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Recurrent benign pleomorphic adenoma of the lacrimal gland – a case report

Nawrót guza mieszanego gruczołu łzowego – opis przypadku

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Summary: Benign pleomorphic adenoma (mixed tumor) is the most frequently occurring tumor of epithelial origin. It occurs in serous glands, most frequently in parotid glands and rarely in small palatine glands, cheek glands, palatine tonsils and the lacrimal gland. This study is a description of the therapeutic management of a patient with recurrent pleomorphic adenoma of the lacrimal gland. In 1996, a 35 years old male patient underwent surgery to remove a tumor of the left orbit. Histopathological examination revealed pleomorphic adenoma. From 1998, the patient had observed increasing exophthalmus of the left eyeball. In 1999, vision distortion and headaches had begun. In 2005, from a cut along the upper-outer edge of the left orbit, an elastic and soft tumor 1 cm in diameter was removed from soft tissues of the upper eyelid. Then, with the use of the side orbitotomy technique, the orbit was opened through incision of the temporal muscle and removal of the orbital side wall. An eyelid nodule of 0.8 mm in diameter was removed together with an elastic and hard tumor of size 2.5 × 1 cm. The result of the histopathological examination of the palpebral tumor was pseudocystic tumor and of the orbital tumor was pleomorphic adenoma. Conclusion: patients resected for pleomorphic adenoma of the lacrimal gland require a long period of postoperative observation and periodic ophthalmologic examination.

Key words: pleomorphic adenoma, lacrimal gland, diagnosis, treatment.

Introduction

Benign pleomorphic adenoma (mixed tumor) is the most frequently occurring tumor of epithelial origin. It occurs in serous glands, most frequently in parotid glands and rarely in small palatine glands, cheek glands, palatine tonsils and the lacrimal gland. It accounts for approximately 50% of the total lacrimal gland tumors of epithelial origin and occurs more frequently in males (60%) than females (40%) between the third and the seventh decade of life (on average around the age of 40). It occurs extremely rarely in juveniles (1).

The lacrimal gland consists of two lobes: the palpebral lobe and the more deeply located orbital lobe, separated by the aponeurosis of the upper eyelid levator muscle. Although the eyelid lobe constitutes between one-third and one-half of the total size of the orbital lobe, the mixed tumor stems mostly from the deep lobe (2). A characteristic quality of the benign pleomorphic adenoma is its very slow pain-free growth and its inclination for local recurrence. As a result of its increasing weight, the adenoma causes displacement of the eyeball most often directed downwards and outwards, which leads to exophthalmus and double vision. A characteristic symptom is also a slowly progressing ptosis. Most patients suffer from such symptoms at least a year before starting treatment (3).

Diagnosis of the mixed tumor is based on clinical and radiological examinations which take the form of computer tomography of the frontal plane, and in some cases also magnetic resonance imaging (4).

Treatment of the benign adenoma revolves around radical surgical resection of the tumor together with pseudocapsule and a margin of orbital tissues. The tendency for local recurrence even many years after the extirpation of the tumor may indicate its malignant transformation. It is characterized by its very rapid growth with accompanying pain and this quickly leads to distortion of vision. So far it has not been discovered whether a tumor resected not radically enough is the cause of its malignant transformation, or whether it is the cause of a new tumor of malignant characteristics (5).

The vast majority of authors advocate the avoidance of aspiration biopsy of the pleomorphic adenoma due to the possibility of damaging the capsule and scattering the malignant cells into neighboring tissues (3,4,5,6).

The aim of this study is a description of the therapeutic management of a patient with recurrent pleomorphic adenoma of the lacrimal gland.

Case report

In 2005, a 35 years old patient (J.K.) came to the Clinical Department of Maxillofacial Surgery in Katowice with symptoms of exophthalmus and vision distortion in the left eye, which made it difficult for him to work. In 1996, he had undergone surgery to remove a tumor of the left orbit. The histopathological examination revealed pleomorphic adenoma. From 1998, the patient had observed increasing exophthalmus of the left eyeball. By 1999, vision distortion and headaches had begun. The patient did not exhibit any general disorders and no history of hereditary disease in the family. The initial examination revealed exophthalmus of the left eyeball, inclined medially downwards. In the upper-outer orbital quadrant (within the eyelid), an elastic tumor was available for palpable examination, movable in relation to neighboring tissues. No diplopia occurred and reaction of pupils was regular. An ophthalmological consultation was carried out and revealed the following symptoms: decreased vision acuity down to 0.5, diplopia, exophthalmometry: RE 20 mm, LE 27 mm. The fundus of the left eye: optic disc II of blurred contours and raised; the retina plicated in the middle.

Computer tomography was carried out using the spiral method, with 0.5 mm layers with and without contrast medium (Fig. 1). The postoperative site contained recurrence of the tumor. Exophthalmus of the left eyeball was recognized. The eye additionally inclined medially due to the weight of the tumor, with no infiltration characteristics. There was loss of bone tissue adjacent to the tumor. The mass of the tumor slightly strengthened after administering the contrast medium. The right orbit and its contents remained unchanged.

The decision for surgical treatment was taken. In September 2005, surgery was carried out. From the cut along the upper-outer edge of the left orbit, an elastic and soft tumor 1 cm in diameter was removed from soft tissues of the upper eyelid. Then, using the side orbitotomy technique, the orbit was opened through incision of the temporal muscle and removal of the orbital side wall. An eyelid nodule of size 0.8 x 1.5 cm was removed together with an elastic and hard tumor of size 1.5 x 3 cm (Fig. 2).

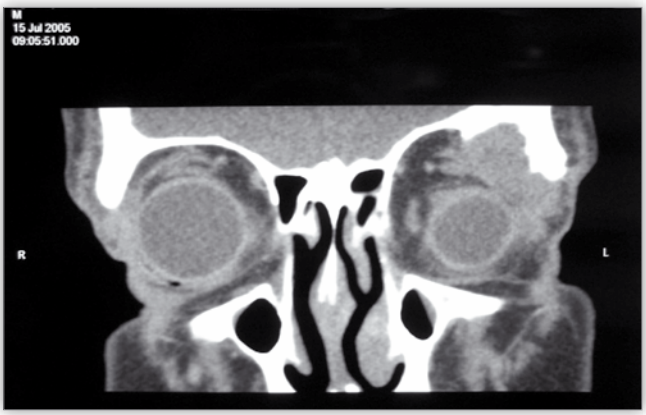

Fig.1. CT scan of the left orbital tumor

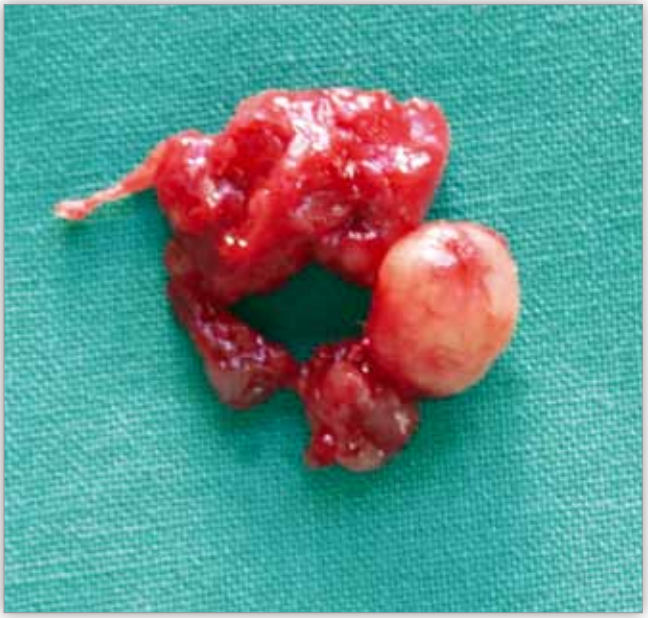

Fig.2. Two parts of palpebral and orbital after tumor removal.

The result of the histopathological examination:

1. Palpebral tumor

A pseudocystic tumor (0.8 x 1.5 cm) with fibrous pseudocapsule was sent for examination. Neoplasmatic cells infiltrated the pseudocapsule of the tumor in some areas (without crossing the pseudocapsule). The tumor showed epithelial cells forming tubular structures arranged in an irregularly anastomotic pattern, lying in a myxoid stroma. The inner layer of the epithelium secreted mucus or underwent squamous metaplasia.

2. Orbital tumor

An elastic, gray-brown tissue fragment (1.5 x 3.0 cm) was sent for examination. The histological section showed fragments of lacrimal gland, fibrous connective tissue, adipose tissue and fragments of bone trabeculae. The numerous focuses of pleomorphic adenoma infiltrated these tissues. Not many epithelial cells lay in a myxoid stroma which predominated in this tumor (Fig. 3).

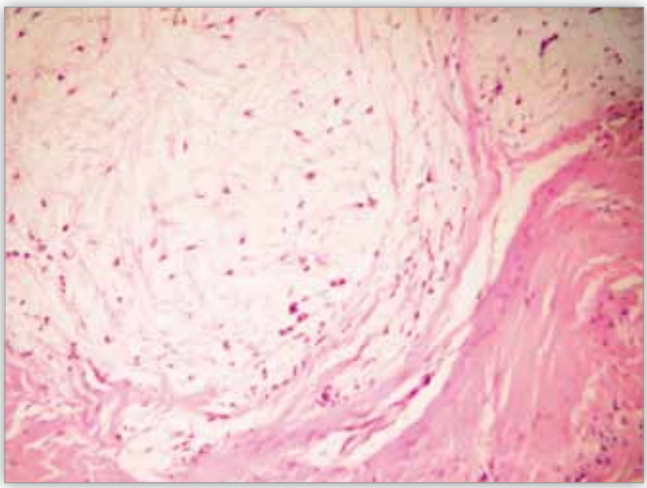

Fig.3. Myxoid stroma of pleomorphic adenoma lying in fibrous connective tissue.Magnification ×200. HE.

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The follow-up course was non-complicated. The patient was referred to the Oncological Institute for consultation but this did not confirm if the tumor was malignant. Periodic check-ups were recommended to be carried out at the operating center. The patient remains under periodic ophthalmological examination.

Discussion

Pleomorphic adenoma is the most frequent tumor of the lacrimal glands (50-60%). The tumor most frequently occurs in young adults (average age 35 years). It occurs twice as often in males than females. It is a locally malignant tumor which may infiltrate its own pseudocapsule and the adjacent periosteum. If not radically resected, it may cause recurrences in soft tissues or in the bone tissue (3). The pseudocapsule is created as a result of pressure of the tumor against the surrounding tissues, but its focal infiltration can nearly always be found. Such a tumor is characterized by a considerable variability of its histopathological image, and hence its name. The histopathological image reveals a myxoid stroma, low cell stroma which hosts high cell areas composed of epithelial cells, forming tubes and ducts incorporated into cords and lobes. The tubes and ducts are lined with two-layer epithelium. The interior layer of the cells may secrete mucus, or undergo squamous epithelium metaplasia. On the other hand, its exterior layer consists of myoepithelial cells which undergo metaplasia forming myxoid, fibrous or cartilaginous stroma (4).

Both benign and malignant orbital tumors should be differentiated from other pathological conditions such as inflammations, vessel malformations, or disorders of the lymphatic system. Appropriate diagnosis of tumor alterations facilitates selection of appropriate therapeutic management. The pleomorphic adenoma is regarded as a benign tumor whose course is initially very slow and hidden. It progresses without destruction of the orbital bones, which, if detected, may indicate the malignant nature of the tumor (1). In our case, in the computer tomography, we observed loss of the bone adjacent to the tumor. Yet, bone destruction may be revealed in the case of pseudotumor or cholesterolic granuloma whose characteristic feature is bleeding inside the tumor. In some cases, mixed tumor may develop from additional or ectopic gland tissue. That was the case with our patient, where under the eyelid there was a separate small nodule 1 cm in diameter resembling a cyst, whereas another tumor was present in the orbit. During the operation, bleeding was of medium intensity, which also bore out the validity of the original diagnosis (2).

In the MRI examination, it is very difficult to judge if we are dealing with benign or malignant pleomorphic adenoma. This is of crucial importance if the appropriate operative technique is to be adopted effectively. Benign tumors are resected by removal of the tumor with a margin of adjacent tissues, while malignant ones require exenteration of the orbit, which is related to considerable maiming of the patient (4). A solution to the problem could lie in aspiration biopsy, but the majority of authors do not recommend this in the case of mixed tumor (1,3,4,5). The paper by Tse and Folberg contains a description of a technique which prevents the possibility of scattering the malignant cells when a diagnostic biopsy is necessary. They covered the place of injection with a few drops of 2- butyl cyanoacrylate. Then, they cut out the frozen piece of tumor and if the diagnosis turned out to be correct, the whole lacrimal gland was resected (7).

Local recurrences of pleomorphic adenoma turn up at different time intervals after the first surgery. The majority of authors consider that approximately 30% of total recurrences are revealed after over 15 years. Around 10% of the total pleomorphic adenoma are transformed into malignant tumors after 20 years from the first surgery, and 20% after 30 years (1). Tse and colleague analyzed the lives of 42 patients treated for benign pleomorphic adenoma in the period between 0.5 and 17 years after surgery. In total, 34 patients did not suffer from recurrences or malignant transformations, and 8 had recurrences, 7 of whom underwent multiple surgeries (7).

In the case presented, the recurrence 9 years after the first surgery could suggest malignant transformation. However, on the basis of computer tomography, the decision was taken to implement a sparing procedure, the validity of which was confirmed by histopathological examination. When pleomorphic adenoma is present, the entire tumor with its pseudocapsule, surrounding levator aponeurosis, and conjunctiva must be excised to avoid recurrences and malignant transformation.

Conclusion

Patients resected for pleomorphic adenoma of the lacrimal gland require a long period of postoperative observation and periodic ophthalmologic examination.

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