, Togo T. Nodular hidradenocarcinoma on the scalp of a young woman: Case report and review of literature. Dermatol Surg. 2004; 30:1265-8.
9. Klein W, Chan E, Seykora JT. Tumors of the epidermal appendages. In: Elder DE, Elenitsas R, Johnson BL, Murphy GF, editors. Lever's histopathology of the skin. 9th ed. Philadelphia: Lippincott Williams & Wilkins, 2005, p.867-926.
10. Tanmoy M, Sampath S, Bhagavatula ID, Asha U, Dhaval S. Malignant nodular hidradenoma of scalp. J Neurosci Rural Pract. 2014; 5(4):423-5.
11. Weedon D. Tumors of cutaneous appendages. In: Weedon D, editor. Weedon's skin pathology. 3rd ed. Edinburgh: Churchill Livingstone Elsevier, 2010, p.758-807.
12. Ko CJ, Cochran AJ, Eng W, Binder SW. Hidradenocarcinoma: A histological and immunohistochemical study. J Cutan Pathol. 2006; 33:726-30.
13. Tolkachjov SN, Hocker TL, Hochwalt PC, Camilleri MJ, Arpey CJ, Brewer JD. *et al*. Mohs micrographic surgery for the treatment of hidradenocarcinoma the Mayo Clinic experience from 1993 to 2013. Dermatol Surg. 2015; 41(2):226-31.
14. Delgado R, Kraus D, Coit DG, Busam KJ. Sentinel lymph node analysis in patients with sweat gland carcinoma. Cancer. 2003; 97(9):2279-84.
15. Harari PM, Shimm DS, Bangert JL, Cassady JR. The role of radiotherapy in the treatment of malignant sweat gland neoplasms. Cancer. 1990; 65(8):1737-40.
16. Nash JW, Barrett TL, Kies M, Ross MI, Sneige N, Diwan AH. *et al*. Metastatic hidradenocarcinoma with demonstration of Her-2/neu gene amplification by fluorescence in situ hybridization potential treatment implications. J Captan Pathol. 2007; 34(1):49-54.
17. Görtler I, Köppl H, Stark GB, Horch RE. Metastatic malignant acrospiroma of the hand. Eur J Surg Oncol. 2001; 27(4):431-5.
18. Garcia-Bonafe MM, Campins MM, Redecilla PH. Malignant nodular hidradenoma on the scalp: Report of a case with fine needle aspiration cytology features and histologic correlation. Acta Cytol. 2009; 53:576-80.
19. Souvatzidis P, Sbano P, Mandato F, Fimiani M, Castelli A. Malignant nodular hidradenoma of the skin: Report of seven cases. J Eur Acad Dermatol Venereol. 2008; 22:549-54.