The Diagnosis and Treatment of Orbital Solitary Fibrous Tumor

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Abstract

Background

Solitary fibrous tumor (SFT) is an uncommon spindle-cell tumor of mesenchymal origin, it commonly occurs within pleura, the occurrence of SFT in the orbit is very rare. The diagnosis of orbital SFT is challenging in clinical, and requires an integrated approach that includes specific clinical, histological, immunohistochemical, and even molecular findings.

Methods

In our retrospective study, we collected 13 patients with orbital SFT. All patients were diagnosed with orbital SFT by postoperative histopathological and immunohistochemical examination. Patient charts and medical records were reviewed for demographic information, relevant medical and family history, clinical presentation, radiological examination, histopathological and immunohistochemical examination, the treatment and prognosis.

Results

This study included 7 males and 6 females. The age ranged from 11 to 78 years. 7 patients presented with predominant features of exophthalmos. 7 patients showed eyeball dislocation. 6 patients with motility disturbances. 9 patients had the palpable masses. 6 patients had the secondary lesions. On CT scan, there were 6 patients were located in the superomedial quadrant of the orbit, 3 patients were located in the inferomedial quadrant of the orbit. The morphology of lesions was solitary ovoid mass in 10 patients, and irregular mass in 3 patients. The CT value of the tumor was 22.8-64.4Hu, with the median of 45.9Hu. On MRI, 3 patients showed hypointense mixed signals on T1WI, 10 patients showed isointense mixed signal on T1WI. 3 patients showed hypointense mixed signals, 4 patients showed isointense mixed signals, 6 patients showed hyperintense signals on T2WI. There were 12 patients showed obviously enhancement, and we found patchy lesions which were no enhancement in the tumor. All patients were treated by surgery. Immunohistochemical analysis showed that the tumor cells exhibited diffuse immunoreactivity for CD34 and CD99 in all the patients. The lesions were positive for Bcl-2 in 11 patients. The lesions were positivity for Ki-67 <5% in one patient, 5%-10% in 10 patients, >10% in 2 patients.

Conclusions

The lesions are often outside the muscular cone, most commonly located at superomedial quadrant and inferomeddial quadrant of the orbit. The density of the lesions is uneven in most cases, the mean CT values of the tumors on CT scans are variable. The signal of lesions on orbital MRI is uncertainty, it is inheterogeneous enhancement, and the lamellar unenhanced regions can also be found in the lesion. The complete gross resection, and even more aggressive wide excision is preferred. Although the Ki-67 labelling index is very low, malignant forms with an increased propensity for local recurrence have been found. A longer follow-up is needed to determine the recurrence rate of the disease, and to identify the causes of recurrence.

Background

Solitary fibrous tumor (SFT) is an uncommon spindle-cell tumor of mesenchymal origin, which was initially described in the pleura by Klemperer and Rabin in 1931[1]. It is originally believed to be of submesothelial origin, however, the histogenesis of SFT is still controversial. The etiology of the neoplasm remains largely unknown, although most of them are benign, many studies show the tumors will behave in an aggressive manner. SFT commonly occurs within pleura, the occurrence of SFT in the orbit is very rare[2]. Owing to its rarity, the diagnosis of orbital SFT is challenging in clinical, and requires an integrated approach that includes specific clinical, histological, immunohistochemical, and even molecular findings[3]. We review the diagnosis and treatment of 13 patients with orbital SFT in detail, our aim is to find the clinical characteristic features and relatively complete treatment experience, and improve the accuracy of diagnostic and treatment.

Patients And Methods

In our retrospective study, we collected 13 patients with orbital SFT from January 2012 to March 2019 in Hebei Eye Hospital. All patients were diagnosed with orbital SFT by postoperative histopathological and immunohistochemical examination. Patient charts and medical records were reviewed for demographic information, relevant medical and family history, clinical presentation, radiological examination, histopathological and immunohistochemical examination, the treatment and prognosis. The study follewd the tenets of the Declaration of Helsinki, and all procedures were approved by the ethics committee of the Hebei Eye Hospital. All patients provided written informed consent before the procedures. All patients included in the study are fully informed about the study and are voluntary for providing data for analysis. All data is recorded and stored in compliance with ethical and data protection guidelines.

Results

Demographic information, relevant medical and family history

13 patients with histopathologic diagnosis of orbital SFT were identified in our hospital from January 2012 to March 2019. This study included 7 males and 6 females. The age ranged from 11 to 78 years, the median age at diagnosis was 43 years. The systemic complications included 3 patients of hypertension, 1 patient of iron deficiency anemia, 1 patient of chronic bronchitis, and 1 patient of sinusitis. None of the patients had the family history.

Clinical presentation

There were 7 patients in right orbit and 6 patients in left orbit. 7 patients (7/13) presented with predominant features of exophthalmos, 2 patients (2/13) with eyelid swelling and 2 patients (2/13) with painful mass. 1 patient (1/13) with swelling of the lacrimal sac and epiphora, and 1 patient (1/13) with visual disturbances. In this study, 8 patients had visual acuity above 20/60, 1 patient was 20/100, and 3

patients had visual acuity lower than 20/200. Among them, 2 patients had no light perception, and all the 2 patients were recurrent orbital SFT. Compared with the contralateral eye, the exophthalmos of the affected eye by the Hertel exophthalmometry was 1-6mm, with the median of 4mm. 7 patients showed eyeball dislocation, 3 patients of them dislocated in inferolateral. The eyeball dislocation in inferomedial, superomedial, superior, inferior, each accounted for 1 patient. 6 patients with motility disturbances, 3 patients of them were significantly restricted in all directions. 2 patients of them were the recurrent cases. 9 patients had the palpable masses, 1 patient of them with obvious tenderness. 6 patients had the secondary lesions. Optic nerve atrophy was observed in 2 recurrent patients, fundus hemorrhage was observed in 1 patient, lacrimal duct obstruction was observed in 2 patients, corneal perforation secondary to exposure keratitis was observed in 1 patient due to severe exophthalmos.

Orbital CT examination

The location of the lesions was identified by orbital CT, there were 6 patients were located in the superomedial quadrant of the orbit, 3 patients of them involved the muscular cone. 3 patients were located in the inferomedial quadrant of the orbit, 2 patients of them involved the lacrimal sac area. 2 patients were located in the inferolateral quadrant, and 1 patient was observed in superolateral quadrant and 1 patient in the medial quadrant of the orbit.

The morphology of lesions was solitary ovoid mass in 10 patients, and irregular mass in 3 patients.

9 patients had well-defined boundary, and 4 patients had unclearly boundary. The lesions adhered closely to the extraocular muscles in 7 patients, of which 3 patients adhered to the optic nerve. The lesions compress the lacrimal sac in 2 patients. The lesions spread to the brain, nasal cavity and eyelids in 1 recurrent patient. There were 9 patients showed compressive bone change. The CT value of the tumor was 22.8-64.4Hu, with the median of 45.9Hu. The standard deviation of CT density in 4 patients was lower than 10% of their own's CT value, and 9 patients were higher than 10% of their own's CT value. The size of the largest lesion was 2.7cm*2.9*4.1cm, and the smallest was 1.0cm*1.3cm*1.4cm (Figure 1).

Orbital MRI examination

T1WI performance: 3 patients showed hypointense mixed signals on T1WI, 10 patients showed isointense mixed signal. The signal intensity on most of lesions was homogeneous, we aslo found patchy hyperintense signals lower than the most lesions. T2WI performance: 3 patients showed hypointense mixed signals, 4 patients showed isointense mixed signals, 6 patients showed hyperintense signals. Only 2 patients showed relatively homogeneous signals, and the other 11 patients showed uneven signals. There were 12 patients showed obviously enhancement, and we found patchy lesions which were no enhancement in the tumor. There was 1 patient with obviously enhanced in the surrounding area, but the enhancement was not obviously found inside the tumor (Figure 2).

The treatment

All patients were treated by surgery. Depending on the location, size, and relationship to surrounding tissue, we determined the surgical approach. 2 patients were treated surgically by lateral orbitotomy. 2 patients were treated surgically by lateral orbitotomy combined with the medial conjunctival approach. 9 patients were treated surgically by anterior orbitotomy. Among 9 patients, there were 3 patients via transcutaneous superomedial routes, 1 patients with transcutaneous superolateral routes, 3 patients with frontoethmoidal medial orbitotomy, 1 patients with transcaruncular medial orbitotomy, 1 patients with transconjunctival orbitotomy. The lesions were completely removed in 11 patients. One recurrent patient underwent a majority resection. Another recurrent patient underwent mass resection combined with orbital exenteration. All patients were not treated with chemotherapy, radiotherapy, and other measures after surgery.

Histopathological and immunohistochemical examination

Among the excised tumors, 8 patients had capsules or pseudocapsules, and 5 patients had no capsules. The sections of the tumor in 6 patients was gray-white, and 7 patients were gray-red. The tumors in 8 patients were brittle, and 5 patients were tough. On histopathological study, the tumor was composed of haphazardly arranged spindle cells with bland nuclei and inconspicuous nucleoli. Immunohistochemical analysis showed that the tumor cells exhibited diffuse immunoreactivity for CD34 and CD99 in all the patients. The lesions were positive for Bcl-2 in 11 patients. The lesions were positivity for Ki-67 <5% in one patient, 5%-10% in 10 patients, >10% in 2 patients. In addition, 7 patients underwent immunohistochemical staining for Vimentin, all the patients showed positive. 3 patients underwent immunohistochemical staining for STAT-6, all the patients showed positive (Figure 3).

The prognosis

The follow-up period was form 1 years and 6 months to 8 years, with the median of 3 years. 2 patients had tumor recurrence, 1 of them took 3 years after surgery, and 1 was 4 years.

Discussion

Clinical features of orbital SFT

With the further understanding of SFT, although it is commonly occurs within pleura, there has been an increase in its incidence in nonpleural sites in recently. SFT is reported in diverse bodily locations, and recognized to have a wider range of clinical and radiological features[4]. Orbital SFT is a kind of extrapleural SFT, and is a rare kind of orbital tumor. Due to its relatively rare, the clinical characteristics have not been fully grasped at present. Although SFT is usually considered as a clinically indolent neoplasm, the orbital SFT is usually more aggressive than pleural form, and the prognosis is substantially unpredictable[3]. Therefore, it has a great clinical significance to study the clinical characteristics and development rules of this tumor.

Orbital SFT has no obvious age preferenction, our study has been documented across a wide age range from 19 to 72 years, and children's patient has also been reported[5]. But from previous reports, as well as our findings, adult cases seem to account for the majority. It has no sex and side of affected eye predilection according to previous reports.

The common manifestations include exophthalmos and eyeball dislocation, andsome patients may be associated with eyelid swelling, visual disturbances, a palpable painless mass, epiphora, and ptosis, periorbital spontaneous pain or tenderness[6]. The majority of patients suffer minor visual impairment. But the patients of recurrent orbital SFT with compressive optic neuropathies or exposure keratitis, often result in severe impairment of visual function, or even loss of vision. In some patients, the lesions are relatively superficial in the orbit, the painless soft tissue masses can be touched. If the mass presses on the dacryocyst, symptoms of epiphora is common. Although the clinical manifestations were diverse, they are not significantly different from other orbital tumors.

Imaging features of orbital SFT

Orbital SFT usually presents as a soft-tissue mass on CT. The tumors are often outside the muscular cone, partial cases are involved in, or within the in the orbital muscle cone. Most commonly located at superomedial quadrant of the orbit, the second is inferomedial quadrant of the orbit. The masses are mostly round or oval in shape. The density of the lesions is uneven in most cases. The mean CT values range from 22.8 Hu to 66.4 Hu in our study, thus the mean CT values of the tumors on CT scans are variable. This is similar to schwannoma, but different from cavernous hemangiomas. The boundary of the tumor is most clear, but it is un clear when it is recurrent cases or the lesion is very large. Remodeling of the adjacent bones can be seen with long-standing orbital SFT[6], due to the compression of the lesions. The change of adjacent bones is found in 9 patients of our group. The disease can spread to the nasal cavity or the brain in the recurrent or long-standing cases.

Although not pathognomonic, homogeneous or heterogeneous-attenuated enhancement is reported to be the most prominent feature of SFT revealed with CT and MR imaging, which is attributed to high vascularity because of the prominent vascular channels within the tumor. And it can see a markedly enhancing mass showing the similar characteristics to those of the internal carotid artery on postcontrast CT or MR images[7]. We found that the MRI signals of orbital SFT are relatively complex in practice. It is mostly medium or low signal on T1WI, but can show arbitrary signal on T2WI, which may be related to the composition of the tumor. Consistent with previous reports, most tumors can be significantly enhanced. But in the majority of cases, it is inheterogeneous enhancement, and the lamellar unenhanced regions can also be found in this study. In a recurrent cases, we find that most lesions are not enhanced, except for cystic enhancement around the lesion.

Due to the uncertainty of lesion signal on MRI, it is necessary to distinguish from orbital hemorrhage and schwannoma. The lesions in hemorrhage is no enhanced on MRI. The composition of orbital schwannoma is complex. Secondary degenerative changes of the tumor, such as cyst formation,

hyalinization, hemorrhage, are relatively common. These lead to extremely complex MRI findings of schwannomas. But the boundary of the schwannoma is very clear, and we can find that the tumor is associated with the corresponding nerve in some lesions. Other orbital tumors for differential consideration include histiocytomas, giant cell angiofibromas, and hemangiopericytomas[8].

Histopathological and immunohistochemical features of orbital SFT

Some tumor specimen has very thin capsules or pseudocapsules, and some tumor specimen has no capsules. The typical SFT phenotype is that the tumor cells are spindle-shaped, and the alternately arranged sparse and dense cells are separated by eosinophilic collagen fiber bands[9]. Normally, immunohistochemical markers are fundamental for the diagnosis, and the combination of immunohistochemical markers CD34 and Bcl-2 increase the diagnostic accuracy[10,11]. The expression of CD34 is usually diffuse and intense, but according to the report, the expression of Bcl-2 is very high in STF and negative in most malignant mesotheliomas. This is similar to our study. Expression of the Ki67 protein (pKi67) is associated with the proliferative activity of intrinsic cell populations in malignant tumors, allowing it to be used as a marker of tumor aggressiveness[12]. Although the Ki-67 labelling index is very low, most cases are <10% in our study, malignant forms with an increased propensity for local recurrence have been reported[11]. Vimentin is a key component of the cytoskeleton, and is particularly important during development and in cancer during epithelial–mesenchymal transition and metastasis[13]. Due to the high expression rate of Vimentin in orbital SFT, we must be alert for recurrence or metastasis of this tumor. In this study, 3 patients were tested for STAT6, and all were strongly positive. Studies have found that the fusion of NAB2 and STAT6 genes can make the STAT6 protein in the cytoplasm enter the nucleus so that the nucleus strongly expresses the STAT6 protein. The immunohistochemical staining method using the STAT6 protein can quickly detect the NAB2-STAT6 gene fusion status in tumor cells, and it has high sensitivity and specificity for diagnosing SFT. STAT6 has a great auxiliary effect on accurately distinguishing SFT from tumors that are similar in morphology but negative for CD34[14]. Therefore, the combined application of Vimentin, STAT6, CD34 and other markers will help the correct diagnosis of SFT.

Treatment and prognosis of orbital SFT

There are few studies examining the various treatment modalities due to its rare. The mainstay of treatment for orbital SFT is en bloc surgical resection with negative margins. If the initial excision is incomplete, the recurrent tumor tends to spread into surrounding tissues and bone, rendering a second excision much more difficult[8]. In our opinion, these tumors may recur and require even more aggressive wide excision[8]. Especially in the cases with unclear boundary of lesions, the capsule is incomplete or no capsule, noncontact excision, and thorough rinsing of the operationg area after extensive excision, may reduce the recurrence rate of the lesion. Currently, there is little evidence that adjuvant chemotherapy and radiation therapy following complete surgical resection is beneficial and thus they are not routinely performed[15,16].

Through our observations, combined with previous reports, the fairly high local recurrence rate underscores their aggressive potential and highlights the importance of prospective recognition[17]. In our case, 2 patients were found to have relapsed, with the lesions growing more rapidly and more invasive after recurrence. In the cohort of recurrent orbital hemangiopericytoma/SFT, median time to recurrence was 4 years underscoring the importance of careful continued follow-up[18]. This group were followed up from 7 months to 8 years, and the median was 2 years 10 months. Thus, a longer follow-up is needed to determine the recurrence rate of the disease, and to identify the causes of recurrence.

Conclusion

The orbital SFT is rare, the clinical manifestations were diverse and not specificity. The lesions are often outside the muscular cone, most commonly located at superomedial quadrant and inferomeddial quadrant of the orbit. The density of the lesions is uneven in most cases, the mean CT values of the tumors on CT scans are variable. The signal of lesions on orbital MRI is uncertainty, it is inheterogeneous enhancement, and the lamellar unenhanced regions can also be found in the lesion. The complete gross resection, and even more aggressive wide excision is preferred. Although the Ki-67 labelling index is very low, malignant forms with an increased propensity for local recurrence have been found. Thus, a longer follow-up is needed to determine the recurrence rate of the disease, and to identify the causes of recurrence.

Abbreviations

SFT: Solitary fibrous tumor

MRI: Magnetic resonance imaging

CT: Computed tomography

T1WI: T1-weighted images

T2WI: T2-weighted images

HE: hematoxylin and eosin

CD34: cluster of differentiation 34

CD99: cluster of differentiation 99

Bcl-2: B-cell lymphoma 2

Declaration

Availability of data and materials

The data shown and/or analyzed during the present investigation are available from the corresponding author upon reasonable request.

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Authors' contributions

MR and YW designed the study, collected and analyzed the data, and wrote the manuscript. RL, YW, JL, JW, LL and YG assessed the patient and checked and revised the manuscript. MR designed the study, performed all the treatments and agreed to be accountable for all aspects of the work. All authors read and approved the final manuscript.

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Ethics declarations

Ethics approval and consent to participate

The study was approved by the Hebei Eye Hospital Review Boardand was conducted in accordance with the tenets of the Declaration of Helsinki. Written consent for publication of the images and identifying clinical details was obtained from the patient's guardians.

Consent for publication

Not applicable.

Competing interests

The authors declare that they have no competing interests.

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Figures

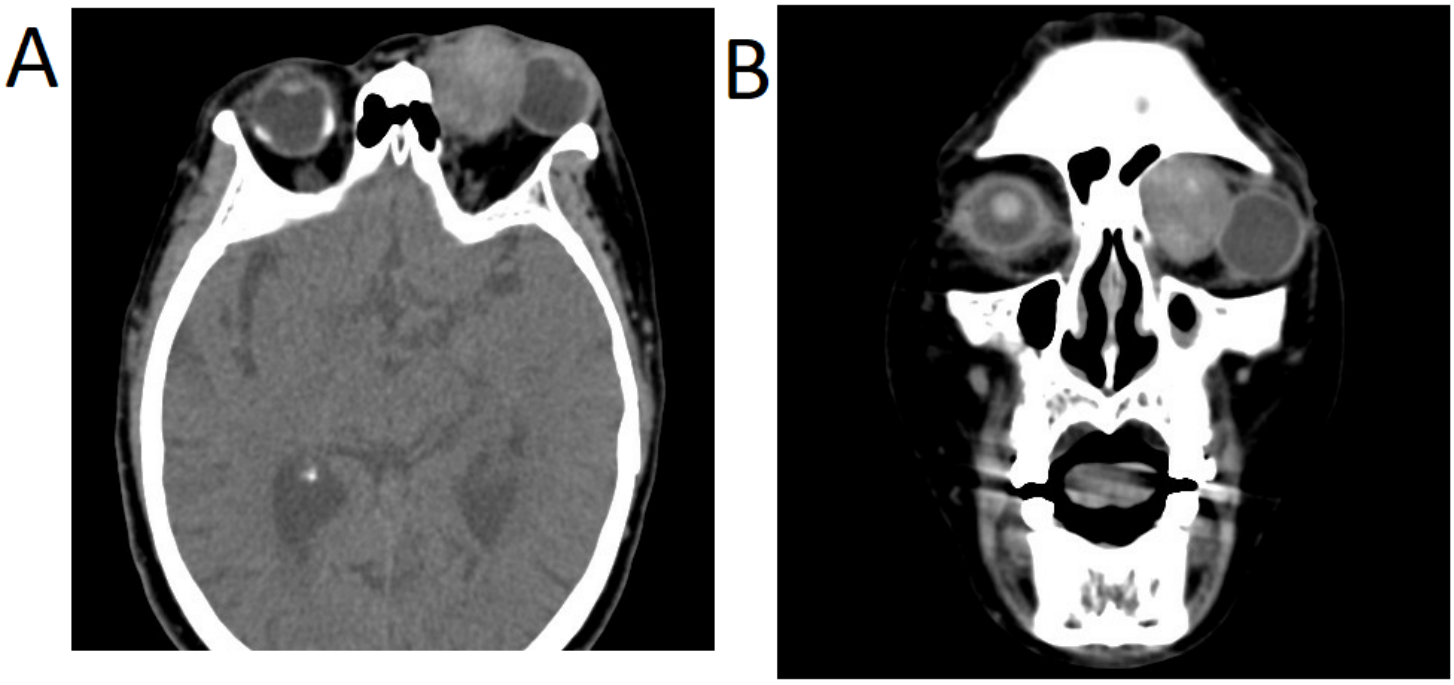

Figure 1

The CT images showed a solitary ovoid mass in the superomedial quadrant of the left orbit. Notes: (A)Axial CT scans. (b) Coronal CT.

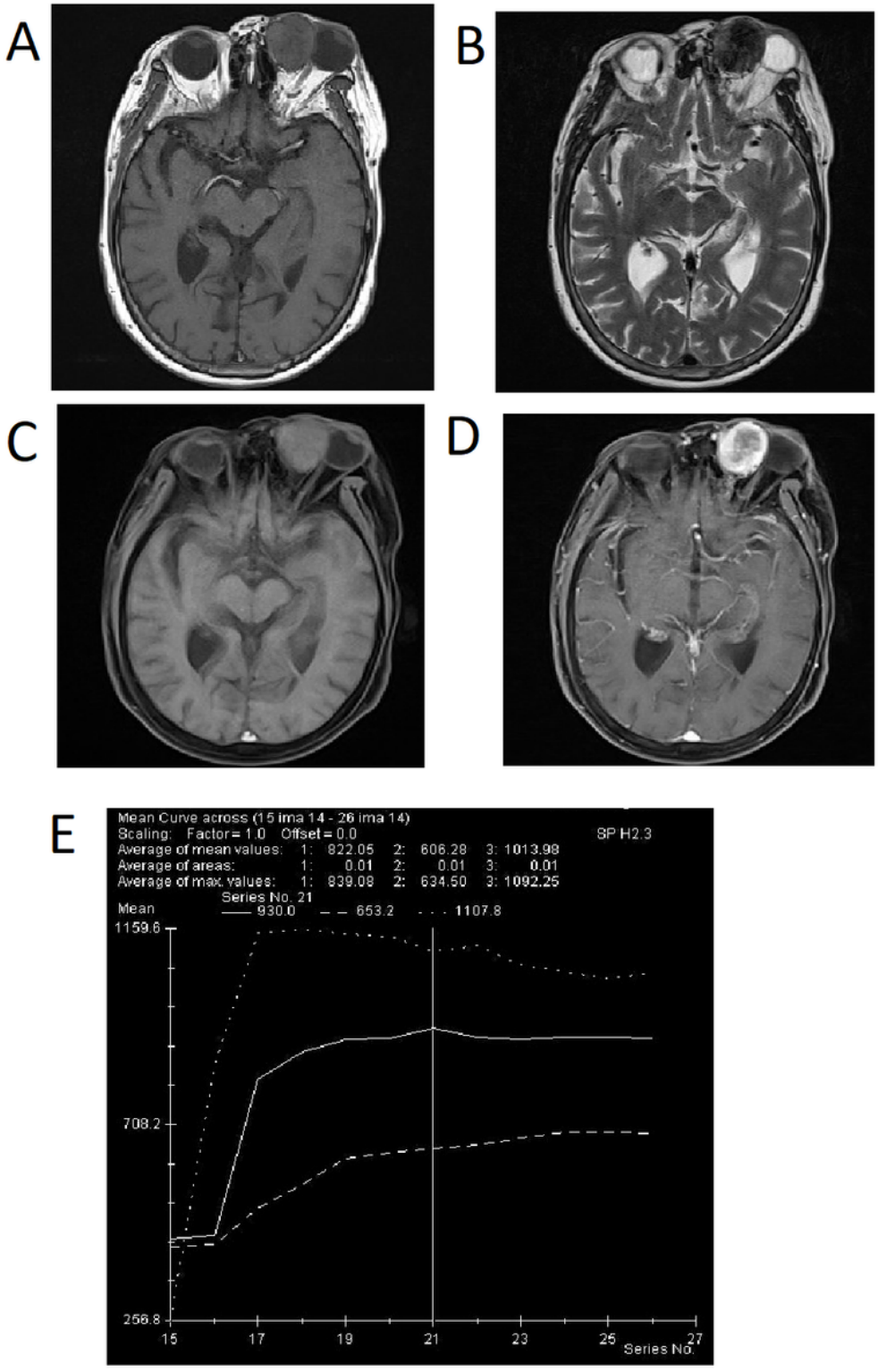

Figure 2

MR scan shows a well-circumscribed circular mass in the superomedial quadrant of the left orbit. Notes: (A)The isointense mixed signal lesion on T1WI. (B) The hypointense mixed signal lesion on T2WI. (C) Post-contrast T1 image. (D) It showed obviously enhancement, and patchy lesions which were no enhancement in the tumor. (E) Magnetic resonance dynamic enhanced curve of the tumor.

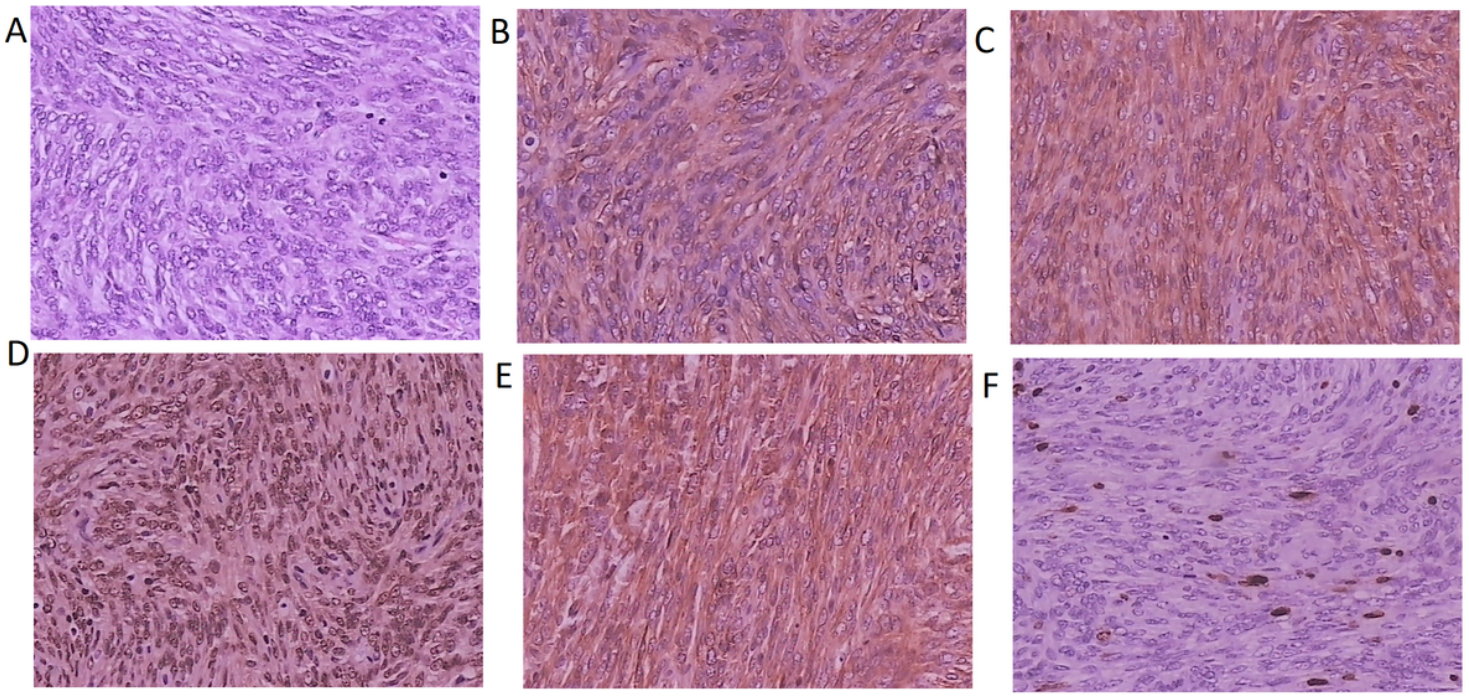

Figure 3

Histopathological and immunohistochemical staining of tumors. Notes: (A) Tumor cells with oval nuclei, conspicuous nucleoli, and mitotic figure are noted. (HE staining; original magnification: ×200). (B) Tumor cells are positive for CD34. (original magnification: ×200). (C) Tumor cells are positive for CD99. (original magnification: ×200). (D) Tumor cells are positive for STAT-6. (original magnification: ×200). (E) The tumor cells are positive for Bcl-2. (original magnification: ×200). (F) The tumor cells are positive for Ki-67 (Close to 10%) (original magnification: ×200).