



UNUSUAL ASSOCIATION OF BAZEX SYNDROME AND A CHOLANGIOCARCINOMA

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ABSTRACT

Acrokeratosis Paraneoplastica or Bazex syndrome is a paraneoplastic process that is usually associated with the diagnosis of squamous cell carcinoma of the upper aerodigestive tract and which affects primarily caucasian males. We describe a typical case of Bazex syndrome in a dark-skinned female associated with cholangiocarcinoma which is to our knowledge the second observation reported in the literature.

KEYWORDS: Acrokeratosis Paraneoplastica, Bazex, cholangiocarcinoma.

INTRODUCTION

Acrokeratosis Paraneoplastica or Bazex syndrome is a dermatologic manifestation described the first time in 1965 as a paraneoplastic process that is usually associated with the diagnosis of squamous cell carcinoma of the upper aerodigestive tract and which affects primarily caucasian males with an average age of 40 years.^[1]

It is characterized by a psoriasiform eruption that favors acral sites, palmoplantar keratosis and symmetric onychodystrophy. Nose and ears are affected in 63% and 79% of patients^[1] respectively. Its physiopathology is still unknown and it often precede the diagnosis of the tumor by an average of 11 months^[2] but it may have a parallel evolution with the co-existing malignancy.^[3]

We describe a typical case of Bazex syndrome in a dark-skinned female associated with cholangiocarcinoma which is unusual and - To our knowledge-the second observation^[4] reported in the literature.

CASE REPORT

Seventy four years old female, dark-skinned (phototype V), admitted in visceral surgery Department for cholangiocarcinoma confirmed histologically based on the endoscopic retrograde cholangio pancreatographic findings and an ultrasound-guided biopsy of the mass, who presented since 7 months (2 months before the cholangiocarcinoma became symptomatic: fatigue, recurrent abdominal pain, nausea, vomiting and quick weight loss) generalized pruritus with erythematous lesions of the face especially the nose and the ear and

extremities extending to the arms and the rest of the body.

our medical advice was sought before the surgery of the patient for his cholangiocarcinoma and our examination had found icterus with well-delimited finely scaly erythematous and violet patches of the dorsal area of the hands; forearms ,arms; legs, feet and the trunk, with excoriations and scaly patches of the scalp in addition to a negative sign of Brocq when scraping lesions, (Figure 1, 2, 3,5) palmoplantar keratoderma (PPK) (Figure 4, 6); the nails examination was disturbed by the application of henna (figure 3, 5).

The main differential diagnoses included psoriasis because of the psoriasiform pattern of the lesions , an eczematous eruption with an irritant or allergic component and pityriasis Rotunda since its occurrence in dark-skinned females.^[5]

The histopathology was nonspecific, with findings of hyperkeratosis, acanthosis and perivascular lymphocytic infiltrate.

As a result, the diagnosis of paraneoplastic dermatosis was established; we had rejected the diagnosis of pityriasis Rotunda because of the presence of pruritus and PPK and we had retained the diagnosis of Bazex syndrome secondary to the cholangiocarcinoma.

And we prescribed for our patient emollients creams and topical corticosteroids with a slight regression of symptoms but we didn't see the patient after the surgery.



Figure 1 : psoriasiform lesions of the forearms



Figure 2: psoriasiform lesions of the ears



Figure 3 : psoriasiform lesions of the dorsum of the hands with unguis dystrophy (covered by the henna)



Figure 4: palmar keratoderma



Figure 5: Psoriasiform lesions of the dorsum of the feet



Figure 6: Plantar keratoderma

DISCUSSION

Bazex syndrome is a rare paraneoplastic eruption described for the first time by Bazex and al. in 1965.^[6] It affects rarely female patients, with only twelve of forty-five cases^[7-28] affected in the largest case series.^[1] The typical clinical features are a non or little pruritic^[29] erythematous psoriasiform eruption that favours acral sites such as the fingers (61%), toes (39%), ears (79%) and nose (63%). Nail involvement is common, and is typified by horizontal and vertical ridging, yellow discoloration, onycholysis, subungual hyperkeratosis and in more severe cases, nail plate atrophy. In our patient

the condition was pruritic which could be exaggerated by the cholestasis since our patient had cholangiocarcinoma and the nail examination was disturbed by the application of Henna.

The clinical manifestations of Bazex syndrome can either precede the diagnosis of the malignancy in 65%–70 % of cases or occur simultaneously in 15%–25% of cases or develop following diagnosis of the malignancy in 10%–15% of cases^[30], in our patient, the psoriasiform rash precede the symptomatic stage of the malignancy by 2 months.

The cutaneous findings of acrokeratosis paraneoplastica of Bazex evolve through three stages, In the first one, erythematous psoriasiform plaques are found on the fingers, toes, helices of ears and nose. Nail folds are usually involved, leading to nail dystrophy, the associated tumor is frequently asymptomatic at this stage. In the second stage, the skin eruption spreads to the palms and soles, with development of palmoplantar keratoderma, the associated tumor could display local symptoms at this stage. In the third stage as the case of our patient, if the tumor remains untreated, the psoriasiform rash could extend to the trunk and limbs.^[30]

Moreover, the histopathological findings of this condition are nonspecific and may include hyperkeratosis, parakeratosis, acanthosis, isolated necrosis of keratinocytes and a perivascular lymphohistiocytic inflammatory infiltrate.^[30]

Bazex syndrome's physiopathology is still not well understood. One postulated mechanism (proposed by Velez and Howard)^[31] is a molecular mimicry, where the immune system is activated against the skin due to cross-reactivity between antigens found in the tumor and the skin. Another postulated mechanism is the secretion of growth factors by the cancer cells, such as epidermal growth factor, transforming growth factor- α and insulin-like growth factor and also zinc deficiency associated to the neoplasm.^[1]

Carcinomas of the upper aerodigestive tract (larynx^[32-36], pharynx^[37], trachea, bronchus^[38-40] and the oesophagus^[41]) are found in more than half of the cases reported in the literature. Associated neoplasms in other locations (skin^[42-46], breast^[47-50], genitourinary tract^[51-53], liver^[54-55], colon and stomach^[56-59], Hodgkin's lymphoma^[60], liposarcoma^[61]) have also been reported.

The improvement of lesions is described in 91% of the cases after the primary tumor is treated. The cutaneous condition is refractory to topical treatment, which may be attempted with coal tar, salicylic acid, vitamin D analogues, corticoids and UVB or UVA radiation. Systemic treatment also presents inconsistent results in studies with etretinate, corticoids and antibiotics.^[1] For our patient, we prescribed emollients creams and topical corticosteroids.

Ultimately, it's important to notice that reappearance of skin lesions after resolution may signify a recurrence of the tumor,^[30] which is a good criteria for follow up of the malignancy.

CONCLUSION

Our patient represents one of the few dark skinned females described with this syndrome and -to our knowledge- the second observation reported having an unusual association with cholangiocarcinoma.

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