Giant metastasizing malignant hidradenoma in a child

Sunil K Bajaj, Ritu Misra, Rohini Gupta, Anju Bansal[1]

Department of Radio Diagnosis, Safdarjung Hospital and Vardhman Mahavir Medical College, [1]Department of Pathology, National Institute of Pathology, Indian Council of Medical Research Laboratory, Safdarjung Hospital Campus, New Delhi, India

ABSTRACT

An 8-year-old girl presented with a scalp swelling. The swelling was recurrent, reappearing everytime after local excision. She underwent surgery and the histopathologic diagnosis was malignant hidradenoma. This very rare and aggressive tumor is known to occur in elderly population and is histopathologically distinct from its commonly occuring benign counterpart**.** Malignant hidradenoma is resistant to chemotherapy and radiotherapy. We empahsize that being cognizant of the possibility of this rare tumor would assist in timely action in the form of wide resection, with possible reduction in morbidity and mortality.

Key words: Clear cell eccrine carcinoma, hidradenoma, malignant nodular hidradenoma, sweat gland carcinoma

Access this article online

Website: www.idoj.in

DOI: 10.4103/2229-5178.190492

Quick Response Code:

Address for correspondence:
Dr. Sunil K. Bajaj, Department of Radio Diagnosis, Safdarjung Hospital and Vardhman Mahavir Medical College, New Delhi - 110 029, India.
E-mail: drskbajaj@rediffmail.com

INTRODUCTION

This is the first reported case of metastasizing nodular hidradenoma in a child in the medical literature. Less than five cases of hidradenoma have been reported in children, none of which were malignant.

Benign hidradenoma isa common tumor of sweat glands, which runs a favourable course. Its malignant counterpart is quite rare; with only 50 cases reported in adults, and none in children. They are aggressive in nature with poor prognosis.

Early diagnosis and wide surgical resection are the mainstay in the management of this malignant tumor, which is not responsive to chemotherapy and radiotherapy (RT). The prognosis is nevertheless poor, with recurrence and fatal outcome.

CASE REPORT

An 8-year-old girl presented to a district hospital in November 2008 with a painless lemon-sized scalp swelling since three months. It was flesh colored with intact overlying skin, and was excised. The mass recurred and was re-excised 11 months later in October 2009. She presented a year later to our referral hospital with another recurrence at the margins. Histopathology records of the previous excisions were not available. She underwent excision of the lesion at our hospital in November 2010 and the histopathology of the excised specimen revealed recurrent hidradenoma of scalp. In August 2011, she had recurrence yet again and underwent excision of lesion with wide margins. The pathology of the specimen showed evidence of bilateral high parietal bone erosion, osseous infiltration, and extradural extension. Histopathology of excised scalp tumor with periosteum revealed skin-covered tumor mass composed of several nodules of tumor cells, which had eosinophilic to clear cytoplasm with round to ovoid nuclei. Cytoplasmic vacuoles representing intracellular lumina formation, an important histopathologic feature, was observed. Mitotic figures and foci of vascular invasion were noted. The tumor tissue was infiltrating the periosteum and reaching up to the resected margins [Figures 1 and 2]. Hence, diagnosis was changed to malignant hidradenoma.

Cite this article as: Bajaj SK, Misra R, Gupta R, Bansal A. Giant metastasizing malignant hidradenoma in a child. Indian Dermatol Online J 2016;7:410-3.

Craniotomy of bilateral parietal bones was done after the final diagnosis of malignancy.

The patient presented with recurrence yet again in 2012 [Figure 3]. The flesh-colored, ulcerated scalp mass had grown to a larger size measuring approximately 13 × 10 × 12 cm. Computed tomography and magnetic resonance imaging done between June and August 2012 revealed a large heterogeneously enhancing mass in the left parieto-occipital region with epidural component and invasion of left transverse sinus [Figure 4]. There were distant metastases into spine, sacrum, both humerii and lungs. Her tumor was staged as stage IV.

Chemotherapy was started with 5Fluorouracil (5FU) and cisplatin, but was discontinued after two cycles due to poor response. She also received 20 Gy of RT in 5 sequences in September 2012 with no visible improvement. The patient was advised palliative care in view of extensive disease and deteriorating condition. Her parents decided to take her back to their native village and never returned for follow-up thereafter.

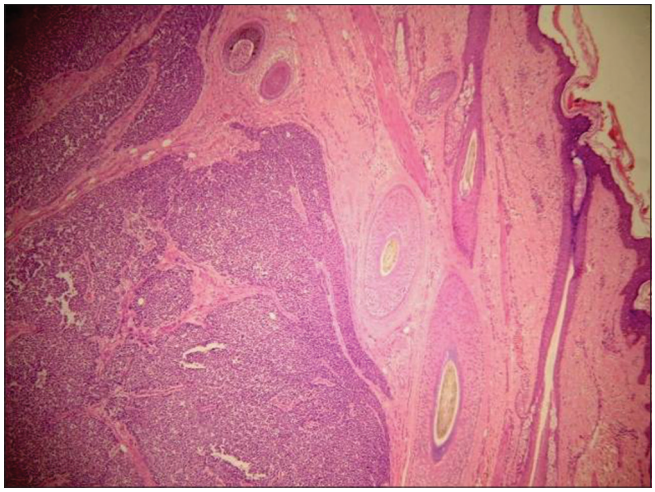

Figure 1: Microphotograph showing multiple lobules of tumor cells seen in the dermis with intact epidermis (H and E ×40)

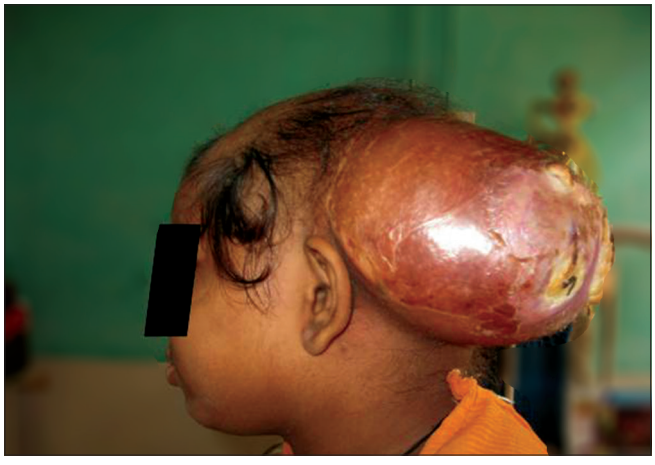

Figure 3: Clinical photograph of the patient

DISCUSSION

Malignant nodular hidradenoma (MNH) is an extremely rare (0.005% among all epithelial neoplasms of skin[1]) and little understood tumor of sweat glands. To date, reported cases in the medical literature number just over 50,[2] mostly as case reports or isolated small series. In 1865, Cornil[3] documented the first case of sweat gland carcinoma. Reviewing the literature, Gates *et al*.[4] assigned a total of eight cases to the nineteenth century. Hidradenoma was first described by Keasbey and Hadley in 1954.[5] Later Stout and Cooley[6] reported a series of 11 well-documented cases of malignant adnexal tumors. In 1968, Berg and McDivitt[7] presented the largest series to date, of 17 cases of hidradenoma among 102 sweat gland carcinomas and were the first to offer classification of this entity. The cell of origin of this tumor is still debatable, whether from eccrine cells, apocrine cells, or folliculosebaceous apocrine unit (fsau) cells. It is increasingly being recognized that most of these tumors have cells from any or all the three units.[8]

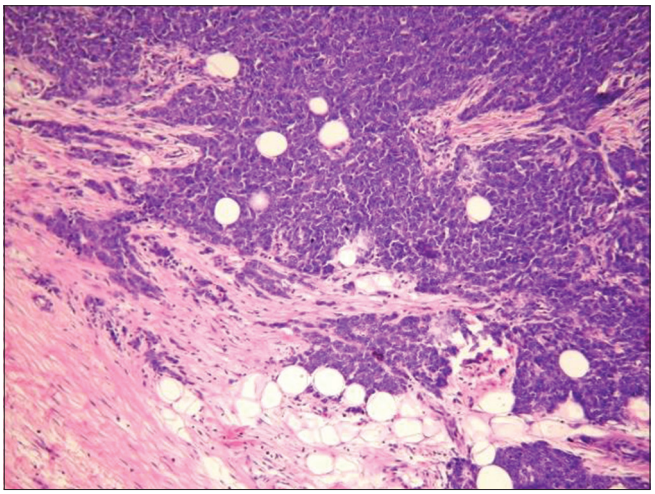

Figure 2: Microphotograph showing cellular tumor with infiltrative borders (H and E ×100)

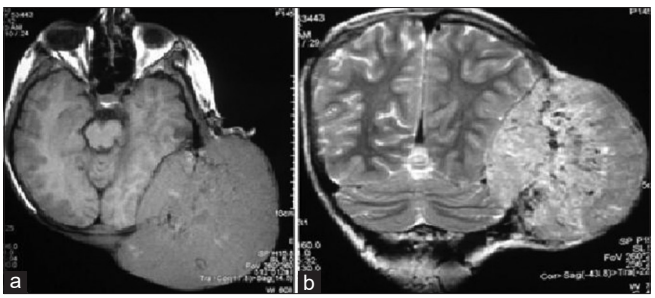

Figure 4: (a) Axial T1W image showing large soft tissue mass in the scalp causing erosion of left parieto-occipital bones and invasion into the extradural space with buckling of white matter. There are few scattered T1 bright hemorrhagic foci within the mass. (b) Coronal T2W image showing multiple radiating flow voids within the mass

There is lack of consensus on the terminology of this condition. These tumors have been variously named as clear cell eccrine carcinoma, nodular hidradenoma, malignant clear cell hidradenoma, solid cystic adenocarcinoma, malignant acrospiroma, and clear cell myoepithelium tumor.

A major predicament faced by the clinicians is the inability to distinguish the malignant variety from its benign counterpart, hidradenoma—a frequently occurring adnexal tumor with favorable prognosis. In contrast, malignant hidradenoma, especially the metastasizing type is extremely aggressive with a very poor clinical outcome.

The distinction is based on lack of circumscription; infiltrative manner of spread; nuclear pleomorphism; perineural and lymphovascular invasion; and increased mitotic numbers as well as hematogenous spread to distant nodes, other organs, bones, and lungs.

Immunochemistry studies have been attempted, but because of paucity of cases, no definite data is yet available. Ki67 and PHH_3 may have a role to play in the diagnosis and differentiation of benign, atypical, and malignant hidradenomas. However, histopathologists are unable to give a conclusive answer. This distinction is of importance for prognostic evaluation, particularly in cases of atypical and malignant variety. Atypical hidradenoma has lower Ki-67 values compared with metastasizing adnexal carcinoma and malignant hidradenoma (*P* value ≤ 0.002). Similarly, PHH_3 is lower in atypical hidradenoma than in malignant hidradenoma. A Ki-67 value > 11% and PHH_3 > 0.7% is likely to be present in malignant rather than atypical hidradenoma.[2] A few other immunostaining markers such as C5/6, CK15, EMA, and p-53 have been found positive in malignant cutaneous adnexal neoplasms and apocrine carcinomas.

The MNH has been reported in elderly people in 6th to 7th decade.[9,10] Metastasizing malignant hidradenoma has not been reported in childhood till date, although three cases of benign hidradenoma in children aged 1, 3, and 13 years have been documented. This patient happens to be the first such case that appeared at the age of 8 years. The tumor recurred every time after surgical excision (5 times) and was so aggressive that it spread to extradural space, vertebrae, humerii, and lungs in a span of less than 4 years. Attempts to arrest its spread by chemotherapy and radiotherapy were unsuccessful.

Hidradenoma typically presents as a slow-growing painless, firm, subcutaneous nodule or plaque. It may be flesh colored, violet, or reddish with intact normal overlying skin; occasionally showing surface ulceration.

More often than not, the diagnosis is based on histopathologic features alone. Malignant hidradenomas have mostly been reported to arise de novo, although a few cases have arisen in pre-existing benign hidradenoma.[11]

Involvement of regional and distant nodes is a known feature in metastatic variety. Metastases to bones, lungs, and other viscera may occur in as high as 60% of patients.[12]

Pathologic features

MNHs are usually large, poorly demarcated, and asymmetric as compared with the typically well-demarcated benign variety. However, definitive histologic features, that distinguish hidradenomas from its benign counterpart or other related malignant tumors are lacking, and diagnosis is based on the extent of invasion. Tumor necrosis, areas of high cellularity, and focal or diffuse areas of marked cytologic atypia are present in some cases. There may be angiolymphatic invasion. Additionally, in some cases, nuclear anaplasia may be minimal to moderate and at times even absent in both the primary and the metastases.[13]

Standard treatment of hidradenoma is wide surgical excision that extends 2 cm beyond tumor margins.[14] Of late, wider excision extending to 5 cm beyond margins has been recommended to reduce incidence of residual cells. Local recurrence rate is 10%–50%.[15]

Where margins are reported positive, adjuvant therapy has been tried. The tumor has largely been reported to be radioinsensitive. Various chemotherapy regimes have been tried using 5FU, cisplatin, and doxorubicin. Oral 5FU (capecitabine) results have been encouraging in a few cases. Herceptin has been tried as targeted therapy in HER1- and HER2-positive tumors. Use of Mohs microsurgery has also been suggested.[16]

Since no uniform treatment guidelines for MNH are established and adjuvant therapies have also not been very effective, targeted therapy using molecular biology techniques might be the answer for this rare but aggressive tumor.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Guerrissi JO, Quiroga JP. Adnexal carcinomas of the head and neck. Indian J Plast Surg 2008;41:229-34.
2. Nazarian RM, Kapur P, Rakheja D, Piris A, Duncan LM, Mihm MC Jr, *et al*. Atypical and malignant hidradenomas: A histological and immunohistochemical study. Mod Pathol 2009;22:600-10.
3. Smith CC. Metastasizing carcinoma of the sweat-glands. Br J Surg 1955;43:80-4.
4. Gates O, Warren S, Warvi WN. Tumors of sweat glands. Am J Pathol 1943;19:591-631.

5. Keasbey LE, Hadley GG. Clear-cell hidradenoma; report of three cases with widespread metastases. Cancer 1954;7:934-52.
6. Stout AP, Cooley SG. Carcinoma of sweat glands. Cancer 1951;4:521-36.
7. Berg J, McDivitt RW. Pathology of sweat gland carcinoma. Pathol Annu 1968;3:123-44.
8. Gauerke S, Driscoll JJ. Hidradenomas: A brief review and future directions. Arch Pathol Lab Med 2010;134:781-5.
9. Hall J, Knee G, A'Hern RP, Clarke J, Glees JP, Ford HT, *et al*. Sweat-gland tumors: A clinical review of case in one centre over 20 years. Clin Oncol (R Coll Radiol) 2006;18:351-9.
10. Oh SH, Kim CH, Hwang E. Giant eccrine hidradenoma mimicking a malignant tumor of the salivary gland. Arch Plast Surg 2014;41:437-9.
11. Hande AH, Sonone AM, Chaudhary MS. Clear cell eccrine carcinoma with comedonecrosis in neck region: A rare case report and review of literature. Pathol Lab Med Int 2010;2:121-6.
12. Mehregan AH, Hashimoto K, Rahbari H. Eccrine adenocarcinoma. A clinicopathologic study of 35 cases. Arch Dermatol 1983;119:104-14.
13. Stromberg BV, Thorne S, Dimino-Emme L, Katz DA, Rouse JW. Malignant clear cell hidradenoma: A case report and literature review. Nebr Med J 1991;76:166-70.
14. Wong TY, Suster S, Nogita T, Duncan LM, Dickersin RG, Mihm MC Jr. Clear cell eccrine carcinomas of the skin. A clinicopathologic study of nine patients. Cancer 1994;73:1631-43.
15. Ashley I, Smith-Reed M, Chernys A. Sweat gland carcinoma. Case report and review of the literature. Dermatol Surg 1997;23:129-33.
16. Wildemore JK, Lee JB, Humphreys TR. Mohs surgery for malignant eccrine neoplasms. Dermatol Surg 2004;30:1574-9.