CASE REPORT

Cystic lesion of the parotid following drug-induced toxic epidermal necrolysis (Lyell’s syndrome)

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A 32-year-old woman developed a unilateral cyst of the duct of parotid gland 4 months after severe oral involvement of drug-induced toxic epidermal necrolysis (TEN). The pathomechanism leading to the TEN epidermal destruction had presumably involved the salivary epithelium as well, leading to the development of the cystic lesion. The patient had low serum lipase levels, but high serum amylase levels at the time of TEN. These serological markers could represent a clue for the risk of developing cystic lesions of the large salivary glands following TEN.

Keywords: cyst; Lyell’s syndrome; parotid; toxic epidermal necrolysis

A 32-year-old woman developed drug-induced toxic epidermal necrolysis (TEN) 48 h after oral intake of ibuprofen for headache. An erythematous rash covered by blisters spread over 64% of the body surface. Severe oral lesions were present associating oropharyngeal erosions and bullae on the palate, tongue and cheek mucosa. Ocular involvement consisted of conjunctival synechiae and corneal ulcerations. The diagnosis of TEN was confirmed by histological examination of a skin biopsy taken 72 h after the initial cutaneous lesions. A subepidermal blister was present with rare inflammatory mononuclear cells scattered between confluent necrotic keratinocytes. The patient was hospitalized for 3 weeks in a burn unit where she was placed on a fluidized bed. She was also treated with cyclosporin IV (5 mg/kg/day) for five consecutive days. Corticosteroids were not administered. During that period of time, the amylase serum values were severely and repeatedly increased during her three-week hospitalization reaching a maximum of 998 IU/ml (normal upper limit: 90 IU/ml). By contrast, the lipase serum values remained near to the upmost normal range limit, occasionally and moderately above it with a peak value of 225 IU/ml (upper limit of normal values: 200 IU/ml). The patient was discharged from the burn unit following complete cutaneous reepidermization and healing of the oropharyngeal ulcerations.

Four months after TEN, she progressively developed an oral swelling lesion nearby the left Stenon’s duct orifice (Fig. 1). Histological examination of the surgically removed lesion showed a unilocular 0.9 cm diameter well circumscribed cyst filled by some amorphous material. The cyst wall was surrounded by a dense fibrous connective tissue containing a mild inflammatory cell infiltrate. The luminal surface of the cyst was lined by an epithelium that exhibited a stratified, squamous or columnar aspect according to the different areas. A salivary duct cyst of the parotid was diagnosed.

Comments

Toxic epidermal necrolysis is a rare potentially lethal disease characterized by sudden necrosis of the epidermis, and of the genital and oropharyngeal mucosae (1). The only recognized cause of the disease is an adverse reaction to drugs, especially sulfonamides, phenytoin, non-steroidal antiinflammatory drugs and allopurinol. Death rate due to TEN reaches about 30%. Pulmonary, myocardial, osseous, renal and gastrointestinal long-term sequelae have been reported (1). However, the most commonly recognized long-term alterations involve the eyes, skin, nails and the genital mucosae (1). Conjunctival retraction, scars and sicca syndrome with severe corneal lesions represent the main ocular complications, sometimes resulting in a major handicap for the patient. Cutaneous sequelae include keloidal scars, alopecia, speckled eruptive melanocytic naevi and hyper- or hypomelanosis. Residual nail dystrophies are frequent. Genital involvement can lead to chronic vulvovaginal synechiae. Oral and lip lesions usually heal without complication, but strictures may develop in the throat and esophagus (1).
Sjögren-like syndrome with xerostomia is also a recognized complication of TEN (2). This condition results from a chronic lymphocytic infiltration of small salivary glands. However, post-TEN cystic lesion of the large salivary glands has not been described so far. We report one adult patient who developed unilateral cystic lesion of the parotid duct following severe oral TEN involvement. Ductal obstruction is thought to be the main etiologic factor of this type of salivary ducts (3). No malignant neoplasm, calculi or mucus plugs were found in the presently reported patient. Hence, the prolonged elevation in serum amylase was probably due to a salivary involvement. In TEN, the incidence of hyperamylasemia is over 30% of the patients, and salivary hyperamylasemia is predominant (9). Hyperamylasemia reflects the extent in mucosal membrane involvement. It seems to have a predictive value of post-TEN Sjögren-like sicca syndrome (9). It is also possible that high serum amylase levels coupled with low lipase serum levels could have a predictive value for the development of post-TEN lesions of the large salivary glands. Indeed, we have another record in our files of a Warthin tumor of the parotid gland diagnosed 10 years following severe TEN. The serum amylase dosages were elevated at the time of skin blistering while serum lipase were in the normal range of values.

In conclusion, we report a TEN patient in whom a cystic lesion developed subsequently in the parotid gland apparatus. It is suggested that such a lesion represents a possible long-term consequence of TEN severely affecting the oral cavity.

References

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