Clinicopathological Characteristics of Three Cases with Recurrent Orbital Solitary Fibrous Tumors: A Retrospective Study and Literature Review

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Abstract

Purpose Solitary fibrous tumor (SFT) is a spindle cell neoplasm that rarely occurs in orbit. This study aimed to report clinical, imaging, and pathological features of three patients with recurrent orbital SFTs.

Methods Clinical, imaging, and pathological data of the three patients were retrospectively reviewed, and the results were compared with those of previously reported cases with recurrent orbital SFT.

Results One female and two male patients (mean age, 54 years old) were included in this study. The present cases and literature review showed that the average time to recurrence in patients who aged under 50 years old was shorter than that in those who aged over 50 years old. The most common site for recurrent orbital SFT was the retrobulbar area of the orbit (23.8%). Imaging examinations showed consistent intensity of MRI signals before and after recurrence. Immunohistochemical results of all cases revealed the expressions of CD34. The mitotic rate increased in 4/8 cases, and the percentage of Ki-67-positive cells was elevated in 5/16 cases.

Conclusion These results suggested that young patients were more likely subjected to recurrent orbital SFT. The postoperative pathological diagnosis revealed that patients with recurrent orbital SFT had more nuclear abnormalities and mitotic activity, as well as a higher percentage of Ki-67-positive cells, indicating that orbital recurrent SFT tended to be malignant according to both morphological features and immunohistochemistry results.

Introduction

Solitary fibrous tumor (SFT) is a spindle cell neoplasm that was first presented in 1931 by Klemperer and Rabin, who reported it as a primary neoplasm of the visceral pleura [1, 2]. SFT demonstrates a different cellular proliferation of bland spindle-shaped cells lacking any pattern of growth, with associated 'ropey' keloidal collagen bundles and characteristic 'staghorn' blood vessels [3]. The hemosiderotic fibrolipomatous tumor is histologically characterized by the presence of mature adipose tissue and spindle cell component accompanied by hemosiderin pigment deposition within macrophages, in the cytoplasm of some of the spindle cells and also within the extracellular stroma. The spindle cell component of these lesions is generally positive for CD34 and negative for CD68, S100, SMA and desmin [4, 5]. The biological course of SFT tends to be benign in nature, while some cases with recurrent SFT and its malignant transformation have been reported in recent years.

SFT of orbit is uncommon and recurrent orbital SFT is even less. To date, no large sample-sized controlled trial on the pathological features of recurrent orbital SFT has been conducted. In the present research, clinical data of 3 patients with recurrent orbital SFT, who were confirmed by pathology, were retrospectively analyzed. The clinical, imaging, and pathological data of patients with two or more episodes were compared. To increase our understanding about the recurrent orbital SFT, literature review of the clinicopathological manifestations of cases with recurrent orbital SFT, including tumor necrosis, mitotic rate, and the percentage of Ki-67-positive cells, was carried out via the PubMed database.

Materials And Methods

Case Series

A retrospective study was performed on 3 patients with recurrent orbital SFT, who were confirmed by pathology and were treated at Ophthalmology Department of the First Affiliated Hospital of Zhejiang University School of Medicine (Hangzhou, China) between 2015 and 2020. Clinical data, including sex, age at the time of diagnosis, recurrence time, symptoms, location and size of the tumor, and treatment history were collected. Imaging examinations mainly included orbital magnetic resonance imaging (MRI) and computed tomography (CT) data. Only Case #1 and Case #2 underwent MRI in our hospital. Pathological examination was conducted using collected specimens, and paraffin-embedded sections were analyzed by histopathology and immunohistochemistry (IHC). IHC results included the expressions of CD34, CD99, CK, SMA, EMA, S-100, and the percentage of Ki-67-positive cells.

The current study was approved by the Ethics Committee of the First Affiliated Hospital of Zhejiang University School of Medicine, and was performed in accordance with the Declaration of Helsinki. Besides, all participants signed the written informed consent form prior to commencing the study.

Literature Review

The PubMed database was used to search for the related literature on the recurrent orbital SFT published from January 1990 to April 2021. The search terms were (“recurrent”) AND (“orbital SFT” OR “orbital solitary fibrous tumor”). A total of 21 articles, which included 45 cases, were retrieved [3, 4, 6–23].

Results

Clinical Features

Two male patients and one female patient were included in the current study. The median age at the time of their first diagnosis was 54 (range, 48–60) years old. Their clinical manifestations included exophthalmos (n = 3), ptosis (n = 2), incomplete closure of the eyelids (n = 2), abnormal eye position (n = 1), and blurred vision (n = 2). On the results of physical examination revealed that the three patients involved in the present study exhibited a huge and incompressible mass behind the eyeball, making the eyeball protruded forward, with the protrusion degree of 22–23 mm compared with the degree of 15 mm for the other eye. The best corrected visual acuity (BCVA) was reduced to 1/20 in Case #1 and 2/20 in Case #2. The dilated conjunctival blood vessels of the involved eye were apparent (n = 3), while on fundus examination, the optic disc edema was raised with an unclear boundary and dilated blood vessels (n = 2), and bleeding could be observed around the optic disc (n = 1). The recurrence time after the first surgery ranged from 9 to 120 months. Two patients did not relapse during a follow-up period of 2 years after the second surgery. Case #3 had 6 relapses in total, he underwent surgery for 4 relapses, and in 2015, he received gamma knife radiosurgery for three times, however, the tumor recurred again in 2016. The patient died of systemic metastasis one year later.

Table 1 summarizes the clinical characteristics of 38 cases with recurrent orbital SFT collected from 18 previously published articles. The patients' mean age was 47 (range of age, 9–86) years old, and 17 (47.2%) patients aged under 50 years old. There were 18 male and 19 female patients, except one case whose sex could not be determined, and no significant difference ($P = 0.33$) was noted between male and female patients. The position of the lesion included superolateral or lacrimal gland fossa ($n = 3$), superomedial or upper eyelid ($n = 4$), inferotemporal ($n = 3$), medial canthal region or lacrimal sac ($n = 3$), postero-lateral ($n = 2$), retrobulbar ($n = 5$), and entire orbit ($n = 1$). Meanwhile, the duration of recurrence after initial surgery ranged from 2 to 240 months. The average time to recurrence after the first surgery for patients who aged ≤ 50 and > 50 years old was 34.4 and 57.1 months, respectively, which showed no significant difference ($P = 0.19$) between the two groups. In addition, 7 (21.2%) patients with SFT had multiple recurrences, and 4 (12.9%) patients had distant metastasis.

Table 1
Clinical manifestations of recurrent orbital SFT cases [3–22]

	Age	Gender	Position	Recurrence time(m)	Multiple recurrences	Transfer
Dorfman et al. (1994)	69	F	NM	48	Y	NM
Ing et al.(1998)	62	M	Entire orbit	240	N	Y
N de Saint Aubain et al. (1999)	68	M	NM	3	N	N
Hasegawa et al. (1999)	48	F	Upper eyelid	84	N	Y
Woo et al. (1999)	23	M	Right medial canthal region	4	N	N
	34	F	Right medial canthal region	84	N	N
Alexandrakis et al. (2000)	14	F	Superomedial	4	N	N
Carrera et al. (2001)	61	M	Superolateral	38	Y	N
Hayashi et al. (2001)	54	M	Inferolateral and intracranial	6	N	N
Polito et al. (2002)	24	M	Lacrimal gland fossa	48	N	N
	70	M	Retrobulbar	48	N	N
Bernardini et al. (2003)	59	F	Between the lacrimal sac and medial rectus muscle	24	N	N
Ness et al. (2005)	36	NM	Upper postero-lateral	24	N	NM
Tam et al. (2008)	20	F	Superomedial	3	N	N
Thomas et al. (2011)	62	F	Retrobulbar	36	N	N
Griepentrog et al.(2013)	9	F	Inferotemporal extraconal and intraconal	120	Y	N
Wang et al. (2013)	31	M	Postero-lateral	72	Y	N

Abbreviations: M, male; F, female; NM, not mentioned; Y, yes; N no

	Age	Gender	Position	Recurrence time(m)	Multiple recurrences	Transfer
Vu et al.(2016)	12	M	Superolateral	4	N	N
Krishnamurthy et al.(2016)	12	F	Superomedial	36	N	N
Smith et al. (2017)	41	F	NM	12	NM	NM
	58	M	NM	9	NM	NM
	58	M	NM	120	NM	NM
	39	M	NM	27	NM	NM
	15	F	NM	6	NM	NM
Sagiv et al. (2019)	36	F	NM	36	N	N
	64	M	Inferotemporal	6	N	Y
	72	M	NM	60	Y	Y
	57	F	NM	72	N	N
	86	F	NM	12	N	N
	59	F	NM	120	N	N
	48	M	NM	NM	N	N
	66	M	NM	108	Y	N
Lester et al. (2020)	37	F	NM	12	N	N
	55	F	NM	NM	N	Y
	64	F	NM	2	N	N
The present report	60	M	Retrobulbar	12	N	N
	48	F	Retrobulbar	9	N	N
	54	M	Retrobulbar	120	Y	Y
Abbreviations: M, male; F, female; NM, not mentioned; Y, yes; N no						

Radiology

Orbital MRI of Case #1 before and after recurrence revealed that there was a mass above the optic nerve in the posterior muscular cone of the left orbit, which was indistinguishable from the optic nerve. The internal signal was uneven, in which a slightly lower value of signal intensity was noted in T1-weighted image (T1WI) (Fig. 1a), a slightly higher value of signal intensity was found in T2-weighted image (T2WI)

(Fig. 1b), and an irregular enhancement could be observed after intravenous gadolinium administration (Fig. 1c). These results were consistent after recurrence (Figs. 1d-f).

In Case #2, the MRI revealed a huge heterogeneous multi-cystic mass occupied the entire posterior orbit, with a size of about 3.4 cm * 3.2 cm. In T1WIs, the intensity of signals was almost equal, while in T2WIs, the intensity of signals was slightly higher. The signals were diffusely and unevenly enhanced on contrast-enhanced MR images. These results were consistent with those observed 9 years before her admission to our hospital.

Besides, Case #3 underwent CT scan for two times in our hospital, and in 2016, the secondary CT scan showed soft tissue masses in the right-sided orbit, maxillary sinus, ethmoid sinus, and frontal sinus.

Pathological Characteristics

Gross pathology showed that the specimens were gray-white or gray-red oval-shaped tissues. Cystic cavities with different sizes were found inside the tumor in Case #2 before and after relapse, and the cystic fluid was yellow green.

Histopathology, in all cases before relapse, revealed that well-arranged spindle cells with oval-to-spindle nuclei and eosinophilic cytoplasm (Fig. 2a) were distributed in the dense collagen fibers with a great number of blood vessels in the background. Blood vessels occasionally showed a staghorn appearance (Fig. 2b). No obvious mitotic activity or necrotic areas were found. After relapse, the spindle cells were arranged irregularly with a higher frequency of chromosomal heteromorphism (Fig. 2c). The number of mitotic figures was > 4/10 high-power fields (HPFs) in the three cases (Fig. 2d). Besides, tumor necrosis was found after recurrence in Case #3.

Results of IHC in Case #1 and Case #3 showed the expressions of CD34, but no expressions of CD99, Bcl-2, CK, SMA, EMA, and S-100. IHC of Case #2 revealed that the tumor cells were positive for CD34 (Fig. 2e), CD99 (Fig. 2f), and Bcl-2 (Fig. 2g), while those were negative for CK, SMA (Fig. 2h), EMA, and S-100. The percentage of Ki-67-positive cells was 3% in Case #1 before recurrence. After relapse, the percentage of Ki-67-positive cells was higher than 10% in Case #1, 15% in Case #2 (Fig. 2i), and 50% in Case #3 after his third surgery.

The pathological manifestations of 5 previously conducted researches and the present study with recurrent orbital SFT before and after recurrence, including tumor necrosis, mitotic rate, and the percentage of Ki-67-positive cells are shown in Table 2. The mitotic rate decreased in only one case, while it remained stable in 3 cases and increased in 4 cases; besides, the percentage of Ki-67-positive cells slightly decreased after recurrence in only one case, while it stably maintained in 10 patients and increased in 5 patients. Tumor necrosis was found in 1 case after recurrence.

Table 2
Pathological manifestations of recurrent orbital SFT cases [3, 4, 10, 14, 17, 23]

	Starting			After relapse		
	Necrosis focus	Mitosis /10hpf	Ki-67(%)	Necrosis focus	Mitosis /10hpf	Ki-67(%)
Hayashi et al. (2001)	NA	5–6	NA	NA	5	NA
Ness et al.(2005)	NA	10%	5–10	NA	10%	5–10
Wang et al. (2013)	NA	NA	< 5	NA	NA	5–10
Sagiv et al. (2019)	NA	NA	NA	NA	NA	NA
	NA	30	65	NA	93	40
	NA	5	1	NA	5	15.5
	NA	NA	NA	NA	10	20
	NA	5	3	NA	NA	NA
	NA	8	5	NA	10	10
	NA	2	10	NA	1	NA
	NA	NA	NA	NA	6	5.5
Yang et al.(2021)	NA	NA	< 10	NA	NA	< 10
	NA	NA	< 10	NA	NA	< 10
	NA	NA	< 10	NA	NA	< 10
	NA	NA	< 10	NA	NA	< 20
	NA	NA	< 10	NA	NA	< 10
	NA	NA	< 10	NA	NA	< 10
	NA	NA	< 10	NA	NA	< 10
	NA	NA	< 10	NA	NA	< 10
	NA	NA	< 10	NA	NA	< 10
	NA	NA	< 10	NA	NA	< 10
The present report	N	N	3	N	> 4	> 10
	N	N	NA	N	> 4	15

Abbreviations: Y, yes; N, no; NA, not available

Starting			After relapse		
NA	NA	NA	Y	A lot of	50
Abbreviations: Y, yes; N, no; NA, not available					

Discussion

SFT is an anatomically ubiquitous mesenchymal neoplasm with an equal gender distribution that often presents as a large, deep-seated soft tissue or visceral mass. It may occur in various extrapleural sites, such as respiratory tract, nose and paranasal sinuses, parotid gland and salivary gland, thyroid, lung, etc [2, 3]. However, since this disease is rare, few cases (within 100) with orbital SFT have been reported, especially recurrent orbital SFT.

In the present research, clinical, imaging, and pathological features of 3 patients with recurrent orbital SFT were retrospectively compared. We also used the PubMed database to search for recurrent orbital SFT, and 21 related literatures were selected to explore their clinical and pathological features [3, 4, 6–23]. To our knowledge, the current study involved the majority of previous researches on recurrent orbital SFT published in English in the recent two decades.

Clinical Manifestations

SFT was originally thought to occur exclusively in the intrathoracic region, while it has been recently described in extrapleural sites (e.g., orbit). SFT of the orbit is a rare lesion, which can be misdiagnosed as hemangioendothelioma, fibrous histiocytoma, meningioma, or neurofibroma. Its main clinical manifestations are painless protrusion of the eyeball, ptosis, diplopia, and limited eye movement. Painless, non-pulsating, incompressible masses can be touched by physical examination [24]. The patients involved in the current study had a mean age of 54 years old, and their clinical manifestations included exophthalmos, ptosis, abnormal eye position, and blurred vision, which were consistent with those reported previously [24–27].

Recurrence time was also counted through literature analysis and the current cases (see Table 1), although the duration of recurrence after the first surgery for patients who aged ≤ 50 and >50 years old showed no significant difference (P = 0.19). The average time to recurrence in patients who aged under 50 years old was shorter than that in those who aged over 50 years old, suggesting that young patients were more likely subjected to recurrent orbital SFT. Moreover, 21.2% of patients with SFT had multiple recurrences, including Case #3 presented herein and those reported previously. A number of scholars have shown that the failure of radical resection is a high risk-factor for tumor recurrence [19]. The tumors of 3 patients reported in the current study were located in the posterior muscular cone of eyeball, which caused difficulty in their surgical removal. After reviewing the literatures, we found that orbital SFT can originate from any part of the orbit, mainly from the superomedial orbital quadrant (20%), medial orbit (15%), and superotemporal area (13%). Lacrimal sac area and inferotemporal area also account for 9.5%

and 7%, respectively [3, 28]. However, after comparing the clinical characteristics of the previously conducted researches and the cases herein presented (Table 1.), the most common site for recurrent orbital SFT was the retrobulbar area of the orbit (23.8%). This site is the most difficult area of orbital surgery, and the surgical approach, surgical method and surgical field exposure may all affect the prognosis [29]. The fact that the tumor was in almost the same place before and after recurrence suggested that the failure of the first surgery to completely remove the tumor, which left residual tumor, could be responsible for the recurrence. In addition to the local recurrence, orbital SFT is also associated with distant metastasis. In the present research, among the 4 cases, only Case #3 had a history of multiple recurrences. As distant metastases scarcely occur, further study needs to be conducted.

Imaging Manifestations

The MRI findings of orbital SFT showed that lesions had equal density with the gray matter on T1WI, and values of signal intensity on T2WIs were controversial [30–33]. Previous studies reported that orbital SFT is rich in fibrous matrix and T2WI should be dominated by a low signal intensity, however, in several reported cases of malignant orbital SFT, cystic degeneration could be detected with a high signal intensity, suggesting that cystic degeneration could be related to malignant transformation [4, 34]. In addition, the contrast-enhanced MRI showed enhanced SFT, while the enhancement can be uniform or uneven, and it may be associated with the distribution of blood vessels and capillary permeability [35–37]. In the current research, 2 patients underwent MRI of the orbit, and the values of signal intensity on T1WI and T2WI before and after the recurrence of SFT were consistent, indicating that MRI signals did not significantly change before and after recurrence. Changes in cystic lesion were both observed before and after recurrence in Case #2. According to the results of IHC, the patient was diagnosed as benign orbital SFT before recurrence and malignant SFT after recurrence, demonstrating that change in cystic lesion may not be the specific marker for differentiating benign SFT from malignant SFT.

Pathological Manifestations

Orbital SFTs are largely benign tumors with few instances of recurrence. Grossly, they could be round- or long oval-shaped specimens. The masses of three cases in the current study were elliptical, all of which were behind the eyeball and confined by the orbital wall. Yang et al pointed out that although a tumor is mainly benign, it may involve the orbital bone and occasionally penetrate the orbital wall and invade the intracranial structure [23]. During the second surgery on Case #1 and Case #2, surgeons found an adhesion between the masses and the orbital bone. In Case #3, the tumor not only involved the right-sided orbit, but also maxillary sinus, ethmoid sinus, and frontal sinus. However, after recurrence, the tumor may inevitably adhere to the bone wall due to the previous surgery, thus, destruction of the tumor on orbital bony fissure before and after recurrence still needs further observation and verification.

In the review of subjects of previous studies, it was found that SFTs shared a histopathologic morphology, which was recognized for spindle cells, remarkably characteristic ‘staghorn’ blood vessels, and matrix with numbers of collagen bundles [24]. The results of IHC showed that CD34 is a useful marker in the diagnosis of SFT. Previous studies reported that SFT expressed CD34 in 90% of cases,

CD99 in 70% of cases, and Bcl-2 in only 30% of cases [26, 37]. In 2016, the World Health Organization (WHO) classified tumors with a classic SFT phenotype as grade I, tumors with intermediate or HPC phenotype as grade II, and tumors with five or more mitoses ($\times 10$ HPFs) as grade III [38]. A number of scholars demonstrated that the presence of mitosis is a poor prognostic indicator, and, Sagiv et al. emphasized that the number of mitotic figures higher than 4/10 HPFs can be a predictor of malignant SFT [4]. In addition, nuclear atypia, cell proliferation, and necrosis were also considered as manifestations of malignant SFT [35]. It has been reported that the percentage of Ki-67-positive cells tends to increase correspondingly in SFT with an invasive tendency, which is clinically significant for the selection of postoperative treatment plan and assessment of tumor prognosis [37, 39–41]. In three cases reported herein, tumor cells were spindle-shaped and densely distributed in collagen fibers. The nuclei were oval, round or spindle-shaped, the cytoplasm was pale and eosinophilic, and branched or antlers-shaped blood vessels could be observed. IHC showed CD34 positive, while S-100 and CK negative, which were consistent with the diagnosis of orbital SFT. In cases #1 and #2, nuclear abnormalities were found for the first time after recurrence, and those cases were diagnosed as malignant SFT with mitotic activity of $\geq 4/10$ HPFs. To date, few reports have concentrated on the recurrent orbital SFT, and there is currently no large sample-sized controlled trial related to the pathological manifestations of recurrent orbital SFT before and after recurrence. In the present study, we compared the pathological manifestations of 24 orbital SFT patients before and after recurrence, including tumor necrosis, mitotic rate, and the percentage of Ki-67-positive cells. As shown in Table 2, the rate of SFT mitosis was elevated in the relapsed eyes, and the proportion of Ki-67 positive cells remained stable in some cases, while it increased in a large proportion after recurrence, indicating that recurrent orbital SFTs tend to increase in malignancy in terms of both morphological features and IHC results. In addition, the pathology testing of Case #3 after repeated recurrences revealed new occurred necrosis, and the percentage of Ki-67-positive cells was about 50%, suggesting that the degree of malignancy may be higher after several recurrences, while the above mentioned results should be verified by further case-control studies with a larger sample size.

In conclusion, the time to recurrence of orbital SFT in the reported patients ranged from 3 months to 20 years, and the average time to recurrence in patients who aged under 50 years old was shorter than that in those who aged over 50 years old. In the current report, 2 patients underwent MRI of the orbit, and the values of signal intensity on T1WI and T2WI before and after recurrence of SFT were consistent, indicating that MRI signals did not significantly change before and after recurrence. Morphological features and IHC results indicated that recurrent orbital SFTs tend to be malignant. The degree of malignancy may increase with the frequency of recurrence. Therefore, orbital SFT requires radical resection to avoid tumor recurrence and change of malignant tendency.

Declarations

Findings:

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Data availability

All data and materials support our published claims and comply with field standards.

Conflicts of interest

There are no conflicts of interest.

Authors' contributions

CJL, WXW, LY and CHG gathered the clinical information and drafted the manuscript. WXW and ZMM collected the clinical data and revised the draft.

Statement:

That the manuscript has been read and approved by all the authors. Each author believes that the manuscript represents honest work.

Ethical Approval:

The current study was approved by the Ethics Committee of the First Affiliated Hospital of Zhejiang University School of Medicine, and was performed in accordance with the Declaration of Helsinki. Besides, all participants signed the written informed consent form prior to commencing the study.

Consent for publication

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Figures

Figure 1

Magnetic resonance imaging (MRI) of the orbit before and after recurrence in case 1 showed retrobulbar tumor in the left orbit. (a) T1-weighted image. (b) T2-weighted image. (c), contrast-enhanced T1-weighted image. (d), T1-weighted image. (e), T2-weighted image. (f), contrast-enhanced T1-weighted image.

Figure 2

Results of histopathology and immunohistochemistry of the tumors before and after recurrence in case #2. (a) tumor with spindle-shaped cells with oval-to-spindle nuclei. (H&E staining; original magnification, x100 times). (b) typical 'spinal cord'-like antlers (H&E staining; original magnification, x400 times). (c) irregularly arranged spindle-shaped cells are more heterogeneous than those reported in 2010 (H&E staining; original magnification, x100 times). (d) > 4 mitotic images (shown in black circle) were found under high power field. (e) tumor cells were positive for CD34 (original magnification, x200 times). (f) tumor cells were positive for CD99 positive (original magnification, x200 times). (g) tumor cells were positive for Bcl-2 (x200 times). (h) tumor cells were negative for SMA (original magnification, x200 times). (i) tumor cells were positive for Ki-67 (about 15%; original magnification, x200 times).