

# Solitary fibrous tumor of the lacrimal gland mimicking pleomorphic adenoma

## *Tumor fibroso solitário de glândula lacrimal simulando adenoma pleomórfico*

EDUARDO DAMOUS FEIJÓ<sup>1</sup>, ANA CARLA DE SOUZA NERY<sup>2</sup>, FÁBIO RAMOS CAIADO<sup>3</sup>, ROBERTO MURILLO LIMONGI<sup>4</sup>

### ABSTRACT

Solitary fibrous tumor (SFT) is a rare mesenchymal spindle-cell neoplasm commonly found in the pleura; it is rare in the orbit and extremely rare in the lacrimal gland. We herein report a case of SFT of the lacrimal gland that mimicked a pleomorphic adenoma. We discuss the clinical, radiological, histopathological, and immunohistochemical findings that provided insight and rationale to accurately diagnose this case.

**Keywords:** Lacrimal apparatus/pathology; Lacrimal apparatus diseases/pathology; Neoplasms/diagnosis; Solitary fibrous tumors; Adenoma; pleomorphic; Diagnosis; differential

### RESUMO

Os autores relatam um caso de tumor fibroso solitário (TFS) de glândula lacrimal simulando um adenoma pleomórfico. O TFS é um raro tumor mesenquimal de células fusiformes, comumente encontrado na membrana pleural, raramente encontrado na órbita e extremamente raro na glândula lacrimal. Os autores discutem os achados clínicos, radiológicos, histológicos e imunohistoquímicos que são a chave para o correto diagnóstico desta rara entidade.

**Descritores:** Aparelho lacrimal/patologia; Doenças do aparelho lacrimal/patologia; Neoplasias/diagnóstico; Tumor fibroso solitário; Adenoma pleomorfo; Diagnóstico diferencial

### INTRODUCTION

Solitary fibrous tumor (SFT) is a rare mesenchymal spindle-cell tumor that can be found at various sites, such as the liver, nasal cavity, orbit, thyroid, meninges, skin, and kidney. However, the most common site of SFTs is the intra-thoracic region<sup>(1-4)</sup>. Although it can affect the orbital region, SFT of the lacrimal gland and lacrimal fossa have rarely been observed and described<sup>(3-5)</sup>. SFT commonly affects middle-aged adults, with equal distribution between genders. SFTs are well circumscribed, and their variably pattern-less spindle-cell lesions and immunopositivity for CD34 led the World Health Organization to redefine orbital SFTs using the same nomenclature for hemangiopericytoma, giant-cell angiofibroma, and fibrous histiocytoma of the orbit<sup>(6-10)</sup>. Here, we describe the first case in Brazilian literature of lacrimal gland SFT mimicking pleomorphic adenoma in a 38-year-old woman, and we have demonstrated that a specific immunohistochemical (IHC) marker, signal transducer and activator of transcription 6 (STAT6), distinguishes SFTs from other tumors that mimic it histologically<sup>(8)</sup>.

### CASE REPORT

A 38-year-old woman presented with swelling in the left superolateral aspect of the orbit (lacrimal gland region) and an S-shaped proptosis for the past 4 years. There was gradual increase in lesion size over time. She had no history of trauma and had no ocular complaints. External examination revealed a palpable tumor in the orbit, specifically in the lacrimal gland. During examination, the mass was soft and did not appear to be attached to the periorbital rim. There

was no proptosis. The Valsalva test was negative, and extraocular motility was normal (Figures 1A and 1B). Magnetic resonance imaging (MRI) revealed an extraconal solid nodule with defined, precise limits in the left lacrimal gland and intense heterogeneous enhancement by contrast, measuring about 2 × 2 × 1.4 cm (Figures 1C and 1D).

The mass was fully excised via a lateral orbitotomy approach, and a macroscopic presumptive diagnosis was typical pleomorphic adenoma. The excised tumor was ovoid and firm, measuring 2 × 2 × 1.5 cm. There were no bone changes caused by the tumor as determined via surgical exploration. Histopathological analysis of the tumor revealed mesenchymal neoplasia with a spindle-cell proliferative lesion and hemangiopericytoma-like pattern encircled by the lacrimal gland parenchyma. Mitotic activity was low. Histomorphology was suggestive of a benign spindle-cell tumor (Figures 2A and 2B). IHC showed that the spindle cells were highly positive for CD34 and STAT6 (Figure 3). S-100 protein and smooth muscle actin were not detected. An SFT was diagnosed based on these IHC and histological findings. During 6 months of follow-up there was no tumor recurrence.

### DISCUSSION

SFT is a rare tumor, originally presumed to be derived from the mesothelium, that affects middle-aged adults, with no apparent sex predilection<sup>(1-5)</sup>. However, these tumors are not often associated with serosal surfaces, such as the nasal cavity and paranasal sinuses, but rather are of a mesenchymal origin.

The orbit is an unusual site for SFT occurrence and generally presents with a slow and non-aggressive course, possibly achieving

Submitted for publication: January 23, 2016  
Accepted for publication: November 7, 2016

<sup>1</sup> Ocular Plastic Sector, Hospital Oftalmológico de Anápolis, Anápolis, GO, Brazil.

<sup>2</sup> Ocular Plastic Sector, Instituto Panamericano da Visão, Goiânia, GO, Brazil.

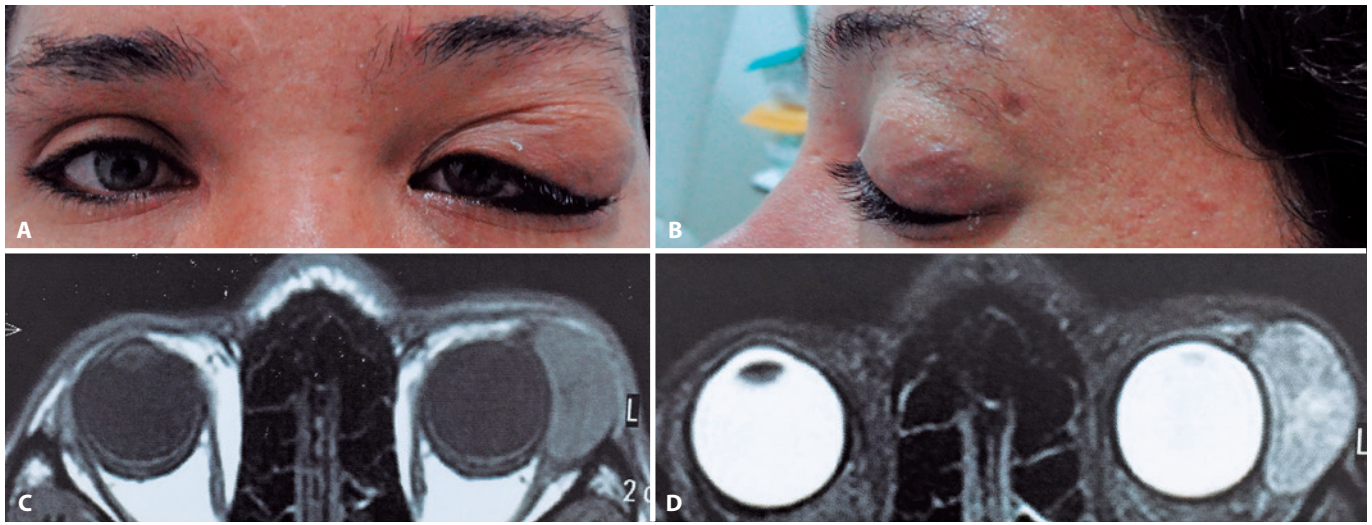
<sup>3</sup> Hospital Oftalmológico de Anápolis, Anápolis, GO, Brazil.

<sup>4</sup> Ocular Plastic Sector, Centro de Referência em Oftalmologia, Universidade Federal de Goiás, Goiânia, GO, Brazil.

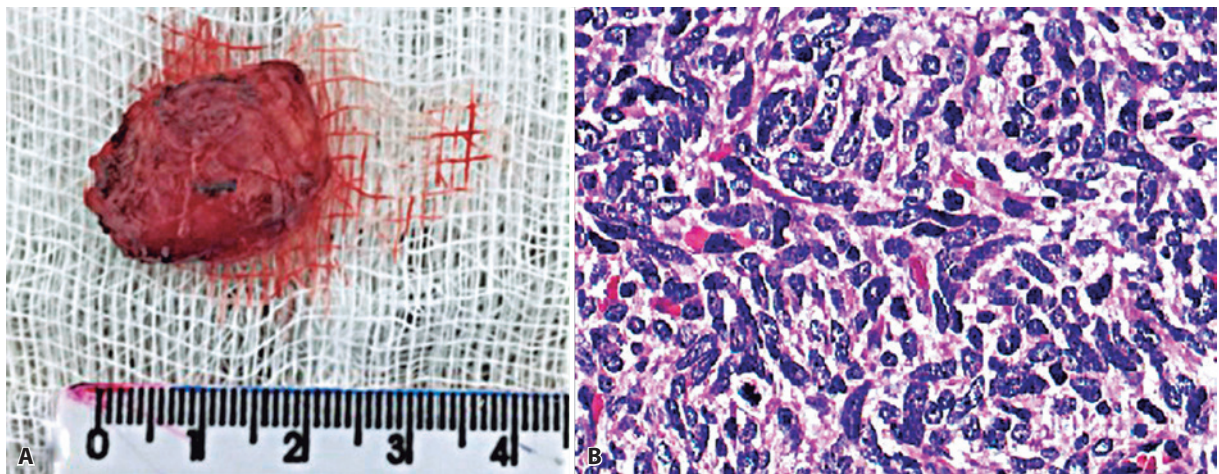
**Funding:** This study was not financially supported.

**Disclosure of potential conflicts of interest:** The authors declare no conflicts of interest.

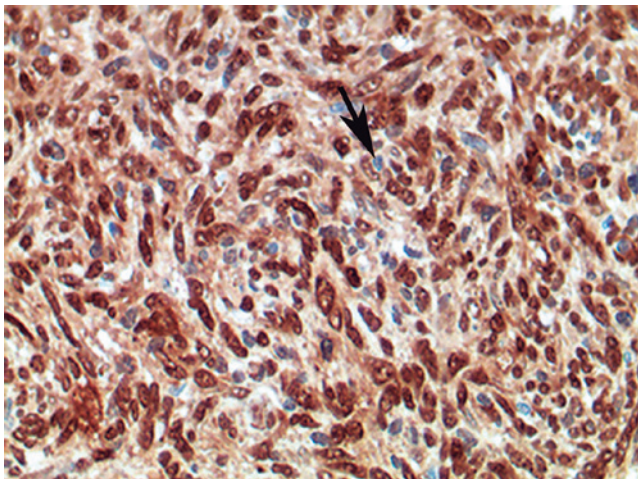
**Corresponding author:** Eduardo D. Feijó. Av. Faiad Hanna, 235 - Anápolis - GO - 75080-410 - Brazil - E-mail: eduardodff@yahoo.com.br



**Figure 1.** A) Tumor in superolateral region of the orbit, as determined via lacrimal gland topography. B) Side view. C and D: MRI revealed an extraconal solid nodule with defined limits in the left lacrimal gland, with intense heterogeneous enhancement by contrast, measuring about 2 × 2 × 1.4 cm, suggestive of pleomorphic adenoma of the lacrimal gland.



**Figure 2.** A) Macroscopic aspect of the tumor, well demarcated, ovoid, and firm, with measurements of 2 × 2 × 1.5 cm. B) The histopathological analysis of the tumor revealed a mesenchymal neoplasm with a spindle-cell proliferative lesion and hemangiopericytoma-like pattern as determined via hematoxylin-eosin staining.



**Figure 3.** Immunohistochemistry demonstrated that spindle-cell nuclei strongly expressed STAT6 (black arrow).

sizes of up to 4 cm<sup>(3-4)</sup>. Our patient also had a slow-growing SFT, with a duration of 4 years and 2 cm in size. In this case, the external examination and MRI suggested pleomorphic adenoma of the lacrimal gland, which is the most common neoplasia of the lacrimal gland.

Dorfman et al.<sup>(4)</sup> also reported three cases of orbital region SFTs in a 44-year-old man, 65-year-old woman, and 69-year-old woman, with MRI and histopathological findings comparable with our case. However, our patient was younger; a 38-year-old woman.

Histopathological analysis has prognostic as well as diagnostic value. SFTs associated with hypercellularity, pleomorphism, and a high mitotic count (2:4 per 10 high magnification fields) are typically malignant, with relapse and distant metastases as possibilities. Vallat-Decouvelaere et al.<sup>(9)</sup> suggested that nuclear atypia, mitosis in 4:10 magnification fields, and necrosis in addition to hypercellularity are considered malignant features of SFTs. In our case, the mitotic count was two per 10-high-power fields. Nuclear pleomorphism, necrosis, or an invasive growth pattern were not observed in this study, suggesting a benign SFT.

Differential diagnoses according to histopathological findings include fibrous histiocytoma, schwannoma, and hemangiopericytoma.



The differential diagnosis between hemangiopericytoma and SFTs is extremely important, because hemangiopericytoma has an aggressive clinical course with high relapse and higher rate of metastasis in contrast to the general benign nature of SFTs<sup>(3-5)</sup>. Hemangiopericytoma classically consists of oval to spindle-shaped cells arranged around vessels. For definitive diagnosis, IHC is crucial. In most cases, an IHC of hemangiopericytoma shows a weak positivity to CD34 and a strong positivity to vimentin. Strong and consistent CD34 immunoreactivity in SFTs is critical to ensuring correct diagnosis<sup>(10)</sup>. The present case showed spindle cells exhibiting strong immunoreactivity to CD34 and nuclear expression of STAT6<sup>(3,4-6)</sup>.

Recent molecular analyses have suggested that most SFTs express STAT6, and thus it is considered a specific marker to this neoplasia. Doyle et al.<sup>(8)</sup> investigated STAT6 nuclear expression by IHC in SFTs outside the central nervous system. They evaluated 60 SFT cases, and 59 (98%) of these cases displayed STAT6 nuclear expression, thus, clearly demonstrating it as a highly sensitive and specific marker for SFTs<sup>(8)</sup>. As aforementioned, SFT is a rare tumor in the lacrimal gland region. Histopathological examination along with IHC findings are essential for correct diagnosis<sup>(6)</sup>, and STAT6 is a suitable IHC marker to distinguish SFTs from other tumors that mimic it histologically, as clearly shown in this case report.

## REFERENCES

1. Ha JK, Park BJ, Kim YH, Lim YJ. Orbital solitary fibrous tumor: a case report and diagnostic clues. *J Korean Neurosurg Soc.* 2009;46(1):77-80.
2. Guillou L, Gebhard S, Coindre JM. Orbital and extraorbital giant cell angiofibroma: a giant cell-rich variant of solitary fibrous tumor? Clinicopathologic and immunohistochemical analysis of a series in favor of a unifying concept. *Am J Surg Pathol.* 2000;24(7):971-9.
3. O'Donovan DA, Bilbao JM, Fazl M, Antonyshyn OM. Solitary fibrous tumour of the orbit. *J Craniofac Surg.* 2001;13(5):641-4.
4. Dorfman DM, To K, Dickersin GR, Rosenberg AE, Pilch BZ. Solitary fibrous tumour of the orbit. *Am J Surg Pathol.* 1994;18(3):281-7.
5. Son H, Yoo SH, As HS, Cho KJ. A solitary fibrous tumor with giant cells in the lacrimal gland: a case study. *Korean J Patholol.* 2013;47(2):158-62.
6. Yoshida A, Tsuta K, Ohno M, Yoshida M, Narita Y, Kawai A, et al. STAT6 immunohistochemistry is helpful in the diagnosis of solitary fibrous tumours. *Am J Surg Pathol.* 2014;38(4):552-9.
7. Hasegawa T, Matsuno Y, Shimoda T, Hasegawa F, Sano T, Hirohashi S. Extrathoracic solitary fibrous tumors: their histological variability and potentially aggressive behavior. *Hum Pathol.* 1999;30(12):1464-73.
8. Doyle LA, Vivero M, Fletcher CD, Mertens F, Hornik JL. Nuclear expression of STAT6 distinguishes solitary fibrous tumor from histologic mimics. *Mod Pathol.* 2014;27(3):390-5.
9. Vallat-Decouvelaere AV, Dry SM, Fletcher CD. Atypical and malignant solitary fibrous tumors in extrathoracic locations: evidence of their comparability to intra-thoracic tumors. *Am J Surg Pathol.* 1998;22(12):1501-11.
10. Alves AP, Felix PR, Cruz AA. Hemangiopericitoma de órbita. *Arq Bras Oftalmol.* 2001; 64(2):159-62.

# Congresso Baiano de Oftalmologia

19 a 21 de outubro de 2017

Hospital São Rafael  
Salvador - BA

## Informações:

Tel.: (71) 3245-3955  
E-mail: sofba@sofba.com.br