Case Report

Microcystic Adnexal Carcinoma of Posterior Neck: A Case Report

Mahjabin Taklif[1], Maryam Abolhasani[1]

1. Department of Pathology, Hazrat-e-Fatemeh Hospital, Iran University of Medical Siences, Tehran.

ABSTRACT

Background: **Microcystic adnexal tumor is a rare sclerosing variant of ductal carcinoma of eccrine sweat glands which is highly invasive. This tumor is often misdiagnosed as other benign or malignant skin lesions and improper treatment is carried on and is associated with high recurrence rate.**

Case presentation: **We reported here in a 59-years old man who underwent incisional biopsy for a congenital lesion on posterior neck which had grown recently. Microscopic examination exhibited an infiltrative tumor as constituted by small cord-like and angulated tubules with tadpole or comma-like shapes, individually set in abundant fibrous stroma in dermis. So, the diagnosis was syringoma. In the next step, the lesion underwent excisional biopsy. Histologically, a tumor located in dermis with extension to subcutis was noted which contained basaloid keratinocytes with occasional horn cysts and abortive hair follicles. In other areas, ducts and gland-like structures lined by two-cell layers predominated. The tumor extended to skeletal muscle and perineurial structures but no significant atypia or mitosis was identified. Eventually, with respect to mentioned features, the diagnosis was microcystic adnexal tumor.**

Conclusion: **It is concluded that thinking about this rare invasive skin tumor with proper use of Mohs' surgery and its correct diagnosis is clinically of high significance to reduce its recurrence rate. Meanwhile, this tumor was noticeably set in a congenital lesion in this reported case.**

Key words: **Syringoma, Adnexal, Microcystic, Mohs' surgery**

Introduction

Syringoma represents as adenoma of intraepidermal eccrine ducts which occurs predominantly in women at puberty or later in life (1, 2). Clinically, it represents as multiple skin-colored or slightly yellowish soft papules with a diameter of 1-2 mm. These papules are commonly found in face and neck. However, in many cases they are limited to lower eyelids (3). Other sites of predilection are cheek, axilla, abdominal wall, and vulva. Histologically, embedded in fibrous stroma are numerous small ducts, the walls of which are lined usually by two rows of epithelial cells (4,5). In most instances, these cells are flat. Occasionally, the cells of the inner row appear vacuolated. The lumina of ducts contain amorphous debris and

Received: 2 January 2006
Accepted: 10 June 2006
Address communications to:Dr. Mahjabin Taklif, Hazrat-e-Fatemeh Hospital, Iran University of Medical Siences,Tehran-IRAN.
Email: robab20002000@yahoo.com

some are comma-like shaped (1, 2).

On the other hand, microcystic adnexal tumor usually presents clinically as skin nodules. It is usually seen in the skin of head and neck. It may best be considered as a sclerosing variant of ductal eccrine carcinoma (1,2). The tumor is a poorly circumscribed dermal tumor that may extend into the subcutis and skeletal muscle. Continuity with the epidermis or follicular epithelium may also be seen. Two components within a desmoplastic stroma may be evident. In some areas, basaloid keratinocytes are seen, containing horn cysts and abortive hair follicles. In other areas, ducts and gland-like structures lined by a two-cell layer predominate. The tumor islands typically reduce in size as the tumor extends deeper into the dermis. The cells may have clear cytoplasm or sebaceous differentiation. Cytologically, the cells are bland without significant atypia, mitosis is rare or absent. Perineurial invasion may be present, a feature that may account for its high recurrence rate (1,2).

Case report

Here we report a skin adnexal lesion with rare manifestations. A 59-years old man referred to us, complaining of a congenital soft tissue lesion on posterior neck which had recently grown. Multiple nodules were seen in gross examination and some of them were ulcerative (Figure1). The general condition of patient was good, blood pressure and laboratory examinations were normal, and the patient had O+ blood group. The patient mentioned no history of previous disease or radiation therapy.

In the first step, the patient underwent superficial incisional biopsy and the lesion was sent for pathological analysis. Macroscopic examination revealed an ovaloid creamy tissue (2 x 1 x 0.5 cm) with a nodular lesion at center (1 x 0.7 x 0.5 cm). Cut section of lesion was creamy brown rubbery. Microscopic examination revealed an infiltrative tumor as constituted by small ducts and tubules. The tubules were small cord-like and angulated, lined by a two-layered epithelium. Ducts were variably sized and shaped. Some of them were comma-shaped or tade pole, the others were dilated and filled with eosinophilic materials. The stroma was fibrotic. High power microscopic examination displayed no atypia or mitosis, so it was diagnosed to be syringoma (Figure 2). According to the large size of the lesion for being syringoma, excisional biopsy was recommended.

In the next step, the whole lesion was excised and a thick skin graft was performed. Macroscopic examination showed an ovaloid piece of skin (8.5 x 6 x 2 cm). Some nodular lesions were noted on surface (5 x 3 x 2 cm). Cut section was non-homogenous and creamy brown. This time, low power microscopic examination revealed basaloid keratinocytes, horn cysts, and abortive hair follicles extending into skeletal muscles and nerves. In some foci, the lesion was in continuity with the epidermis. At high power examination, no atypia or mitosis was seen. All margins were involved. Regarding microscopic findings and references, it was proved to be microcystic adnexal carcinoma (Figure 3) and wider excision for preventing local recurrence was recommended.

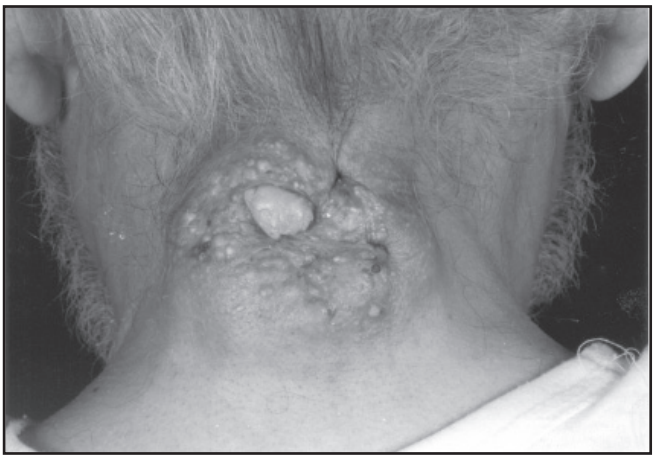

Figure 1 : Multiple ulcerative nodules

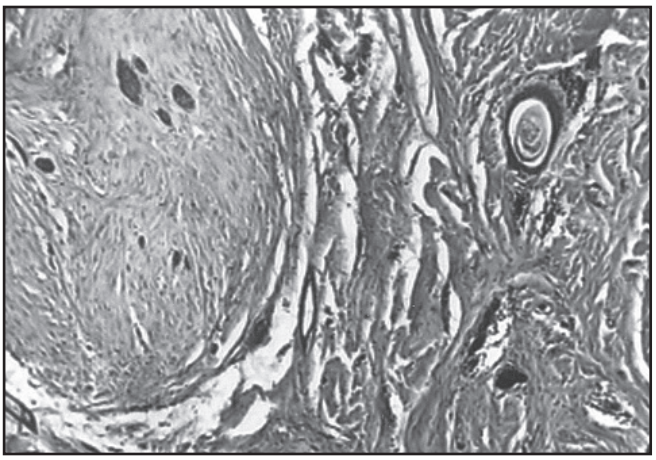

Figure 2 : First biopsy, Variably sized and shaped ducts.comma-shaped or tade pole(LPF)

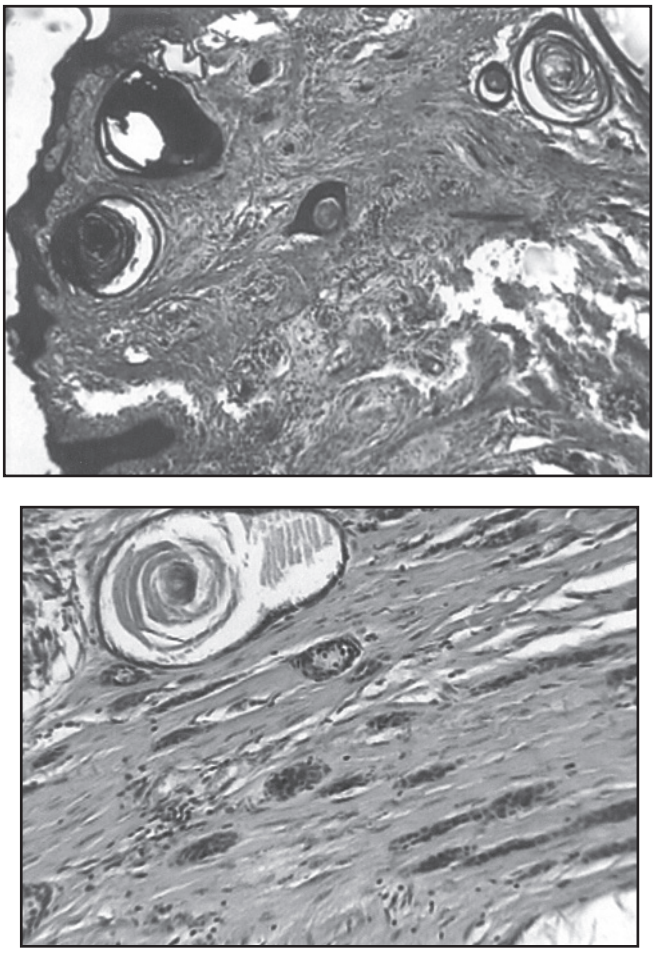

Figure 3: Exisional biopsy, basaloid keratinocytes, horn cysts, and abortive hair follicles (LPF & HPF)

Discussion

The mentioned case differed from syringoma in two respects. First, syringoma usually occurs after puberty, while this case was set on a congenital lesion. The other point was that syringoma is 1-2 mm in size, while the size of mentioned lesion was very large. These contrasts lead us to investigate further (6). In reviewed literature, two cases of syringoma with abnormal and giant size had been reported in scrotum and axilla, but both were chondroid in nature (7,8,9) and usual syringoma of this size has not been reported yet. After resection of the whole lesion and microscopic evaluation, findings such as giant size, extension to subcutis, muscle and perineurial structures led us to suspect malignancy. In all examined sections, no atypia or mitosis was identified and all above-mentioned criteria resulted in the diagnosis of microcystic adnexal carcinoma (1,10, 2).

Microcystic adnexal tumor may best be considered as a sclerosing variant of ductal eccrine carcinoma. It is usually seen in the skin of upper lid, but can occasionally be seen on the chin, nasolabial fold, or cheek (1). Most of the affected patients are at their fourth and fifth decades of life (11). The tumor frequently recurs locally but usually do not metastasize, however, few cases of lung and lymph node metastases have also been reported (12,13,14). This tumor is deeply invasive with occasional infiltration of perichondrium, periosteum, and/or perineurium (15). It often extends beyond the clinical margins with local spreading in the dermal, subcutaneous, and perineural tissue planes. It has a high local recurrence rate after local standard excision. This tumor which was first reported to be a distinct skin tumor entity by Goldstein and colleagues in 1982 (15,10) and 276 cases of which thus far reported (13, 14, 17), is often misdiagnosed (14-17). There are also reports that this tumor was first diagnosed as squamous cell carcinoma, basal cell carcinoma (16-17), desmoplastic trichoepithelioma (15), and even rhinophymaoma (18). This tumor should be considered in the differential diagnosis of slowly-growing tumors in the head and neck region (15), particularly when the tumor extends to the base of biopsy in a syringomatous or trichoepitheliomatous-like proliferation (1). Some of the patients have had a history of radiation treatment (12-14, 19) and one case has been reported to have an immunodeficiency syndrome (11). In one study, quantitative DNA analysis revealed a tumor with a diploid stem line and only few aneuploid cells (18). Immunoperoxidase staining for carcinoembrionic antigen stains the glandular but not the pilar structures (1). Local standard excision and Mohs micrographic surgery are two choice treatment of this tumor. Local standard excision has lead to high recurrence rate up to 85% but the 5-year recurrence rate of Mohs micrographic surgery was low and about 5%. In this regard, Mohs' micrographic surgery is a surgical technique that seeks to ensure the clearance of cutaneous tumors while maximizing normal tissue conservation. This is accomplished through the sequential removal of thin layers of tissue in which the entire peripheral and deep margins are examined for

residual tumor (16, 20-22). This procedure was once carried out for the treatment of squamous cell and basal cell carcinomas but now is practiced in the management of other skin tumors, such as microcystic adnexal carcinoma, melanoma, Merkel cell carcinoma, dermatofibrosarcoma protuberans, and extramammary Paget's disease (22). Long-term follow up of patients with imaging studies has been advocated (14).

If this tumor is diagnosed too late, it can be inoperable because of its infiltrative growth (15). When extirpation entails sufficiently large morbidity, given the low rate of metastases and mortality, observation is a reasonable alternative (13). Another large-sized case of microcystic adnexal tumor of face was reported in literature (13) and one case arised from a previous facial lesion in a patient with history of radiation treatment for acne after fifteen years (19). This would be an interesting point that none of the reported cases had already mentioned this lesion set in a congenital lesion.

References

1- Elder D, Elenitsas R, Christine Jaworsky C, Johnson B Jr.Lever's Histopathology of Skin. 9th ed. Philadelphia: Lippincott- raven;2005.

2- Weedon D. Skin Pathology. 2th ed. London:Churchill livingstone;2004.

3- Rosai J.Ackerman's Surgical Pathology. 9th ed. Edinburgh:Mosby; 2004.

4- Kumar V, Abbas AK, Fausto N.Pathologic Basis of Disease. 7th ed. Philadelphia: W. B. Saunders Company; 2005.

5- Mills SE, Carter D,. Greenson JK, Oberman HA., Reuter V, Stoler MH. Sternberg's Diagnostic Surgical Pathology. 4th ed. Philadelphia: Lippincott Williams & Wilkins; 2004.

6- Glatt HJ, Proia AD, Tsoy EA et al. Malignant Syringoma of the Eyelid. Ophthalmol 1984 Aug, 91(8): 987-90.

7- Chillon DF, Martin NM, Mora NC, et al, Giant Chondroid Syringoma of the Scrotum. First Reported Case. Arch Esp Urol 2001 Dec, 54(10): 1121-3.

8- Hardisson D, Linares MD, Nistal M . Giant Chondroid Syringoma of Axilla. J Cutan Med Surg 1998, 3(2): 115-7.

9- Sliwa Hahnle K, Obers V, Lakhoo M, Saadia R. Chondroid Syringoma of the Abdominal Wall. A Case Report and Review of the Literature. S Afr J Surg, 1996 Feb, 34(1): 46-8.

10- Goldstein DJ, Barr RJ, Santa Cruz DJ. Microcystic Adnexal Carcinoma, A Distinct Clinicopathologic Entity. Cancer566:(3)50 1982 -72.

11- Lei JY, Wang Y, Jaffe ES, et al. Microcystic adnexal carcinoma associated with primary immunodeficiency, recurrent diffuse herpes simplex virus infection, and cutaneous T-cell lymphoma. Am J Dermatopathol 2000 22(6):524-9.

12- Ban M, Sugie S, Kamiya H, Kitajima Y. Microcystic adnexal carcinoma with lymph node metastasis. Dermatology 2003 207(4):395-7.

13- Eisen DB, Zloty D. Microcystic adnexal carcinoma involving a large portion of the face: when is surgery not reasonable? Dermatol Surg 2005 1472-7, discussion1478.

14-Gabillot-CarreM,WeillF,MamelleG,etal.Microcystic adnexal carcinoma: report of seven cases including one with lung metastasis. Dermatology 2006 212(3):221-8.

15- Fischer S, Breuninger H, Metzler G, Hoffmann J. Microcystic adnexal carcinoma: an often misdiagnosed, locally aggressive growing skin tumor. J Craniofac Surg 2005 16(1):53-8.

16- Leibovitch I, Huilgol SC, Selva D, Lun K, Richards S, Paver R. Microcystic adnexal carcinoma: treatment with Mohs micrographic surgery. J Am Acad Dermatol 2005 Feb, 52(2):295-300.

17- Leibovitch I, Huilgol SC, Richards S, Paver R, Selva D. Periocular microcystic adnexal carcinoma: management and outcome with Mohs' micrographic surgery. Ophthalmologica 2006 220(2):109-13.

18- Bewer F, Forster C, Welkoborsky HJ. Microcystic adnexal carcinoma (malignant syringoma) of the nose: case report and review of the literature. Laryngorhinootologie 2004 83(2):113-6.

19- Delshad E, Ratner D. Microcystic adnexal carcinoma. Skin med 2004 Nov-Dec, 3(6):341-3.

20- Khachemoune A, Olbricht SM, Johnson DS. Microcystic adnexal carcinoma: report of four cases treated with Mohs' micrographic surgical technique. Int J Dermatol 2005 44(6):507-12.

21-Leibovitch I, Huilgol SC, Selva D, Paver R, Richards S. Cutaneous lip tumours treated with Mohs micrographic surgery: clinical features and surgical outcome. Br J Dermatol 2005 153(6):1147-52.

22- Pennington BE, Leffell DJ. Mohs micrographic surgery: established uses and emerging trends Oncology (Williston Park) 2005 19(9):1165-71; discussion 1171-2, 1175.