

## Solitary Fibrous Tumour of the Lacrimal Gland with Apparent hemangiopericytoma – Like Characteristics: A Case Study

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### Abstract

Hemangiopericytoma (HPC) is a rare tumor originating from the mesenchyme. We will describe the clinical presentations, radiological and operative findings, and pathological features of a patient with lacrimal gland HPC. The patient was a 26-year-old female who presented with swelling of the left upper eye lid progressing over the past 12 months. On ophthalmological examination, no visual impairment was detected, but induration of the left eye was found in the area of the lacrimal gland. Orbital magnetic resonance imaging showed tumorous mass, of size 20 × 8 × 22 mm affecting the lacrimal gland. The patient underwent an anterior orbitotomy, which removed the palpebral portion of the lacrimal gland. Histology did not clarify the diagnosis. Three months after surgery, the tumor size had increased. MRI showed progression of the findings. It was therefore decided to perform a lateral orbitotomy with radical extirpation of the tumor. The morphology of the tumor, including positivity of the immunohistochemical marker CD34, showed predominant characteristics of the HPC. The patient has been subject to a follow-up for the past 84 months and no signs of recurrence or metastases were observed during this period.

**Keywords:** Hemangiopericytoma; Lacrimal gland; Radical extirpation

### Introduction

Hemangiopericytoma (HPC) is a rare mesenchymal tumor thought to originate from the pericytes, first identified by Zimmermann [1]. Other investigators believe that HPCs arise from pluripotential perivascular cells, so they can occur anywhere the capillaries are found [2].

Not all HPCs are typical, and some present overlapping features with other vascular and mesenchymal tumors. Immunohistochemistry helps by excluding other differential diagnoses, but some cases with unusual findings can pose a challenge to general pathologists [3].

Differential diagnosis must consider the following disorders - chronic inflammation, Mikulicz's or Sjogren's syndrome, histiocytosis, pseudotumor, lymphoma.

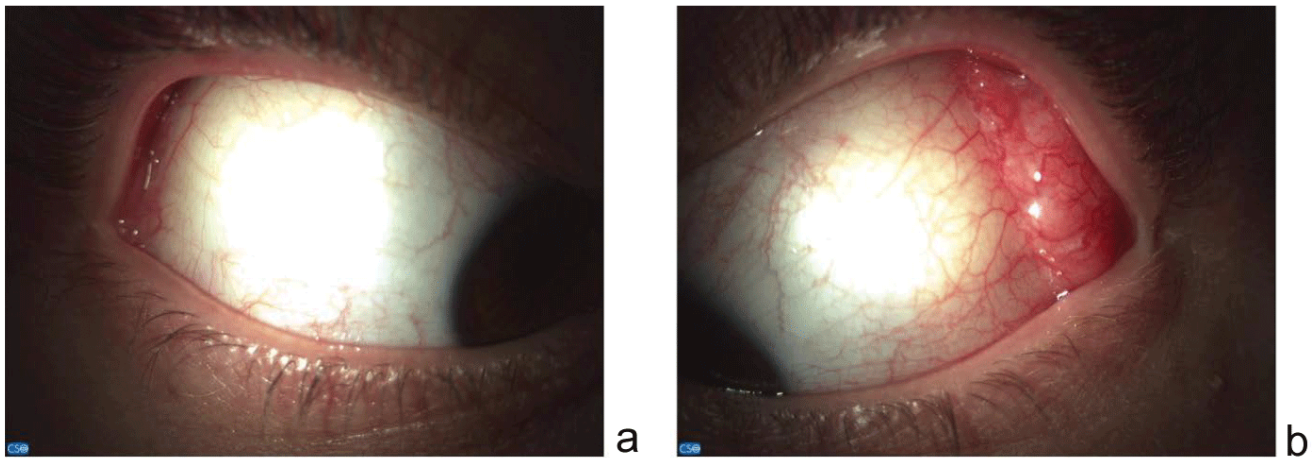
HPC is found in various locations, including the orbit. Orbital HPC is often described as synonymous with solitary fibrous tumor (SFT), giant cell angiofibroma (GCA), and fibrous histiocytoma, as they all belong to a spectrum of collagen-rich fibroblastic tumors that are often CD34-positive and have overlapping histopathologic features [4].

In their work, the Korean authors published a case report of a solitary fibrous tumor in the lacrimal gland, which is the eighth case described in available literature [5]. Our case report presents the ninth case of lacrimal gland HPC.

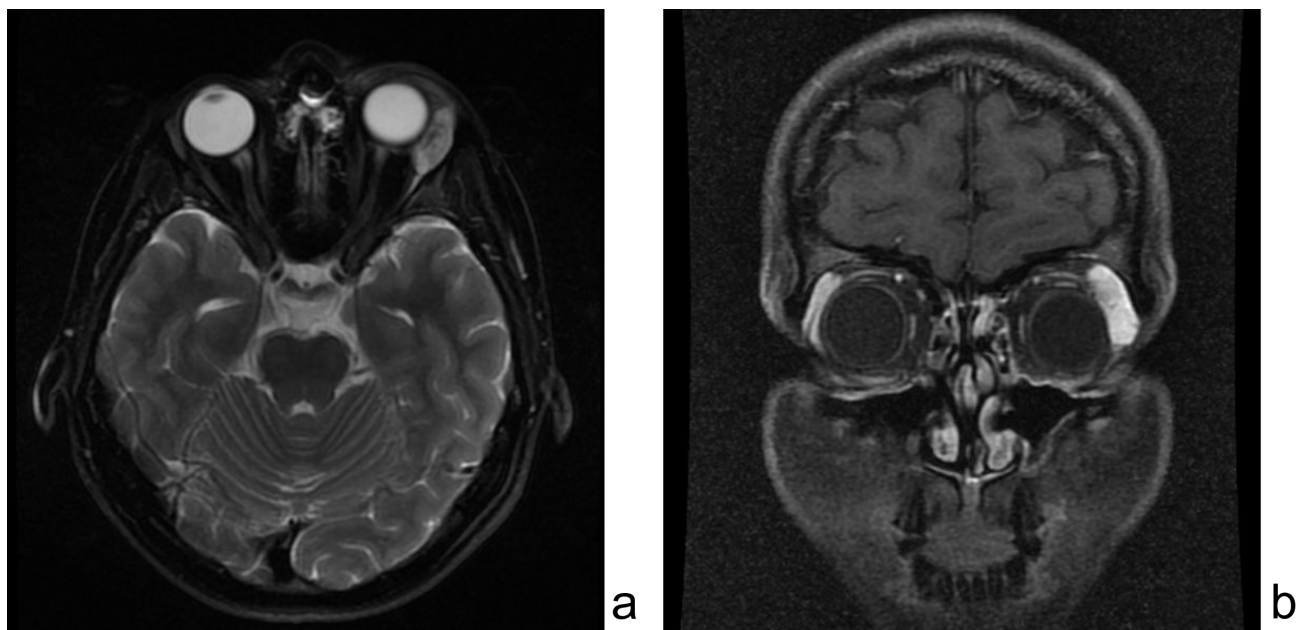
### Case Report

A 26-year-old, otherwise healthy, woman noticed swelling of her left upper eyelid and conjunctivitis at the age of 24. She was examined by her ophthalmologist, who recommended local corticosteroids and antibiotics. The condition did not show any improvement and the patient was then switched to systemic corticosteroids, which resulted in a temporary improvement of upper eye lid oedema. With respect to long-term worsening of the condition (progression over the past 12 months), the patient was referred to our facility for assessment of the lacrimal gland induration. During examination, the non-inflammatory swelling of the outer third of the left upper eyelid was found. It was soft and painless on palpation. Inverting and lifting the eyelid revealed bulging tumorous lobate formation in the palpebral portion of the lacrimal gland. Conjunctival vessels were dilated over the prominence. Ocular findings, including the position of the eye, its motility and intraocular findings were within physiological limits (Figure 1).

For an accurate diagnosis we performed MR examination of orbits, which showed an enlarged left lacrimal gland (20 × 8 × 22 mm, 1.8 ccm). This gland was of inhomogenous high signal on T<sub>2</sub> weighted images and homogenous low signal on T<sub>1</sub> weighted images. After intravenous administration of gadolinium contrast media, there was homogenous high increase of signal intensity in enlarged left lacrimal gland. The gland was sharply margined and infiltration of surrounding tissues was not observed (Figure 2).



**Figure 1:** (a) Image of the upper external quadrant of the right eye-physiological findings, (b) tumor masses in the palpebral portion of the lacrimal gland of the left eye. Images were made on the day of the first examination.



**Figure 2:** (a) Axial T<sub>2</sub> weighted MR image with fat suppression and (b) coronal T<sub>1</sub> weighted post contrast MR image with fat suppression showing tumor mass in the left lacrimal gland area.

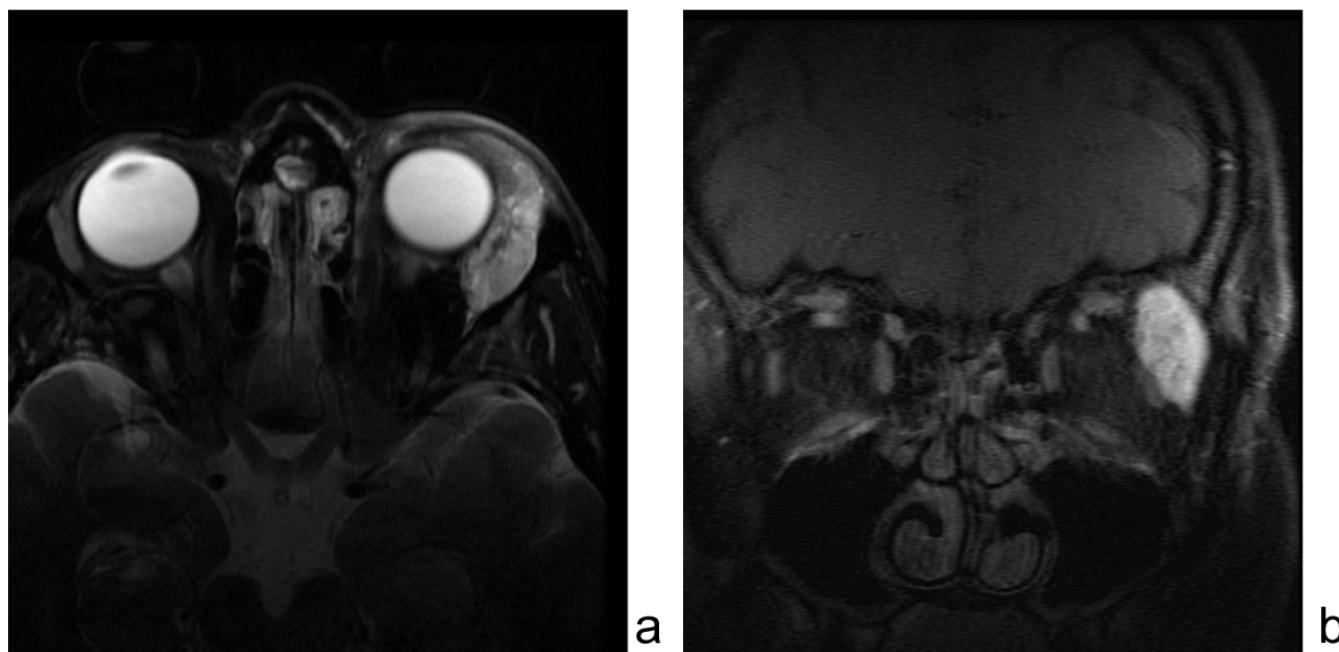
Because the clinical findings showed no signs of pleomorphic adenoma, carcinoma or lymphoma, we decided to perform a partial surgical removal of the tumorous mass in order to refine the diagnosis. After peritomy of conjunctiva in its outer half, we resected the whole palpebral portion of the lacrimal gland, which showed no macroscopic signs of pathological changes. The procedure lasted 45 minutes and was performed under uncomplicated anaesthesia.

The size of the biopsy sample was 10 × 10 mm. The pathological microscopy showed organoid tissue of the lacrimal gland with a very mild increase of round cell count in the stroma, typical accumulation of lymphocytes around the orifice of the lacrimal duct, which was

mildly dilated. The diagnosis remained unclear despite this examination.

The postoperative period was uneventful. Three months after the surgery, the tumor size increased and swelling of the outer third of the left upper eye lid recurred.

The follow-up MR examination showed progress of enlargement of the left lacrimal gland (29 × 12 × 24 mm, 4.2 ccm). In homogenous increased signal of the gland on T<sub>2</sub> weighted MR images and homogenous post contrast strong enhancement on T<sub>1</sub> weighted MR images were unchanged comparing the previous MR examination (Figure 3).



**Figure 3:** Follow-up MR examination performed 4 months after the initial one demonstrated tumor progression. (a) Axial T<sub>2</sub> weighted image with fat suppression, (b) coronal T<sub>1</sub> weighted image with fat suppression after contrast media administration.

Clinically, we observed swelling of the outer third of the upper eyelid. The palpation showed painless soft swelling. The axial protrusion of the left eye was 4 mm in comparison with the right eye. After lifting the upper eyelid, tumor masses had an appearance that was similar to the initial findings. Conjunctival hyperaemia was present above the swelling. Other ophthalmological findings were within physiological limits.

Although the patient was without any subjective symptoms, we decided to perform radical surgery, one year after the initial surgery. During lateral orbitotomy (according to Krönlein), the whole lacrimal gland was removed. Its parenchyma was tender and haemorrhagic. The tumor size was 30 × 20 × 10 mm. The course of the upper eyelid elevating muscle was monitored during the surgery. The temporal rectus muscle was not affected by the surgery (Figure 4).

Intraoperative biopsy ruled out lymphoma, but the pathologist expressed a suspicion of a mesenchymal tumor. The surgery lasted two hours and was without complications. It was performed under uneventful general anaesthesia (Figure 4). Bleeding occurred during the first postoperative day and a revision of the surgical wound was performed. A subsequent postoperative course was without complications.

Histological examination revealed a mesenchymal tumor with non-uniform microscopic structure. Sections of the tumor with decreased cellularity and local hyaline connective tissue transformation alternated with sections with increased cellularity and branching staghorn vessels. The tumor cells were oval or spindle-shaped, without significant polymorphism. Numerous small blood vessels dominated certain parts of the tumor. KI 67 proliferative activity was low. The

morphology of the tumor, including CD34 immunohistochemical positivity showed predominant characteristics of a HPC (Figure 5).

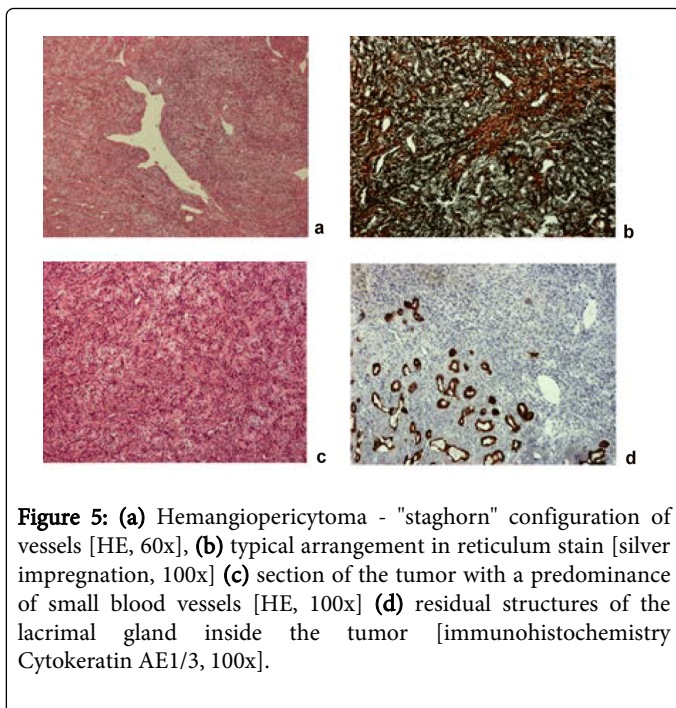
The follow-up MRI performed one year after the second surgery showed physiological findings.

The patient has been under regular monitoring and no signs of recurrence or metastasis have been observed since the second surgery 84 months ago. A decreased production of tears in the operated eye is compensated for by the application of artificial tears.



**Figure 4:** Intraoperative finding of a HPC with the size of 30 × 20 × 10 mm, which was radically removed through lateral orbitotomy.





**Figure 5:** (a) Hemangiopericytoma - "staghorn" configuration of vessels [HE, 60x], (b) typical arrangement in reticulum stain [silver impregnation, 100x] (c) section of the tumor with a predominance of small blood vessels [HE, 100x] (d) residual structures of the lacrimal gland inside the tumor [immunohistochemistry Cytokeratin AE1/3, 100x].

## Discussion

Haemangiopericytoma of the lacrimal gland is a very rare disease. The HPC case which we presented, ninth according to the literature, confirms this fact. Diagnostic difficulties are therefore justifiable.

Sullivan et al. [6] summarized typical characteristics of HPC as the following combination of clinical and radiological features suggestive of hemangiopericytoma. 1. Painless non-axial proptosis with downward displacement of the globe. 2. Intermittent upper lid swelling. 3. A soft, superiorly located mass with poorly defined borders, especially with a blue hue. 4. A superiorly located, rounded or elongated extraconal mass on CT, isodense with brain, with smooth, well-defined borders and moderate to marked enhancement with the injection of intravenous contrast medium. 5. Significant blush in all three phases of carotid angiography, without prominent arteriovenous shunting. Once hemangiopericytoma is suspected, complete surgical excision is recommended.

Our patient presented a similar course of the disease.

On computerized tomography, it generally appears as a well-circumscribed mass, which dramatically enhances after an injection of contrast material. On magnetic resonance images, it is isointense in  $T_1$ , while in  $T_2$  it can show slightly high intensity [3].

Meyer et al. describe HPC as a solitary tumor that tends to recur, and in some cases, may be malignant [7]. Our patient remained without recurrence, after the radical removal of the HPC 84 months ago.

Treatment of HPC affecting the orbit depends on the size of the tumor. Surgical removal may be associated with an exenteration of the orbit [8] or radical excision of the tumor [9]. Pihlblad and Schaefer described the elimination of the tumor using embolization and subsequent extirpation after biopsy confirmation of HPC [10]. Setzkorn et al. reported two recurrences, which were treated surgically, with the third relapse treated by radiotherapy. The patient was still

disease free 78 months after radiotherapy and experienced no side effects [9]. Due to the rarity of HPC in the orbit, radiotherapy is used in the case of recurrence [9,11].

Earlier reports in literature describe metastases of orbital HPCs in approximately 12% - 45% of cases [12]. The most common sites of metastases are the lung, mediastinum, liver, and bone. Unfortunately, the aggressive behavior of HPC is unpredictable, as it happens that apparently benign tumors eventually spread systemically. Nevertheless, some criteria has been used to classify a tumor as malignant: large diameter (5 cm), increased mitotic rate (four or more figures per ten high-power fields), a high degree of cellularity, immature and pleomorphic neoplastic cells, foci of necrosis, and hemorrhage [13,14].

Treatment of our patient was also radical, and the patient remained without signs of recurrence in the long term.

## Conclusion

Lacrimal gland hemangiopericytoma is a very rare disease (nine cases were described).

Literature data regarding its behavior in the lacrimal gland is poor. Presented data suggest that patients with hemangiopericytoma, whose biological properties are difficult to predict, must be subject to long-term clinical and radiological monitoring.

MRI, arteriography (DSA) may contribute to preoperative diagnosis. The basic therapeutic method involves the removal of the affected gland while maintaining the function of surrounding anatomical structures. Further biological behaviour of HPC can be predicted based on the KI 67 proliferation index. HPC located in the orbit tends to relapse and metastasis according to the literature. Patients with these tumors must be therefore under long-term clinical and radiological monitoring. If all conditions allow, we will clearly prefer surgical treatment with complete removal of the tumor in the future.

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## Conflicts of Interest

No potential conflict of interest relevant to this article was reported.

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