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OLGU SUNUMU / CASE REPORT

Lacrimal gland tumor presenting with proptosis and retinal folds

Proptosis ve retinal ırışıklık ile prezente lakrimal bez tümörü

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ABSTRACT

A 47-year-old male patient, who complained of a small, painless, progressively enlarging lesion in the temporal of the orbit, was examined for ophthalmologic findings. Visual acuity was about ½ meters in the right eye and 20/20 fingers in the left eye. Direct and indirect pupillary reactions were normal, there was limitation in ocular movements. The amount of ptosis was 35 mm in the right and 20 mm in the left eye. There was macular pigment epithelial atrophy and chorioretinal folds in the temporal quadrant of the right fundus. In the pathological examination, the mass was evaluated as pleomorphic adenocarcinoma of the lacrimal gland. Although pleomorphic adenoma (benign mixed tumors) is an encapsulated tumor of the lacrimal gland with an excellent prognosis, it may undergo malignant transformation in rare cases.

Keywords: Lacrimal gland, pleomorphic adenocarcinoma, orbital masses

ÖZET

Orbitanın temporalinde küçük, ağrısız, giderek genişleyen lezyondan yakınan 47 yaşındaki erkek hasta oftalmolojik bulguları nedeniyle muayene edildi. Görme keskinlikleri sağ gözde yaklaşık ½ metre ve sol gözde 20/20 parmak sayıyordu. Direkt ve indirekt pupil reaksiyonları normaldi, oküler hareketlerde kısıtlılık vardı. Pitoz miktarı sağda 35 mm, sol gözde 20 mm idi. Sağ fundusun temporal kadranında maküler pigment epitel atrofisi ve korioretinal kıvrımlar vardı. Yapılan patolojik incelemede kitle lakrimal bezin pleomorfik adenokarsinomunu olarak değerlendirildi. Pleomorfik adenom (iyi huylu karma tümörler), mükemmel prognozlu, lakrimal bezin kapsüllenmiş bir tümörü olmasına rağmen, nadir durumlarda malign transformasyona da uğrayabilir.

Anahtar Kelimeler: Lakrimal bez, pleomorfik adenokarsinom, orbital kitleler

INTRODUCTION

Approximately 5% to 10% of orbital masses are composed of space-occupying lesions of the lacrimal gland and its fossa. Based on the clinicopathologic studies lesions of the lacrimal gland are originated from 50% epithelial elements and 50% are of nonepithelial elements. Of nonepithelial lesions, 50% are lymphoid tumors and 50% are infections and inflammatory pseudotumors. Epithelial tumors of the lacrimal gland are 50% pleomorphic adenomas (benign mixed tumors), 25% adenoid cystic carcinoma, and the remainders are other types of carcinoma. An acute presentation without adjacent bony changes is suggestive of inflammatory disorders. Insidious painless onset (less than 1 year) in an old age group with radiographic evidence of a lesion molding or conforming to ocular and bony contours rather than indenting adjacent structures are hallmarks of lymphoproliferative diseases. Subacute presentation of short duration (usually 4 to 6 months) and radiographic evidence of infiltration of adjacent structures, irregular erosions or destruction of bone, and calcification are distinctive of malignant epithelial neoplasms. A chronic presentation without the pain associated with a radiographic finding of lacrimal fossa remodeling is suggestive of benign lacrimal gland tumors.[1]

The pleomorphic adenoma (benign mixed tumors) is an encapsulated epithelial tumor of lacrimal gland with an excellent prognosis for vision and survival following complete excision. In rare instances this tumor can undergo malignant transformation, particularly following incomplete surgical removal.[2-4]

Here we want to talk about a patient who is asymptomatic for many years but presenting proptosis and progressive visual loss about 1 year. We considered it to be a benign mixed tumor of the lacrimal gland and decided to remove it through the lateral orbitotomy approach.

CASE REPORT

A-47- year-old man was complaining about progressive visual loss and proptosis in the right eye for about 1 year. He stated that a small painless lesion, which was first appeared in his temporal aspect of orbit 8 years ago, was gradually expanding. During the ophthalmologic examination, the corrected visual acuities were counting fingers about ½ meters in the right eye and 20/20 in the left. The anterior segment, direct and indirect pupillary reactions were all normal. The ocular pressures were 18 in the right and 12 mmHg in the left eye. There were macular pigment epithelial atrophy and chorioretinal folds in the temporal quadrant of the right fundus (Figure 1-A). The amount of ptosis by Hertel exophtalmometry was 35 mm. There was a restriction in elevations and looking up in and out (Figure 2; A 1-3). Only green lights were seen by Worth four dot test. The lesion was about 2x2 cm in size, firm and immobile. Chorioretinal folds were seen in fundus fluorescein angiographic evaluation (Figure 1 B). There was no abnormality in hematologic evaluation. Coronal computed tomography demonstrated an extraconal placed 3x2.5 cm well-circumscribed lesion and no bony destructions in the adjacent structures. There was a focal thickening of right maxillary sinus mucosa (Figure 3). According to its clinical appearance we considered it to be a

pleomorphic adenoma (benign mixed tumors) of the lacrimal gland and decided to complete the removal of the lesion through the lateral orbitotomy approach.

Histopathologic study of this specimen revealed a malignant mixed tumor that was most likely arising from the lacrimal gland. The lesion was 3x3 cm in size, composing of solid and tubular structures of benign epithelial and myoepithelial cells in the myxoid stroma, large pleomorphic vesicular nuclei, large eosinophilic cytoplasm cells of malignant glandular structures. These cells were showing atypical mitosis. There was a capsular invasion but no vascular and perineural invasion (Figure 4). 1 month after the operation the corrected visual acuity was counting finger about 1 meter in the right eye. There was no restriction in looking right and left upwards (Figure 2;B1-6) and chorioretinal folds were decreased. As there was residual mass, radiotherapy was performed and the residual mass was disappeared. The patient was followed up to 4 years without any problem. (Figure 5 and 6)

DISCUSSION

The incidence of pleomorphic adenocarcinoma (malignant mixed tumor) of the lacrimal gland has been reported as 4% to 15%. The average age at diagnosis is 50 years but ranges from 15 to 80 years and has a predilection for men. This tumor may arise from an incomplete excision of a benign adenom or as malignant transformation years after the diagnosis of a presumed benign adenoma. When an orbital mass associated with proptosis for several years develops a sudden increase in proptosis, malignant transformation of pleomorphic adenoma should be considered.[1,2]

Grossly, the tumor is well-circumscribed with a pseudocapsule. Microscopically, tumor infiltration into adjacent soft tissues and bone may be seen.[1] Chorioretinal folds are a common manifestation of orbital disease. These folds can be seen in association with orbital masses, miscellaneous inflammatory conditions, pseudotumor, Graves' ophthalmopathy, mucoceles,

optic neuropathy. They have also been described as a manifestation of primary ocular disease, including hyperopia, hypotony, posterior scleritis, detachments of the retina and choroid, subretinal neovascular membranes, scleral buckling, uveitis, choroidal tumors, and trauma. Sometimes folds can occur with no identified associated condition.[5]

Tumors such as hemangiomas, metastatic neoplasms, and optic nerve meningiomas, are located within the muscle cone and can press on the globe posteriorly, producing exophthalmos, flattening of the globe, and shifting of the refractive error towards hyperopia. Intraconal tumors tend to produce straight folds that radiate away from the optic disk. An extraconal tumor may press directly on the extraocular muscles and Tenon's capsule, as well as on the sclera. Because of anterior segment distortion, astigmatic refractive errors are commonly induced. Extraconal tumors produce curved folds with the convex side directed away from the optic disk, but the nerve head is usually located outside the region of folds. Mucoceles, dermoids, tumors of the lacrimal gland, and orbital meningiomas are typical extraconal tumors associated with choroidal folds.[5,6] Incidental equatorial folds created by encircling procedures to repair retinal detachments do not cause distortion or refractive errors themselves, but the scleral buckle in this situation produces relative myopia.[6] The exact mechanism of chorioretinal folds formation is not known. It has been suggested that ocular compression by an orbital mass is the primary cause. However, folds are not induced by scleral indentation, and rapidly expanding orbital masses may lead to the scleral indentation in the absence of chorioretinal folds. Therefore, other changes must occur to account for their presence and some explanations have been proposed. One explanation is that they are produced by tension or shortening of the extent of Bruch's membrane due to choroidal edema, scleral thickening, or shrinkage of the globe. Traction on the optic nerve or compressive forces within the choroid may also be the factors. A localized increase in choroidal thickness, causing Bruch's membrane to be thrown into folds has also been suggested. The close

anatomical attachment of Bruch's membrane to the underlying choriocapillaris supports this view. Degeneration of elastic tissue and a reduction in the area of the inner surface of the sclera due to thickening have been suggested to be factors as well.[5] Chorioretinal folds running through the fovea may cause metamorphopsia and a reduction of visual acuity that cannot be eliminated by corrective lenses.[6] Our patient was complaining about a small painless masses in his temporal aspect of orbita, that was gradually expanding for 8 years. As there was no pain but proptosis, we considered it to be a pleomorphic adenoma, an epithelial tumor of the lacrimal gland. Because of the direct pressure effect of the tumor to retina, there were chorioretinal folds and macular pigment epithelial atrophy. That produced visual loss and metamorphopsia. If chorioretinal folds remain for months, retinal pigment epithelium (RPE) in the troughs may undergo hypertrophy and hyperplasia from being mechanically compacted. Even if the folds resolve, this linear pigmentation may persist, RPE atrophy and apparent fractures of Bruch's membrane may also be seen along the folds, especially in older patients.[6] In our patient, by the removal of the tumor there was no the pressure effect on the retina, so retinal folds were disappeared but pigment epithelial atrophy was seen

The best approach in the treatment of lacrimal gland tumor must be complete surgical resection of the tumor within its capsule. Even with complete resection, mortality remains high, with 50% of patients live for 12 years. Patients with pleomorphic adenocarcinomas transformed from bening adenoma lived longer (mean 19 years). Intracranial spread and metastasis to the lung, chest wall, sacrum, and scapula have been reported.[1] After removal the orbital mass, control tomography was taken and as there was residual mass, we decided to perform radioteraphy and the patient was followed up to two years without having any complains.

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Figure Legends

Figure 1-A: Colour fundus photograph of the patient's right eye

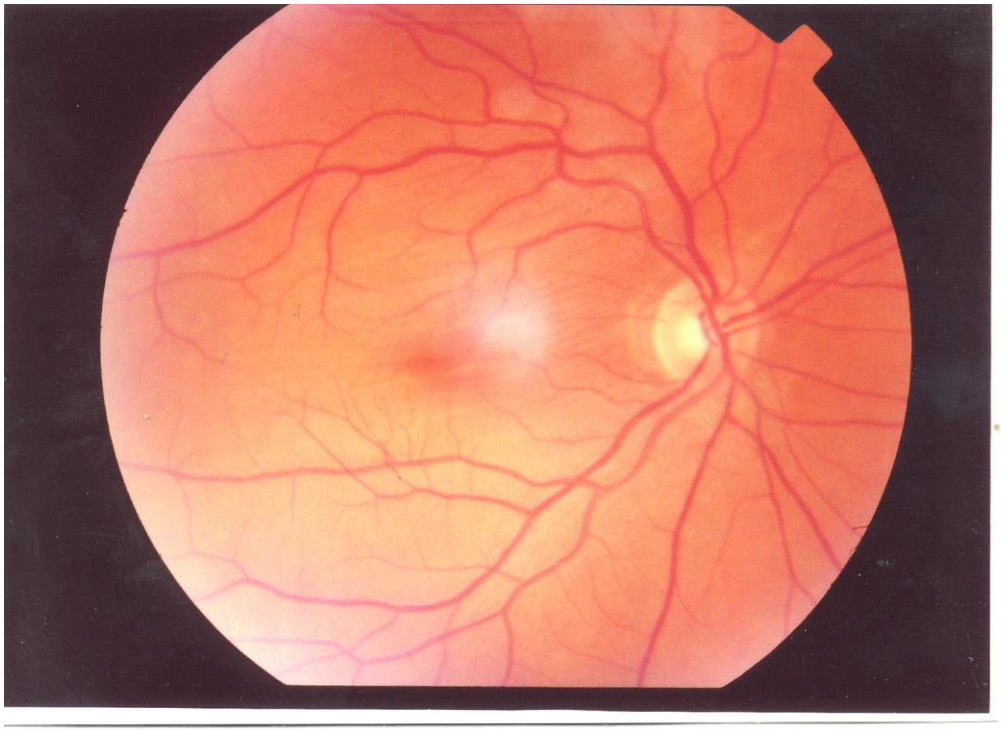


Figure 1-B: Fundus flouressein angiography of the right eye

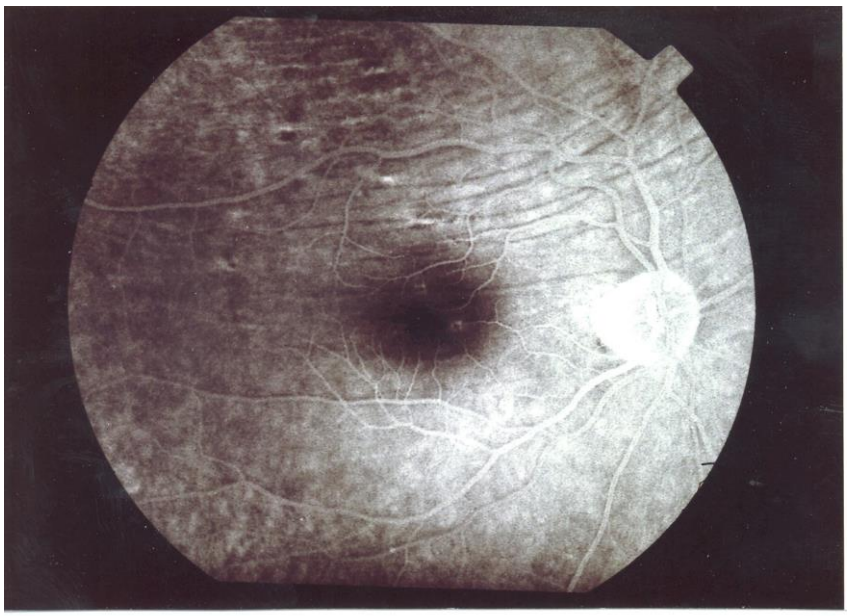


Figure 2-A1-3: Preoperative view of the patient in primary position and in looking up and down

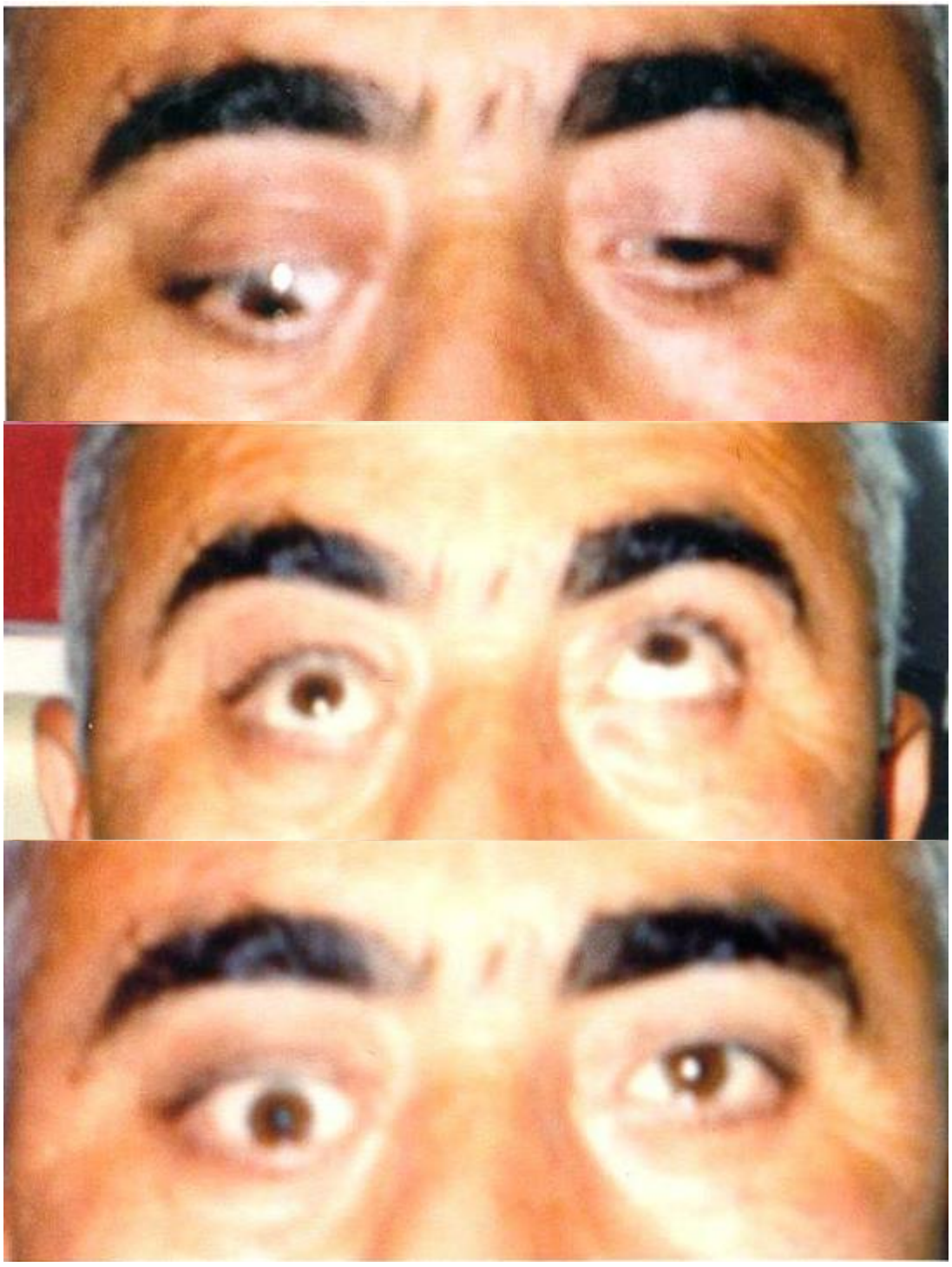
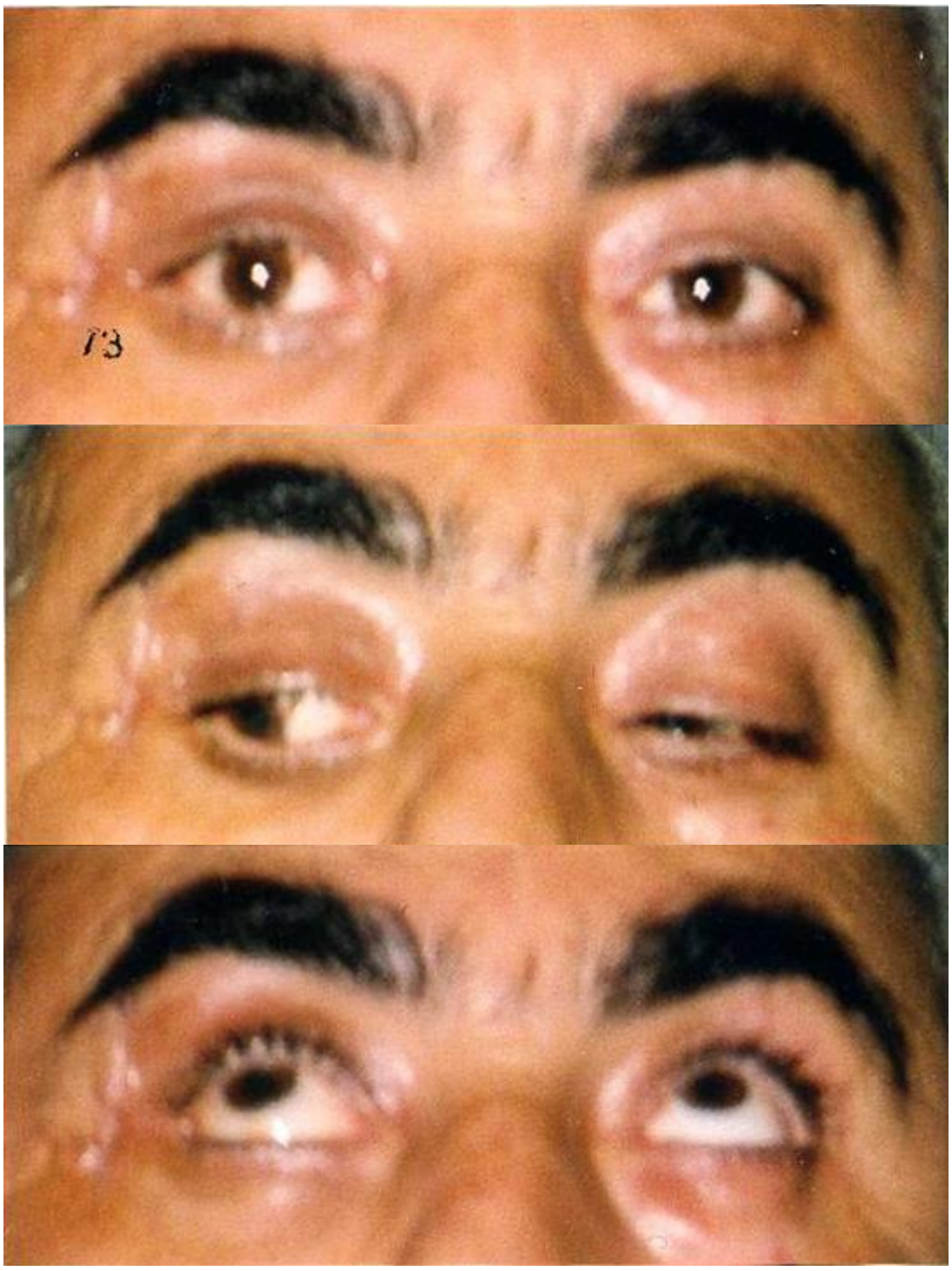


Figure 2-B1-6: Postoperative view of the paient in primary position, looking up, up and left, up and right, down and right positions



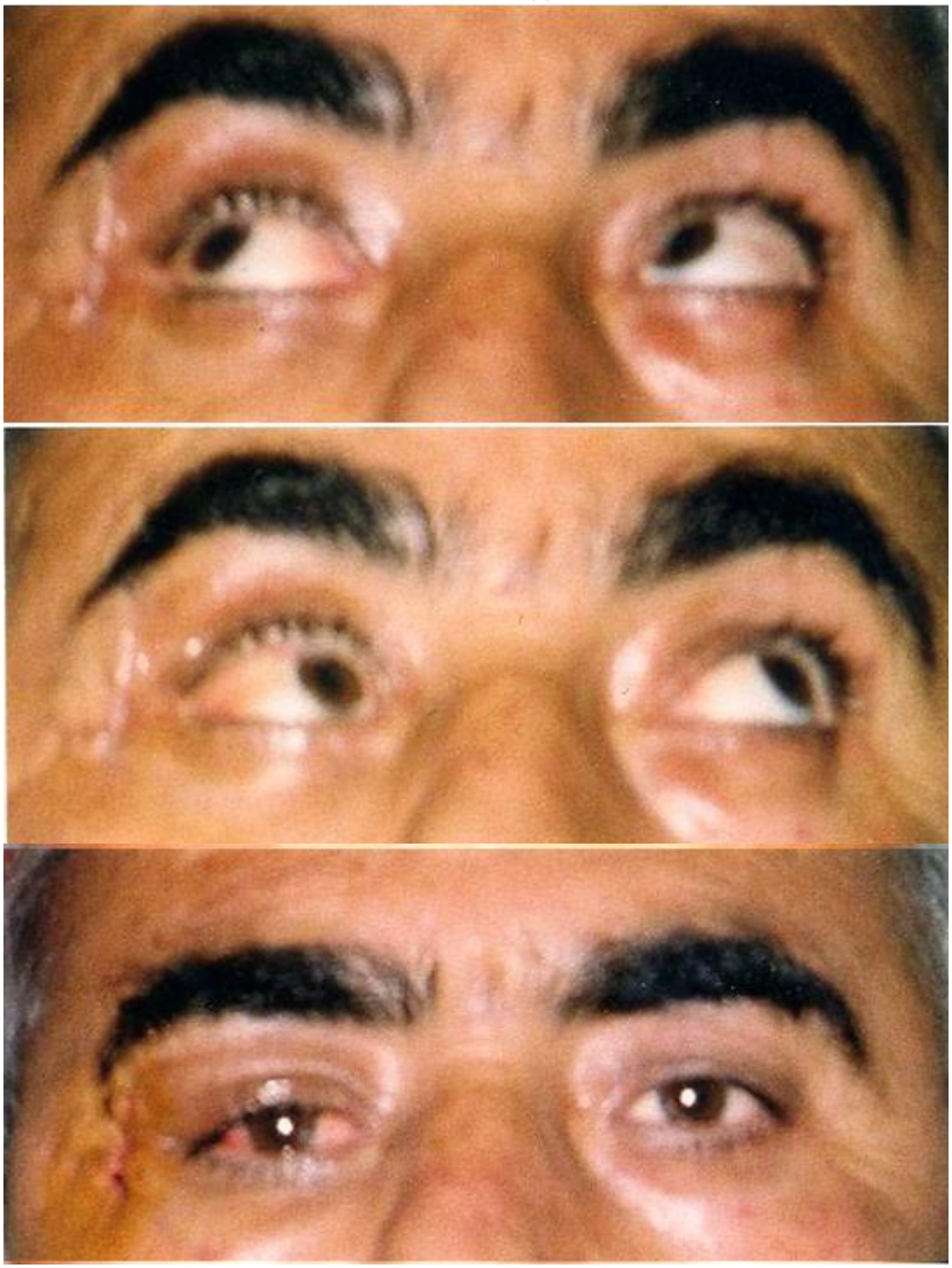


Figure 3: Preoperative view of the tumor in coronary computed tomography

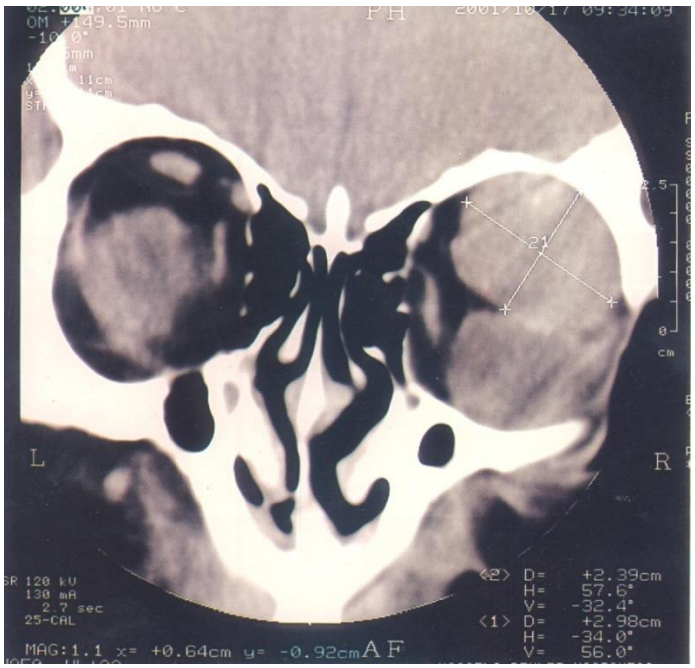


Figure 4: Histopathologic examination of the tumor. Glandular structures in fibrotic stroma (Hematoxylin-Eosin x 10)

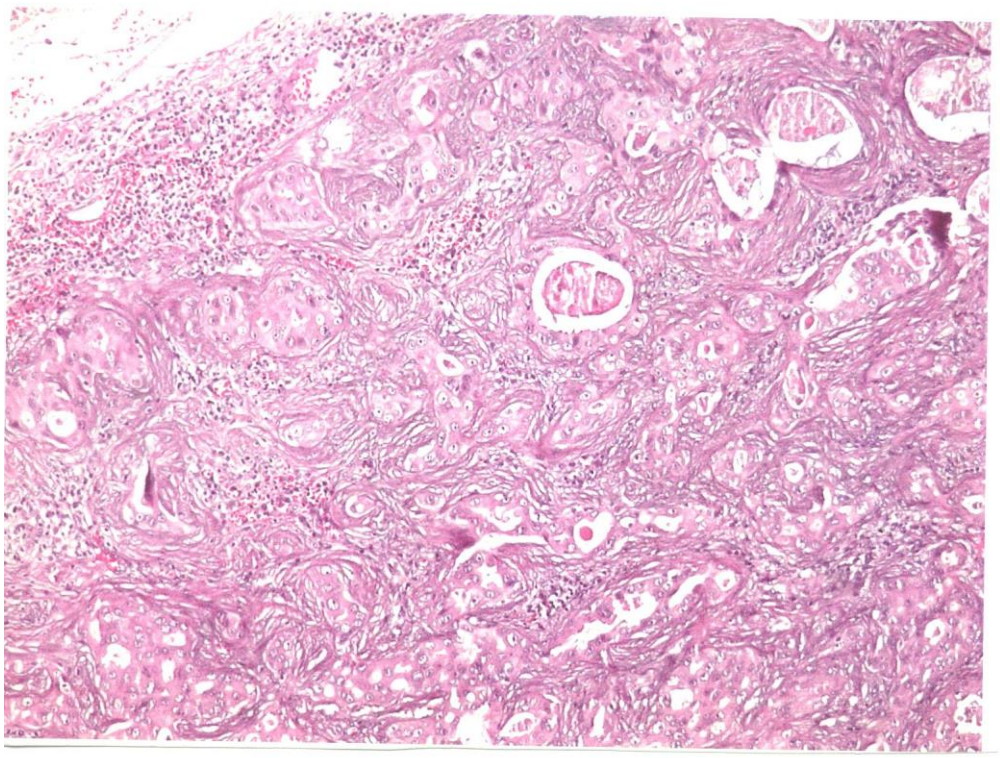


Figure 5: Postoperative view of the residual mass in coronary computed tomography

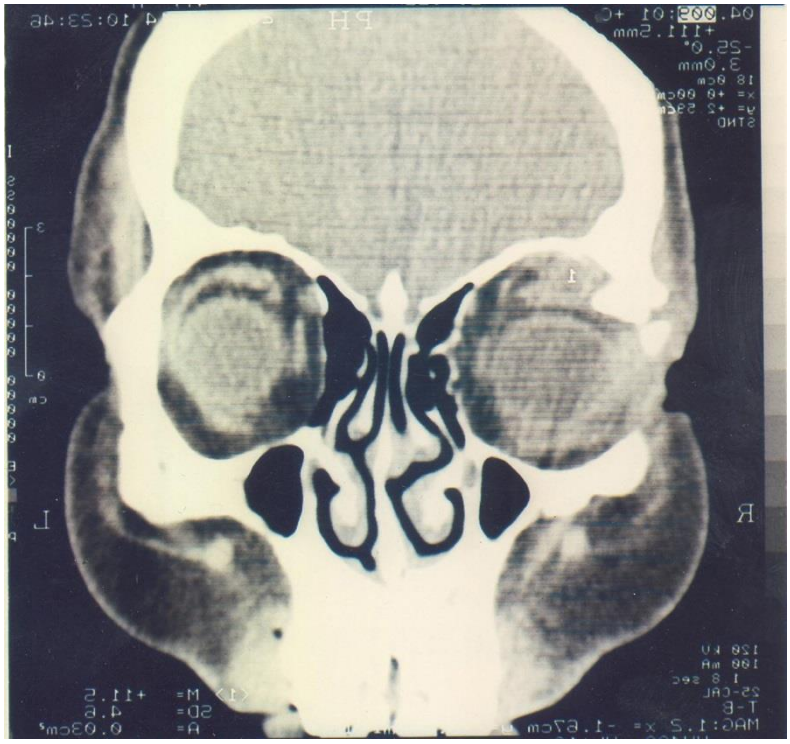


Figure 6: After radiation therapy, the regression of residual mass was seen in controle coronal computed tomography.