

Case Report

Orbital Solitary Fibrous Tumor : A Case Report and Diagnostic Clues

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Orbital solitary fibrous tumor (SFT) is a rare tumor originating from the mesenchyme. We describe the clinical presentations, radiological and operative findings, and pathological features of a patient with orbital SFT. The patient was a 46-year-old female who presented with progressive proptosis advanced for 20 months. On ophthalmological examination, no visual impairment was detected, but left eye was found to be obviously protruded on exophthalmometry. Orbital magnetic resonance imaging showed a $2.5 \times 2 \times 2$ cm, intensely enhanced mass in the left orbit, which compressed the eyeball forward and the optic nerve downward. The patient underwent frontal craniotomy with superior orbitotomy and gross total resection was performed for the tumor. The histopathological diagnosis including immunohistochemistry was a SFT. After the surgery, proptosis was markedly relieved without visual impairment. Although orbital SFT is extremely rare, it should be considered in the differential diagnosis of orbital tumors. Clinical presentations such as painless proptosis and CD34 immunoreactivity play a significant role in differentiating orbital SFT from other spindle-cell neoplasms of the orbit.

KEY WORDS : Orbital · Solitary fibrous tumor · Mesenchymal origin tumor · CD34.

INTRODUCTION

Solitary fibrous tumor (SFT) is a rare spindle-cell tumor of mesenchymal origin that arises most often in the pleura, but may also occur at other organs, including lung, mediastinum, pericardium, peritoneum, retroperitoneal space, pelvis, adrenal gland, kidney, liver, periosteum, salivary gland, thyroid gland, lacrimal gland, breast, nasopharynx, orbit, urogenital system, skin, meninges and spinal cord^{3,10,11,13,14,19,21,24,27,33}. Since the first report of orbital SFT by Westra et al.³⁴ in 1994, fewer than 60 cases of orbital SFTs have been reported to date. Orbital SFT has possibly been underdiagnosed in the past, because of its rarity in extra-pleural organs and histological similarity to other spindle cell tumors. Currently, with the development of immunohistochemical method, the number of reported cases have been increasing⁹. One of the most common symptoms is unilateral painless proptosis, which can be accompanied by visual disturbance, palpable orbital mass, ocular mobility

disturbance, hyperglobus or blepharoptosis from the secondary mass effect in the orbit²⁴. As a general rule, orbital SFT shows an indolent and nonaggressive clinical course. It is known that a complete cure can be achieved with gross total resection^{2,15,17}.

CASE REPORT

A 46-year-old female presented with a 20 months history of progressive proptosis. On ophthalmologic examination, normal visual acuity, normal visual field and full range of extraocular movement were present. On funduscopy, there were no findings indicating optic nerve atrophy or papilledema. On exophthalmometry, the exophthalmos of the left eye was measured as 18 mm. Orbital computed tomography (CT) scan showed a $2.5 \times 2 \times 2$ cm sized well-demarcated left extraconal superomedial orbital mass (Fig. 1). No evidence of calcification or bony destruction was observed. Orbital magnetic resonance images (MRI) showed a mass located in the upper and medial quadrant of the left orbit with low signal intensity on T1-weighted images and mixed signal intensity on T2-weighted images, which was enhanced strongly with gadolinium (Fig. 2). The mass was compressing the eyeball forward and the optic nerve down-

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ward. The patient underwent left frontal craniotomy with superior orbitotomy under cerebrospinal fluid lumbar drainage. Opening periorbita exposed fat tissue, which was carefully dissected laterally and levator palpebrae muscle underneath was retracted away from the midline. Then, a well demarcated solid mass was identified occupying the left orbit superoposteriorly. The mass was located within a clear dissecting plane isolated from the underlying extraocular muscles with no evidence of local invasion. It was too hard to be grinded with an ultrasonic aspirator. Instead, we removed it with piecemeal fashion using a knife on a step-by-step basis until gross total resection was obtained. After the removal of the mass gross totally, we checked extraocular muscles, optic nerve and ophthalmic artery undamaged, and the surgical procedure was finished after suturing periorbita. Pathological examination disclosed a spindle-cell tumor in a dense fibrous tissue. Immunohistochemical

stains of the tumor demonstrated a positive staining for vimentin and CD34 but negative for smooth muscle actin (SMA), cytokeratin and S-100 protein (Fig. 3). With these data, it was finally diagnosed as a SFT. After the surgery, proptosis was completely relieved with visual acuity and the function of all extraocular muscles remained intact. Three months after surgery, there was no enhanced mass on orbital MRI (Fig. 2).

DISCUSSION

Patient suffering orbital SFT presents usually with unilateral proptosis and a strongly enhancing, well-circumscribed mass on MRI. Other orbital neoplasms, such as fibrous histiocytoma, nerve sheath meningioma, schwannoma and hemangiopericytoma, share a similar clinical presentation. Furthermore, spindle-cell tumors in orbit have microscopic resemblance to SFT which exhibit focal hemangiopericytoma-like staghorn, fibrous histiocytoma-like storiform pattern, synovial sarcomatous and neural-like palisading regional architecture¹⁾. For these reasons, it is possible that former orbital SFT have been misdiagnosed. Recently, however, advanced immunohistochemical techniques have facilitated a clear distinction.

On immunohistochemical findings, SFT has the strong and diffuse positivity to CD34, vimentin and BCL2, and negativity to desmin, cytokeratin, factor VIII related antigen, S-100 protein, muscle specific actin and SMA^{8,10,15,29)}. Smooth muscle tumors such as leiomyoma and leiomyosarcoma, on the other hand, show the positivity to actin and desmin, and negativity to CD34 and BCL2.

Neural tumors such as schwannoma and neurofibroma show focal positivity to BCL2 and CD34, and strong positivity to S-100 protein^{22,29)}.

Strong and consistent CD34 immunoreactivity in SFT can be the important clue for histopathological distinction from other spindle shaped cell tumors, such as meningioma, fibrous histiocytoma, schwannoma and hemangiopericytoma^{2,8,10,12,15,18)}. Of these tumors, a differential diagnosis to hemangiopericytoma with SFT is mandatory because hemangiopericytoma has malignant clinical course with recurrence rate of 83%, rate of distant metastasis of 27%, mortality rate of 22%, as opposed to general benign

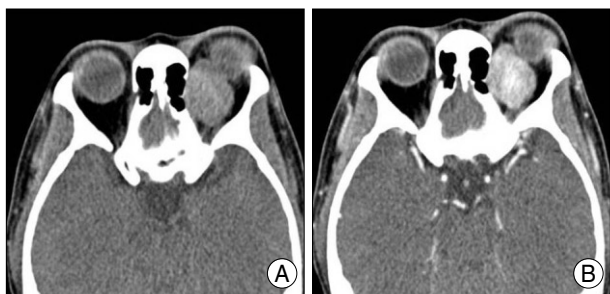


Fig. 1. Orbital computed tomography (CT) scan showing 25 × 20 × 20 mm sized well-demarcated extraconal superomedial orbital mass. A : Precontrast CT scan. B : Postcontrast CT scan showing intensely enhancing mass.

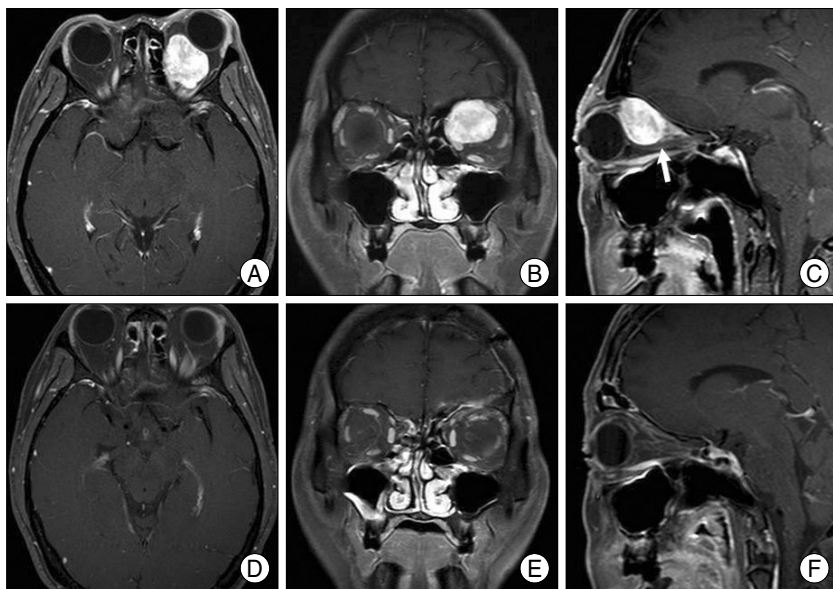


Fig. 2. A, B and C : Preoperative orbital magnetic resonance image (MRI) show that optic nerve (white arrow at C) are compressed and downward displaced by strong enhancing mass located in the upper and medial quadrant of the left orbit. D, E and F : On MRI performed 3 months later, removal state of left retrobulbar mass and relieved state of proptosis of the left eyeball were seen.

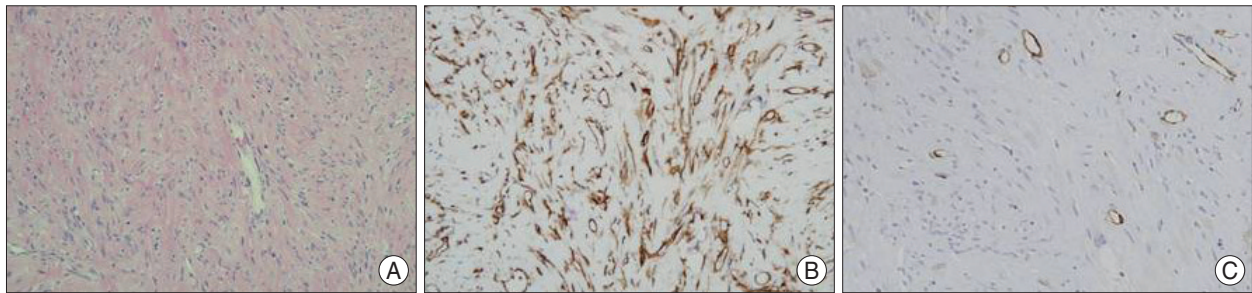


Fig. 3. A : Spindle cell tumor with a characteristic cracking artifact between cells and collagen (H & E, $\times 200$). B : Strong and diffuse positivity to CD34 ($\times 200$). C : Negativity to smooth muscle actin ($\times 200$).

nature of SFT^{4,31}). The hemangiopericytoma is typically composed of small, oval to slightly spindle shaped cells, and it also has a lot of branching, thin-walled vessels of varying caliber and “staghorn” vascular pattern. Immunohistochemical analysis of hemangiopericytoma reveals strong positivity to vimentin, factor XIIIa and Leu-7, and weak positivity to desmin and cytokeratin which are similar characteristics sharing with SFT. However, SFT shows strong positivity to CD34, while hemangiopericytoma shows weak positivity^{25,30}.

Orbital SFT often has some analogy with optic nerve sheath meningioma on the radiologic findings, for which a differential diagnosis is important clinically. In cases of SFT, there are no findings such as psammoma body, whorl formation of tumor cells or amorphous calcification, which are commonly found in meningiomas. Besides, in meningioma, focal staining to BCL2 or CD34, and intense staining to cytokeratin were observed^{25,28-30}. On symptomatology, optic nerve sheath meningioma, as it gradually increases in size, usually causes a disturbance in the blood flow to the adjacent area of the optic nerve²³. For this reason, visual symptoms and signs including impairment of visual field and acuity, disturbance in color vision and development of diplopia tend to occur earlier than proptosis. The majority of patients with orbital SFT, however, present with unilateral painless proptosis as the initial clinical feature because of the mass effect compressing both eyeball and optic nerve.

The probability of local recurrence related to incomplete resection has led surgical en bloc removal to be recommended as the treatment of choice in SFT^{2,5,13,14,21,24,27}. No previous studies have demonstrated the effect of chemotherapy or radiotherapy^{6,7,26}. The possibility for malignant transformation is closely associated with histologic features². The findings, such as nuclear atypia, increased cellularity, necrosis and greater than 4 mitoses/10 high-power fields, are suggestions of the malignant potential of SFT³². The prognosis in SFT, however, most likely depends upon complete resection rather than histologic findings because the clinical behavior of SFT is not in agreement with the

histologic prognostic factors in all cases^{16,20,24}.

The most frequent clinical presentation of orbital SFT is unilateral painless proptosis, which led the ophthalmologists to report most of the cases so far. In majority, orbital SFT originated from lacrimal glands according to the previous reports, in which orbitotomy alone could give successful surgical results^{17,21,24,26,35}. But, SFT arising from mesenchymal orbital connective tissue could have its location within the retro-orbital space where frontal craniotomy with superior orbitotomy is essential like what is performed to remove optic nerve sheath meningioma.

CONCLUSION

We experienced a rare case of orbital SFT. Although extremely rare, orbital SFT should be included in the differential diagnosis of a well-circumscribed and intensely enhancing orbital mass on MRI. Clinical presentation such as painless proptosis and CD34 immunoreactivity play a significant role in differentiating orbital SFT from other spindle-cell neoplasms of the orbit.

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