ABSTRACT

Purpose/methods: To describe one case of invasive squamous cell carcinoma of the conjunctiva in a 74-year-old woman, who two years previously had presented with a lesion which appeared to be an anterior nodular scleritis.

Results/conclusions: An anterior nodular scleritis, which did not respond to therapy, preceded the development of a squamous cell carcinoma of the conjunctiva adjacent to it. In our patient, the conjunctival squamous cell carcinoma could have masqueraded as a scleritis, delaying the correct diagnosis and allowing orbital spread of the tumor. A diagnosis of neoplasia must be considered when a scleritis shows an atypical appearance or does not respond to the usual therapy (Arch Soc Esp Oftalmol 2007; 82: 237-240).

Key words: Scleritis, squamous cell carcinoma, ocular invasion, conjunctiva-uveitis-sclerokeratitis.

RESUMEN

Objetivo/método: Se presenta una escleritis nodular anterior de dos años de evolución en una mujer de 74 años, que desarrolla posteriormente un carcinoma conjuntival escamoso invasivo, en la zona adyacente al nódulo de escleritis.

Resultados/conclusiones: El desarrollo de un carcinoma escamoso conjuntival que tiene como precedente la aparición adyacente de una escleritis nodular anterior que no respondió al tratamiento. En nuestra paciente la escleritis iniciada dos años antes pudo enmascarar el carcinoma escamoso, retrasando el diagnóstico y permitiendo la invasión orbitaria del tumor. Ante una escleritis de presentación atípica o sin respuesta al tratamiento debe considerarse la posibilidad de una neoplasia.

Palabras clave: Escleritis, carcinoma de células escamosas, invasión ocular-conjuntiva-uveitis-esclerokeratitis.
INTRODUCTION

The squamous cell conjunctiva carcinoma is an infrequent tumor (1) which develops in transition zones of the eye surface (conjunctiva or cornea) and expresses as a reddish-graying gelatinous mass (2). Occasionally it may begin without obvious mass and go unnoticed a long time, or appear as a scleritis. This paper presents an unusual form of squamous cell conjunctiva carcinoma in the environment of a long-evolution anterior scleritis refractory to treatment. Considering the overlapping of the alarm signs of the carcinoma with those of the sclerokeratitis, the delay in diagnosing carcinoma enhanced orbitary extension.

CASE REPORT

A 71-year old healthy Caucasian woman, diagnosed with anterior nodular scleritis in her left eye (LE). At the time her visual acuity (VA) was of 20/30 in RE and 20/50 in LE.

The 1.5 mm whitish scleral nodule was in the inferior nasal sector at 2mm from the limbus, under normal ophthalmological exploration. The systemic study revealed a slight reduction of IgM and lymphocitosis of 48%, with the remaining specific uveitis tests giving normal results. An empirical treatment was established with oral steroids, with paradoxical response and clear signs of clinical worsening. The patient rejected the use of immunosuppressants. After 6 months of treatment with steroids without controlling the symptoms, an intraocular pressure (IOP) was detected in the LE of 30 mmHg without response to pressure-reducing drugs. After 10 months of evolution the process worsened, with greater nodular elevation, corneal edema and ulceration of the perilesional conjunctiva (fig. 1). The possibility of a biopsy was discarded due to the patient’s refusal and a potential risk of perforation. After 16 months, treatment was initiated with Methotrexate, which considerably worsened the clinical symptoms, and IOP of 45 mmHg which did not react to the treatment. After 21 months, the scleral node changed, showing a scleroconjunctival destructuring and a reddish gelatinous flat mass (fig. 2). A conjunctival biopsy was performed which confirmed the presence of a squamous conjunctiva carcinoma.

The orbitary and neck TC revealed the involvement of the medial rectum and the lachrymal sac of the LE. Due to the evolution of the process, an exenteration was performed with rotational flap of the temporal muscle. The anatomic and pathological study revealed the scleral nodular lesion and the adjacent conjunctiva carcinoma (figs. 3 and 4). After one year of said surgery, the patient remains without symptoms or relapse, and the extension study gave negative results.

DISCUSSION

Neoplastic lesions of the conjunctival squamous epithelium include type 2 displasia, intra-epithelial neoplasia and squamous cell carcinoma.
The squamous cell carcinoma has a low prevalence, between 0.02 y 3.5 /100,000, lower in high latitudes (3). It appears between age 60-70 and is located in interpalpebral exposed areas, in epithelium transition areas (limbus). Risk factors associated to its appearance are exposure to UV rays and pigmentary xeroderma (above all in young patients). The existence of burns and injuries in the ocular surface could also facilitate the appearance of the carcinoma. The invasive squamous cell conjunctiva carcinoma is defined by the infiltration of the basal membrane by atypical cells. Orbitaly invasion appears in 12-16% (4).

Even though the clinical expression of the carcinoma as a reddish gelatinous mass in the interpalpebral region is highly suggestive. Occasionally, it can appear in a non-typical way as a whitish lesion, which leads to it being mistaken with other processes and generating a masquerading syndrome (2).

The literature does not describe many cases of invasive squamous cell conjunctiva carcinoma associated to scleritis (5) or sclerokeratitis (2). This case is exceptional because of the presentation of the nodular sclerokeratitis and the late appearance of an adjacent squamous conjunctival carcinoma which was confirmed subsequently in the anatomic and pathological examination.

Said study shows the tumoral process (Fig. 3) with abundant nests of cells with keratin and mitosis (rapid growth) and little lymphocytic inflammatory infiltrate. Here the tumoral process grows from the surface (conjunctiva) instead of from scleral or corneal areas. Figure 4 shows the merger of both lesions, the scleral one with its accumulation of lymphocytes without tumoral cells and, detached from it, a tumoral niche which is not related to the previous lesion.

The atypical behavior of a scleritis must lead us to consider other diagnostic alternatives such as conjunctival carcinoma. It is a subject of debate whether scleritis is the precursor of carcinoma due to a local irritation cause produced by the chronic inflammation of the area (if they are two independent processes which coincide in time giving rise to a masquerading process) or from the beginning the lesion was a tumor with a high inflammatory component.

In any case, any delay in the diagnostic of a non-typical squamous conjunctival carcinoma enhances its infraorbitaly extension.

By way of conclusion, a diagnostic of neoplasia must be considered in the presence of a scleritis which is atypical or does not respond to treatment.

REFERENCES

