Case report

PERINEURAL INVASION IN SOLITARY KERATOACANTHOMA: A MALIGNANT FEATURE?

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ABSTRACT

Solitary keratoacanthoma is a very frequent neoplasm, whose biology, however, still remains unexplained. In fact, as our case shows, it is always considered a benign neoplasm, even though it may present malignant features. So, where does the truth lie? Our case cannot solve the problem but it stresses the importance of collecting further information about the biology of keratoacanthoma.

KEY WORDS

keratoacanthoma, neoplasm invasiveness, prognosis

INTRODUCTION

Solitary keratoacanthoma (SKA) is usually considered a benign, self-healing skin lesion, although it may sometimes exhibit confounding features which would seem to indicate a malignant attitude.
We present a case which confirms this dual identity of SKA and underlines the paucity of information about its biology.

CASE REPORT

A 80-year-old male was examined by a surgeon because of a swelling in the left zygomatic region. Macroscopically, the lesion consisted of a firm, red, dome-shaped nodule, 1 cm in diameter, with a crater in its center.

Histological examination of the surgical specimen, performed by embedding in paraffin and staining with haematoxilyn-eosin, showed a roughly symmetrical, crateriform lesion composed peripherally of variably thickened squamous epithelium. The cells had glassy, eosinophilic cytoplasm and there were occasional mitotic figures; cytologic atypia was lacking. The central crater was filled with orthokeratotic keratin with occasional flakes of parakeratosis (Fig. 1). Diffuse perineural infiltration was present in the deep dermis and subcutaneous tissue away from the main lesion with neoplastic epithelium intimately involving the nerves, either by completely enveloping them or by coursing immediately adjacent. Foci of perineural infiltration were adjacent to the deep surgical margin of the specimen but apparently they did not cross it (Fig. 2, 3). Fibrosis around the involved nerves was focally striking. The tumor cells infiltrating the nerves were morphologically like those constituting the main neoplastic mass. The diagnosis was Solitary keratoacanthoma with diffuse perineural

infiltration.

Six months later the patient is well and no signs of relapse or metastases are present.

DISCUSSION

Biology of SKA is still misunderstood: although the vast majority of cases is self-healing and surgical removal is generally aimed at ascertaining the true nature of any swelling, different and confounding information is available in literature about a possible attribution of SKA to malignant neoplasms.

In 1993, Hodak et al. (1) stated that "SKA is a squamous-cell carcinoma". They based their opinion on three cases of SKA with metastases observed by them and on a case of giant KA (2) considered by Hodak et al.(1), the only true (one out of 9) metastasizing KA previously reported in Literature. The authors, supporting the opinion expressed by other authors (2-5), claimed that "KA is squamous-cell carcinoma from the outset, just as is solar keratosis, arsenical keratosis, radiation keratosis, and Bowen's disease". Now, if we consider that the reported diseases should really be classified as premalignant and/or

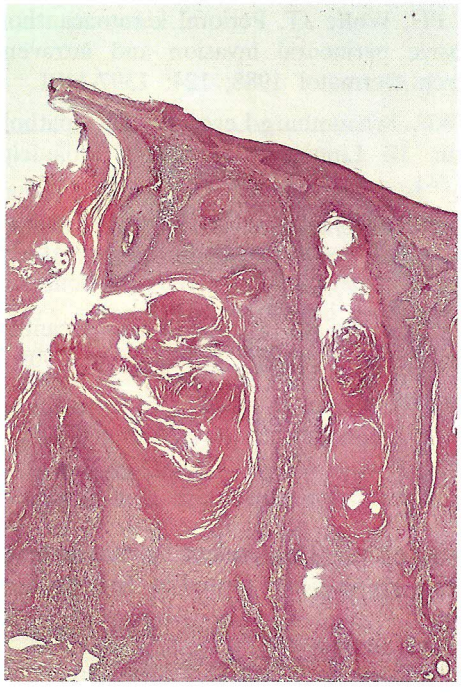

Fig. 1. Side of the central crater of the lesion, H&E 6x

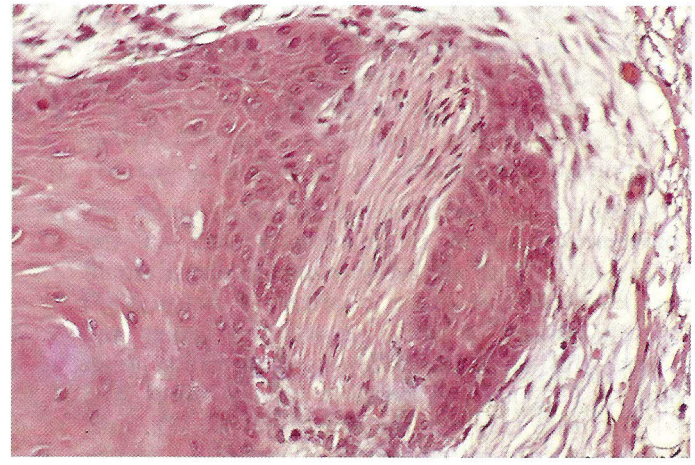

Fig. 2. Perineural infiltration near the main mass, H&E 40x

malignant lesions, slowly but fatally progressing to invasive forms, and that, on the other hand, judging by the very small number of cases reported in literature, the tendency of SKA to metastasize is exceedingly uncommon, there is a striking discrepancy between the huge number of SKAs with benign clinical and histological features and their supposed malignant attitude. In other words, we may state that the very few reported cases with metastases could be considered at the most "the exception proving the rule".

In the cases reported above, metastases were present in lymph nodes and distant districts without any evidence of local aggressive behaviour of the SKAs. On the contrary, our case and few others reported in literature, are characterized by local features of malignancy like perineural infiltration and vascular invasion, without evidence of distant metastases. Vascular invasion was occasionally reported both alone (6) or associated with perineural invasion (7,8)

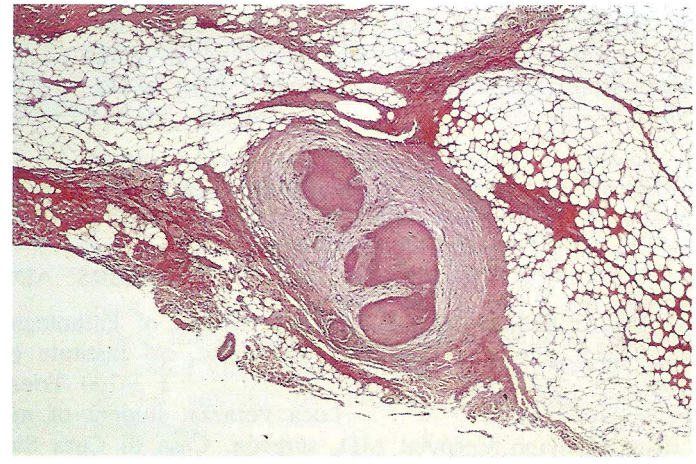

Fig. 3. Perineural infiltration with desmoplastic reaction away from the main mass, H&E 24x

and according to Calonje and Wilson Jones (6) "whatever the explanation could be, the presence of intravascular invasion in keratoacanthomas does not seem to increase the risk of metastasis or recurrence". In fact, none of the reported cases showed further recurrences or metastases in the follow-up period.

According to Lever and Schaumburg-Lever (9) "perineural invasion is occasionally seen in the proliferative phase of keratoacanthoma and should not be misinterpreted as evidence of malignancy". In fact, out of 18 cases reported by Lapins and Helwig (10) in which follow-up information from three to 12 years was available for two thirds, and out of 6 cases reported by Janecka et al. (7), none showed evidence of recurrences or metastases; among four patients studied by Cooper and Wolfe (8), 2 SKAs recurred soon after excision but no further recurrences were observed after follow-up periods of 7 and 24 months.

In conclusion, what to say?

SKA must be considered an unexplained neoplasm which, when presenting histological features of malignancy hardly ever exhibits a malignant behaviour. However, even when it lacks (as is the rule) malignancy features, it may be very aggressive.

Clinically, the statement of Robbins and Bailey (11) that "Any deviation from the classic pattern of development or any unexpected associated clinical signs should alert the physician to the possibility of an alternative diagnosis" may be considered very useful in the practice. But for the pathologist what is the truth?

We think that the topic needs further contributions and we hope that our case will be contributive in this way.

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