Malignant chondroid syringoma in a West African cancer institute: A case report

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1. Introduction

Malignant chondroid syringoma (MCS) is a tumor of sweat glands that preferentially interested ends [1]. Unlike chondroid syringoma which is a rare benign tumor of the skin appendages with very slow growing [2], MCS, also called malignant mixed tumor of the skin, is extremely rare [3] with poor prognosis. We report a case of malignant chondroid syringoma of the axillary region in our practice at Joliot Curie Cancer Center in Dakar.

2. Case presentation

It was about a 53 year old patient who consulted for a mass of the right deltoid area. Eighteen months before he noticed a painless nodule of the right shoulder which has risen steadily in volume with the appearance of axillary nodule. The examination revealed a 10 cm mass at the right deltoid region with edema and skin involvement (Fig. S1). MRI of the right shoulder objectified a necrotic tumor at the lower back part of the deltoid and triceps infiltrating the invasive axillary region with axillary malignant lymph node. Microscopic examination showed in the deltoid lesion, dermis covered by normal skin and occupied by a dense fibro-collagen tissue, and few inflammatory, polymorphic, predominantly perivascular cells making a scar. At the axillary mass initially considered as a lymph node, microscopic examination revealed tumor fragments without lymphoid tissue. It showed extended necrosis, bays and cords with cells sometimes small, compact, round nuclei, dense chromatin, to scanty cytoplasm, poorly defined, sometimes trabecular with larger cells, eosinophilic cytoplasm fairly abundant, forming acinar and alveolar provisions. Sometimes they took a signet ring appearance in vacuolated cytoplasm, or plasmacytoid eosinophils. Spans and cords were separated by a myxoid stroma with many stellate cells, myoepithelial types with slightly irregular nuclei. Moreover, there was many peri-nervous and muscle tissue infiltration. Some isolated cells were PAS positive. In summary there was a biphasic tumor tissue cell, epithelial and myoepithelial evolving carcinoma or malignant mixed tumor. At Immunohistochemistry, the epithelial cells expressed cytokeratin gaits AE1/AE3, PS100 and P63. The spindle-shaped and stellate cells expressed actin and rarely PS100. All in favor of a malignant chondroid syringoma. As the patient was opposed to a radical surgical treatment, an adryamyacin-cisplatin based chemotherapy was instituted. The evolution was marked by growth with increased volume of the lesions that had become painful. Death occurred 8 months after diagnosis.

3. Discussion

MCS was first described by Hirsch and Helwig in 1961 [4]. The term of “atypical mixed tumor of the skin” is used to describe tumors characterized by histological stigma of malignancy, recurrence, local invasion and satellite tumor nodules without proven metastases [5].
Epidemiologically, malignant chondroid syringomas have a female predilection while their benign counterparts are more common in men. Clinically, they are characterized by a faster growth and a size significantly greater than that of benign mixed tumors [6]. There is no evidence of a specific risk factor. Trauma has been suggested as a possible predisposing factor for the development of both malignant and benign chondroid syringoma [1].

Unlike the benign variety that preferentially seat at the head and neck, malignant chondroid syringoma seat preferentially in extremities and trunk [1,5].

Malignant chondroid syringoma can appear de novo or more rarely be developed from a cartilaginous syringoma [2]. In fact, rarely however, a chondroid syringoma after many years of evolution, can undergo malignant transformation and be metastatic.

The pathological diagnosis of adnexal tumors of the skin is difficult. It is made on material of fine needle aspiration, biopsy or surgery [6]. It is related firstly to the wide variety of tumors and their variants and on the other hand to the differentiation of adnexal lines of frequency in the same tumor [3]. The chondroid syringomas are included in myoepithelial skin tumors. The chondroid syringoma has an epithelial component with eccrine or apocrine differentiation, and myoepithelial component of varying importance with stellate myoepithelial cells or plasmacytoid chondro-myxoid in a matrix [1,5]. These characteristics were found during pathological examination of our patient. The immunostaining showed a positivity of epithelial markers (cytokeratin), epithelial structures, vimentin, PS100, actin and calponin for both smooth muscle and myoepithelial cells. It was a significant contribution for our patient. Basal cells and adenoid cystic carcinomas seem to be strong differential diagnosis [7].

There is no evidence of the effectiveness of chemotherapy and radiotherapy in the treatment of SCM. Some authors used chemotherapy based on 5-FU, prednisone and Cytoxan and radiotherapy [5]. Early wide excision and margins appears to be the most reliable treatment [2]. For other authors adjuvant radiotherapy is useful to decrease recurrence [8]. Chemotherapy made in the case of our patient, based on adryamycin and cisplatin, did not prevent progression and death.

The clinical course of MCS is deemed unpredictable. The spread can be lymphatic or blood-borne [5]. The two most common sites for distant metastases are lungs and bones [9].

4. Conclusion

The MCS is an extremely rare malignant tumor of sweat glands. Pathological examination is the key of diagnosis, showing an epithelial component, eccrine or apocrine differentiation and myoepithelial component of varying importance. The immunostaining is very helpful because there is a positivity for epithelial and myoepithelial markers. For the treatment, an early and large excision with clear margins is the most reliable treatment. Chemotherapy and radiotherapy are not effective. The prognosis depends on early diagnosis and complete resection and remains poor.

Conflict of interest
None declared.

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Ethical approval
Because of the case report we did not specifically required ethics committee.

Consent
Written informed consent was obtained from the patient’s legal.

Author contribution
Ka and Gnangnon conceived this presentation while Diouf, Dieng, Gaye, Thiam and Dem participated in quality control of this manuscript. All authors read and approved the final manuscript.

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Appendix A. Supplementary data
Supplementary data associated with this article can be found, in the online version, at http://dx.doi.org/10.1016/j.ijscr.2016.06.029.

References
[1] S. Vohra, W.A. Bates, S.L. Baithun, A rare adenaxial tumor of the hallux: malignant chondroid syringoma, Foot 6 (1996) 175–177.
[2] M. Varsori, S. Dettwiler, K. Chaloupka, Syringome chondroide de la paupière: à propos d’un cas, J. Fr. Ophtalmol. 11 (2001) 80–82.
[3] P. Shashikala, H.R. Chandrashekhar, S. Sharma, K.K. Suresh, Malignant chondroid syringoma, Indian J. Dermatol. Venereol. Leprol. 70 (2004) 175–176.
[4] D. Tural, F. Selkubiricik, F. Gunvar, A. Karismaz, S. Serdengecti, Facial localization of malignant chondroid syringoma: a rare case report, Case Rep. Oncol. Med. (2013) 907980.
[5] J.A. Kamarashev, S. Kaddu, et al., Non melanoma skin cancers: appendageal tumors, in: Reinhard Dummer (Ed.), Skin Cancer – A Worldwide Perspective, Springer, Berlin, 2010, pp. 121–176.
[6] W. Agrawal, R.L. Gupta, S. Kumar, K. Mishra, Malignant chondroid syringoma, J. Dermatol. 25 (8) (1998) 547–549.
[7] C. Requena, S. Brotons, O. Sanmartin, B. Llombart, V. Traves, C. Guilen, L. Requena, Malignant syringoma of the face with bone invasion, Am. J. Dermatopathol. 35 (5) (2013) 395–398.
[8] J.J. Hong, J.F. Elmore, C.J. Drachenber, M.C. Jacobs, O.M. Salazar, Role of radiation therapy in the management of malignant chondroid syringoma, Dermatol. Surg. 21 (9) (1995) 781–785.
[9] G. Hermann, D. Moss, K.I. Norton, M.E. Guttenberg, Case report 450: skeletal metastasis secondary to malignant chondroid syringoma, Skeletal Radiol. 16 (1987) 657–659.

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