Synchronous atypical fibroxanthoma and Bowen’s disease of the head and neck in an otherwise normal patient – a case report and review of literature

Simultanes Auftreten eines atypischen Fibroxanthoms und Plattenepithelkarzinoms im Kopf-Hals-Bereich – Fallbericht und Literaturübersicht

Abstract

Although both atypical fibroxanthoma and squamous cell carcinoma arise on top of sun-damaged skin of the elderly, there is no evidence in literature reporting a synchronous presentation of primary lesions of both malignancies in the head and neck regions. We report a case of synchronous atypical fibroxanthoma and squamous cell carcinoma in situ (Bowen’s disease) of the head and neck in an otherwise normal old Caucasian male patient. We reviewed the literature for cases of head and neck atypical fibroxanthoma in association with other skin malignancies with an overview over the risk factors and modalities of treatment. We would like to raise the awareness for the concept of multiple synchronous primary malignant lesions and the importance to anticipate and differentiate between different pathologies in order to provide adequate investigations and treatment for the patient.

Keywords: atypical fibroxanthoma, squamous cell carcinoma, synchronous cancers

Zusammenfassung


Introduction

The risk factors that contribute to the development of the non-melanoma skin cancer (NMSC) include ethnicity, age, gender, chronic exposure to chemical and physical mutagens, besides genetic factors [1]. Excessive exposure to ultraviolet (UV) radiation, especially type B (UVB), has been associated with a high risk of developing skin cancers, namely squamous cell carcinoma (SCC) and basal cell carcinoma (BCC). The DNA damage due to UVB results...
in actinic keratoses (AK), solar lentigo, and Dermatoheli-
osis (i.e., “photoaging”) [2]. A small percentage of squamous cell carcinomas, however, do not follow these pathological changes stimulated by UV radiation exposure. In such cases, skin cancer may arise in skin wounds, and immunosuppressed persons may have higher incidence [3]. By far, the most frequent types of epithelial skin cancers are BCC and SCC [4], [5]. Atypical fibroxanthoma (AFX) is relatively uncommon, and was first was first defined by Helwig in 1961 to describe some cutaneous tumors with marked cellular pleomorphism but benign clinical course [6]. AFX is considered now a malignant fibrohistiocytic neoplasm that most commonly arises on sun damaged skin of elderly individuals [7].

Patients with head and neck skin cancers frequently develop multiple skin cancers. According to the timing of presentation, these multiple lesions could be classified as synchronous or metachronous metastases. Multiple synchronous primary lesions (SEER Program Coding and Staging Manual 2004, Revision 1) are those primary lesions with different pathology, occurring within 2 months of each other. Being aware of this concept is important in the clinical practice in order to be able to treat and investigate the patient adequately according to the presented lesions. Bearing in mind that a patient may have a totally different synchronous primary lesion can influence further investigations of the patient and can avoid inappropriate management procedures that may be considered undertreatment or sometimes an overtreatment.

We present here a case of synchronous AFX and SCC in situ occurring in the head and neck region.

Case report

We report a case of synchronous AFX and SCC in situ (Bowen’s disease) in the skin of the Head and neck region. The patient is a 74-year-old Caucasian male, working as field engineer, with no history of irradiation or immunosuppressive therapy. The patient presented to the out patient clinic with multiple scalp and nasal lesions (Figure 1). Immunohistochemically confirmed biopsies from the scalp reported atypical fibroxanthoma (positive for vimentin, negative for S100, CD31, and cytokeratin) (Figure 2). Two other suspected lesions from the left side of the nose and the right side of the scalp were biopsied and revealed squamous cell carcinoma in situ (Bowen’s disease). The lesions were reported to be surrounded with atrophic multifocal actinic keratosis, as well as massive solar elastosis. Definitive treatment of the patient was done by wide surgical excision of the lesions with appropriate safety margins (RO resection) (Figure 3; left). A scalp lesion was closed with split thickness skin graft, and the nasal defect was treated with a delayed glabellar local flap (Figure 3; right). The postoperative period passed uneventful and the patient was regularly followed up for eventual appearance of other lesions, recurrence or metastasis (Figure 4). Five months later, the patient developed another small (1 cm) SCC lesion at the back of the scalp that was surgically removed (RO) and the defect was covered using a full thickness skin graft. The patient has undergone an esthetic correction operation for the nose aperture 6 months later. No recurrence or metastasis was diagnosed through a 3-year follow up period. The patient was advised to avoid direct sun light exposure, was encouraged to make regular follow up visits, and to promptly seek medical advice on appearance of any suspicious new skin lesions.

Discussion

In our case, the AFX stained negative for cytokeratin. Mirza and Weedon previously studied 89 cases of AFX and found cytokeratin expression in only one of the lesions that displayed dual features of AFX and squamous cell carcinoma. Based on these data, they concluded that it appears unlikely that AFX is a variant of squamous cell carcinoma [8]. This confirms the unique identity of each of the tumours and thus defines both tumours occurring in the same patient as multiple synchronous primary tumours. In our case the lesions were reported to be surrounded with atrophic multifocal actinic keratosis, as well as massive solar elastosis. Actinic keratoses is defined as a precursor to SCC, and studies have shown that most cases of SCC arise from AK lesions [9], [10], [11].

To the best of our knowledge, this is the first report of synchronous AFX and SCC (CIS) occurring in the region of the head and neck in a non-XP (Xeroderma Pigmentosa) patient occurring on the native skin of the patient (not on burn scars or grafted skin). Youssef et al. previously reported about a case of synchronous AFX and basosquamous cell carcinoma in the face. The 6-year-old girl however was suffering from XP [12]. A 66-year-old male patient was reported with several squamous cell carcinomas and a myxoid atypical fibroxanthoma of the skin. The tumours developed in burn scars that the patient had had for more than 50 years and that, in part, undergone actinic elastosis [13]. A reported case of atypical fibroxanthoma, lentigo malignant melanoma and squamous cell carcinoma developed in the site of a thermal burn that had previously been treated with multiple skin grafts. The SCC and melanoma in that case occurred synchronously but they occurred 10 years after the AFX [14]. It was reported in literature that some cases of AFX were associated with SCC and other NMSCs but this was never reported to occur synchronously [7], [15], [16], [17], [18].

Possible causes for this multiplicity were thoroughly investigated in literature. A decrease in the cellular tolerance of U.V. radiation was demonstrated for individuals with multiple epidermal cancers. Ultraviolet light, which is the major etiology of human skin cancer, will cause mutations in the P53 gene. Such mutations occur in more than one-half of nonmelanoma squamous cell cancer and precancer. It was concluded that P53 patches, estim-
Figure 1: Left: Actinic keratosis of the scalp, the arrow points at the SCC in situ lesion of the scalp. Right: The arrow points at the SCC in situ lesion of the nose.

Figure 2: Left: The pleomorphic dermal tumor abuts against the epidermis. Secondary hemorrhages are present. The flattened epithelial layer corresponds to an atrophic actinic keratosis. Right: AFX lesion of the scalp (H&E stain 200X), cytologically malignant spindle and epithelioid cells are present.

Figure 3: Left: The lesions after excision; intraoperative, Right: Delayed glabellar flap
ated to be approximately 100,000 times as common as dysplasia, have a very small or even no precancerous potential. Their common presence demonstrates that human epidermis contains a large number of P53 mutations apparently without detrimental effect. The only result of the mutation may be a ‘benign clonal keratinocytes proliferation’. So, P53 patches are frequent companions of different types of squamous cell neoplasia, but the genetic links that would support their status as early precursors of malignant neoplasia were still lacking. Treatment modalities for primary NMSC tumours include surgical excision, radiotherapy, curettage with electrocautery, cryotherapy, laser destruction, intralesional Interferon, and some topical drug regimens. Surgical excision is the most effective treatment but renders the most tissue loss which requires functional and esthetic reconstruction in cases of head and neck cancers. Moh’s surgery may be used for large, aggressive, recurrent or location-sensitive lesions. It is the most definitive treatment but costly and time intensive. A study showed Mayo Clinic’s experience in treating AFX with Moh’s micrographic surgery (MMS) and concluded that microscopic control of the surgical margins with MMS in the treatment of AFX results in a lower recurrence rate than that with wide local excision and conserves normal tissue [19]. Radiotherapy is less effective than surgery (four-year follow-up); can cause dyspigmentation, telangiectasia and not suitable for sensitive skin as xeroderma pigmentosa patients [20], [21], [22]. Curettage with electrocautery and cryotherapy have got higher recurrence rate than surgery and radiotherapy. Also pain, leaking, and wound infection are not infrequent. Laser destruction for BCC has better cosmetic results but still with higher clinical recurrence rate than cryotherapy [20].

**Conclusion**

We report a case of synchronous atypical fibroxanthoma and SCC in situ (Bowen’s disease) of the head and neck in an otherwise normal old Caucasian. To the best of our knowledge this is the first case to be reported in literature with such a synchronous presentation in an otherwise normal patient. We emphasize bearing in mind the possibility of a synchronous presentation of different primary malignant lesions – especially in patients with actinic keratosis – which can help provide appropriate investigations and treatment procedures.

**Notes**

**Competing interests**

The authors declare that they have no competing interests.

**References**


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