**Name of Journal:** *World Journal of Clinical Cases*

**Manuscript NO:** 70345

**Manuscript Type:** CASE REPORT

**Primary orbital monophasic synovial sarcoma with calcification: A case report**

Ren MY *et al*. Orbital monophasic synovial sarcoma

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**Author contributions:** Ren MY conceptualized this study, made the literature review and wrote the first draft of this paper; Li J, Li RM, Wu YX, Han RJ, and Zhang C made the literature review; all authors revised the paper and approved the final version for submission.

**Supported by** the Science and Technology Planning Project of Xingtai, No.2019ZC246.

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**Received:** July 31, 2021

**Revised:** October 28, 2021

**Accepted:** December 28, 2021

**Published online:** February 16, 2022

**Abstract**

***BACKGROUND***

Synovial sarcoma is a malignant mesenchymal neoplasm with variable epithelial differentiation. Most synovial sarcoma cases are reported in young adults and can arise in any body site. Primary orbital synovial sarcoma is rare.

***CASE SUMMARY***

An 8-year-old east Asian girl with 1-mo history of gradual painless proptosis and lacrimation of the right eye was admitted. The patient presented with painless proptosis, downward eyeball displacement, and upward movement disorders. According to clinical manifestations, imaging examinations and postoperative immunohistochemical examinations, the diagnosis was monophasicsynovial sarcoma with calcification. The patient underwent anterior orbitotomy for removal of the right orbital mass under general anesthesia. The diagnosis of monophasicsynovial sarcoma with calcification was confirmed through histological and immunohistochemical examination. The follow-up period was 6 mo, and no recurrence was observed during this period.

***CONCLUSION***

Primary orbital monophasicsynovial sarcoma is rare. Surgical resection with adjuvant or neoadjuvant radiotherapy is highly effective for localized tumors.

**Key Words:** Orbital tumor; Synovial sarcoma; Calcification; Histological; Case report

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**Citation:** Ren MY, Li J, Li RM, Wu YX, Han RJ, Zhang C. Primary orbital monophasic synovial sarcoma with calcification: A case report. *World J Clin Cases* 2022; 10(5): 1623-1629

**URL:** https://www.wjgnet.com/2307-8960/full/v10/i5/1623.htm

**DOI:** https://dx.doi.org/10.12998/wjcc.v10.i5.1623

**Core Tip:** We describe a patient with 1-mo history of gradual painless proptosis and lacrimation of the right eye. The patient underwent anterior orbitotomy for removal of the right orbital mass under general anesthesia. The diagnosis of monophasicsynovial sarcoma with calcification was confirmed through histological and immunohistochemical examination. The follow-up period was 6 mo, and no recurrence was observed during this period. This case illustrates that the tumor may present with similar features as a benign tumor. Comprehensive analysis of clinical, radiological and pathological findings is important for making the right diagnosis.

**INTRODUCTION**

Synovial sarcoma is a malignant mesenchymal neoplasm with variable epithelial differentiation. It mainly occurs in young adults and can arise at several sites[1]. It mainly commonly occurs in deep soft tissue of the extremities in adolescents and young adults[2]. Synovial sarcoma of the head and neck region is rare, and only a few cases of sarcoma arising from the orbit have been reported[3,4]. The current study reports a case of primary orbital monophasic synovial sarcoma which was characterized by calcification in an 8-year-old patient.

**CASE PRESENTATION**

***Chief complaints***

An 8-year-old East Asian girl with 1-mo history of gradual painless proptosis and lacrimation of the right eye was admitted to our hospital.

***History of present illness***

The patient presented with gradual painless proptosis and lacrimation of the right eye for 1 mo. The proptosis was gradual and painless. No special treatment was performed, and there was no other significant change in ocular symptoms.

***History of past illness***

The patient had no history of any previous disease.

***Personal and family history***

There was no family history of malignant neoplasm.

***Physical examination***

Physical examination showed no neurological signs. The patient presented with painless proptosis, downward eyeball displacement, and upward movement disorders. Ocular examination showed that the binocular best-corrected visual acuity was 20/20. Eye examination did not show any significant eyelid, conjunctival, corneal and lenticular abnormalities, and fundus examination did not show any abnormalities. Hertel exophthalmometry analysis showed that the right eye was 18 mm and whereas the left eye was 13 mm, and the interorbital distance was 91 mm. Intraocular pressure was 13 mmHg in the right eye and 11 mmHg in the left eye.

***Laboratory examinations***

Blood and urine tests were normal.

***Imaging examinations***

A/B-scan showed moderate echogenic lesions in the right eye orbital. Echoes were uneven, well-distributed and sound transmission was normal. Patchy strong echoes and sound shadows were detected (Figure 1). Orbital computed tomography showed a well-defined soft tissue density mass in the right orbit, with flaky high-density shadows observed inside the right orbit. The size of the mass was approximately 20 mm × 20 mm × 19 mm, and exophthalmos; extraocular muscles and optic nerve were compressed (Figure 2). Orbital magnetic resonance imaging showed a circular-like mass in the right orbital. T1-weighted imaging (T1WI) showed moderate signals, whereas T2-weighted imaging (T2WI) showed mixed signals, with a high number of moderately high signals. T1WI and T2WI were characterized by low-signal regions. Most of the lesion was significantly and unevenly enhanced, whereas local lesions did not exhibit any enhancement (Figure 3).

***Primary diagnosis***

Based on the findings described above, the preliminary diagnosis was rhabdomyosarcoma or other malignant neoplasm.

**FINAL DIAGNOSIS**

Histological examination showed that the tumor was monophasicsynovial sarcoma with calcification. Immunohistochemical analysis showed positive staining for CD34, CD99, Bcl-2, CKpan, TLE1, INI-1 and Ki-67 (25%), and negative staining for SMA, vimentin, myogenin, myoglobin, Syn, CgA, NSE, S-100, PGP9.5, EMA, CK7, CK (AE1/AE3), CD65, calretinin, TTF1 and MUC-4 (Figure 4).

**TREATMENT**

After preoperative examination, the patient underwent anterior orbitotomy for removal of the right orbital mass under general anesthesia. The operation showed an oval tumor above the optic nerve in the right orbit. The tumor margins were well defined; however, the tumor was large, reddish, unmovable, and adhesive to the levator palpebrae muscle (Figure 5). The levator palpebrae muscle was cut along its path, and the tumor was carefully separated from the muscle and removed. The levator palpebrae muscle was sutured before the end of the operation. After treatment, the patient was transferred to the tumor hospital and underwent systemic chemotherapy.

**OUTCOME AND FOLLOW-UP**

The follow-up period was 6 mo, and no recurrence was observed during this period.

**DISCUSSION**

Synovial sarcoma accounts for 10%–20% of soft tissue sarcomas. It is a high-grade soft-tissue sarcoma occurring mainly in older children and young adults. Approximately 7% of soft tissue sarcomas occur in the head and neck region, and synovial sarcoma represents < 0.1% of all head and neck cancers[5,6]. Orbital synovial sarcoma is a rare malignancy. Therefore, diagnosis of orbital synovial sarcoma in clinical practice is challenging, and requires an integrated approach that incorporates specific clinical, histological, immunohistochemical and molecular analyses.

Synovial sarcoma is a rare orbital tumor and the clinical characteristics have not been fully elucidated. Clinical manifestations include gradual painless proptosis, eyelid swelling, a palpable painless mass, epiphora, ptosis, and periorbital spontaneous pain or tenderness. However, these clinical manifestations are not unique to synovial sarcoma. Characteristic findings are not reported in current imaging studies due to the small number of cases. A case of monophasic synovial sarcoma primarily arising in the left superonasal orbital region was reported in a 24-year-old woman, which was clinically mistaken for a periocular cyst[6]. However, the lesion was characterized by calcification similar to the current case.

The calcification can be pathologically divided into dystrophic and metastatic. In the current case, the growth of the lesion was rapid, resulting in ischemia and necrosis of the tumor. Significant calcification of the lesion may be caused by dystrophic calcification. Occurrence of a lesion with calcification in the orbit of a pediatric patient, orbital vascular lesions or malformations, and orbital malignancies should be explored when carrying out diagnosis. Irregular calcification is common in malignant and partially benign tumors. Orbital tumors in children are associated with a higher incidence of rhabdomyosarcoma[7]. In addition, most common soft-tissue sarcomas in children are reported in the head and neck, with 10% of all cases occurring in the orbit. A detailed history is essential if a child is suspected to have rhabdomyosarcoma[8]. A case of recurrent primary orbital calcified synovial sarcoma in a young woman was previously reported[4]. In addition, diagnosis should distinguish the primary lesion from metastatic lesions, as metastatic synovial sarcomas are characterized by poor prognosis[9]. The current case showed no other systemic lesions and was a primary tumor and not a recurrent case.

Orbital vascular lesions or malformations, other benign lesions and orbital malignancies should be considered when there is a lesion characterized by calcification in the orbit during childhood. Irregular calcification is common in malignant and partially benign lesions. In the current case, orbital computed tomography and magnetic resonance imaging showed a well-defined soft tissue dense mass in the right orbit. The tumor may present with similar features as benign tumors. Differential diagnosis may identify findings that do not fit preliminary diagnosis of benign tumors. In such cases, comprehensive consideration of clinical, radiological and pathological findings is important[10].

Synovial sarcoma is a highly malignant soft tissue sarcoma, with poor survival of patients. Conventional treatment approach is surgical resection with adjuvant or neoadjuvant radiotherapy, which are highly effective for localized tumors. Synovial sarcoma is sensitive to chemotherapy. Ifosfamide alone and ifosfamide combinations are effective for treatment of synovial sarcoma[5,11,12]. Combination of doxorubicin and ifosfamide is the preferred first-line therapy for patients with metastatic tumors. Sequential doxorubicin and ifosfamide can be considered for localized tumors. Pazopanib and trabectedin are effective as second-line therapy and for subsequent treatment[11].

A previous study reported high local recurrence rates despite surgery and postoperative radiotherapy and adjuvant chemotherapy, and distant metastasis rates were not reduced by these approaches[2]. The disease is characterized by early and late recurrences, and the 10-year disease-free survival is ~50%[5]. Several new approaches for treatment of metastatic synovial sarcoma are currently under investigation, both at preclinical and clinical levels, including receptor tyrosine kinase inhibitors, epigenetic modulators, compounds interfering with DNA damage response, and immunotherapy[11].

Histological analysis shows that synovial sarcoma is monophasic, biphasic, or poorly differentiated and exhibits a specific chromosomal translocation t (X; 18) (p11.2; q11.2) in > 95% of cases[6]. Genetic analysis shows that synovial sarcoma tumors have a characteristic fusion protein, SS18–SSX, implicated in promoting disease development. BRD9 is a component of SS18–SSX containing BAF complexes in synovial sarcoma cells. Studies report that BRD9 is implicated in oncogenic mechanisms underlying the SS18–SSX fusion in synovial sarcoma and targeted degradation of BRD9 is a potential therapeutic approach for treatment of synovial sarcoma[13].

**CONCLUSION**

These findings show that primary orbital synovial sarcoma with calcification is rare, and clinical manifestations and imaging results are not specific. The tumor may exhibit similar features as a benign tumor. Therefore, these cases require comprehensive clinical, radiological and pathological analysis to achieve the right diagnosis. The conventional treatment is surgical resection with adjuvant or neoadjuvant radiotherapy, which are highly effective for localized tumors. However, a longer follow-up time is required to determine effectiveness of the treatment.

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**Footnotes**

**Informed consent statement:** Informed written consent was obtained from the patient for publication of this report and any accompanying images.

**Conflict-of-interest statement:** The authors declare that they have no conflict of interest.

**CARE Checklist (2016) statement:** The authors have read the CARE Checklist (2016), and the manuscript was prepared and revised according to the CARE Checklist (2016).

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**Provenance and peer review:** Unsolicited article; Externally peer reviewed.

**Peer-review model:** Single blind

**Peer-review started:** July 31, 2021

**First decision:** October 25, 2021

**Article in press:** December 28, 2021

**Specialty type:** Ophthalmology

**Country/Territory of origin:** China

**Peer-review report’s scientific quality classification**

Grade A (Excellent): A

Grade B (Very good): 0

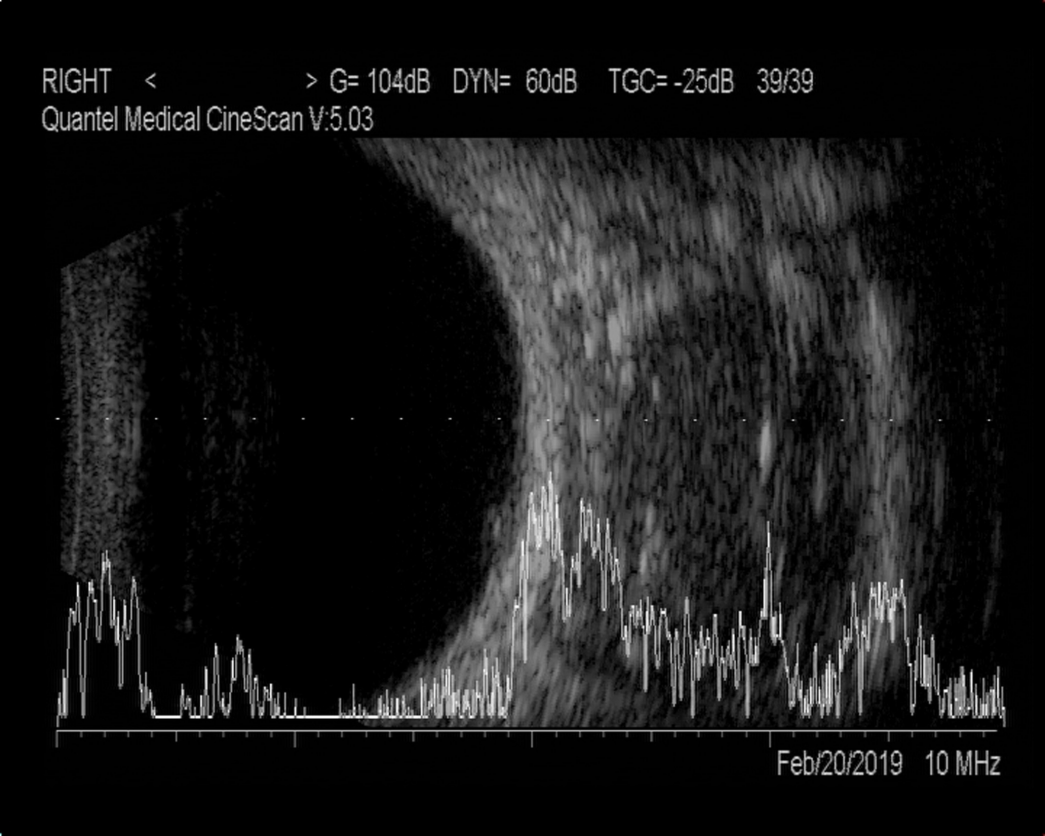
Grade C (Good): 0

Grade D (Fair): 0

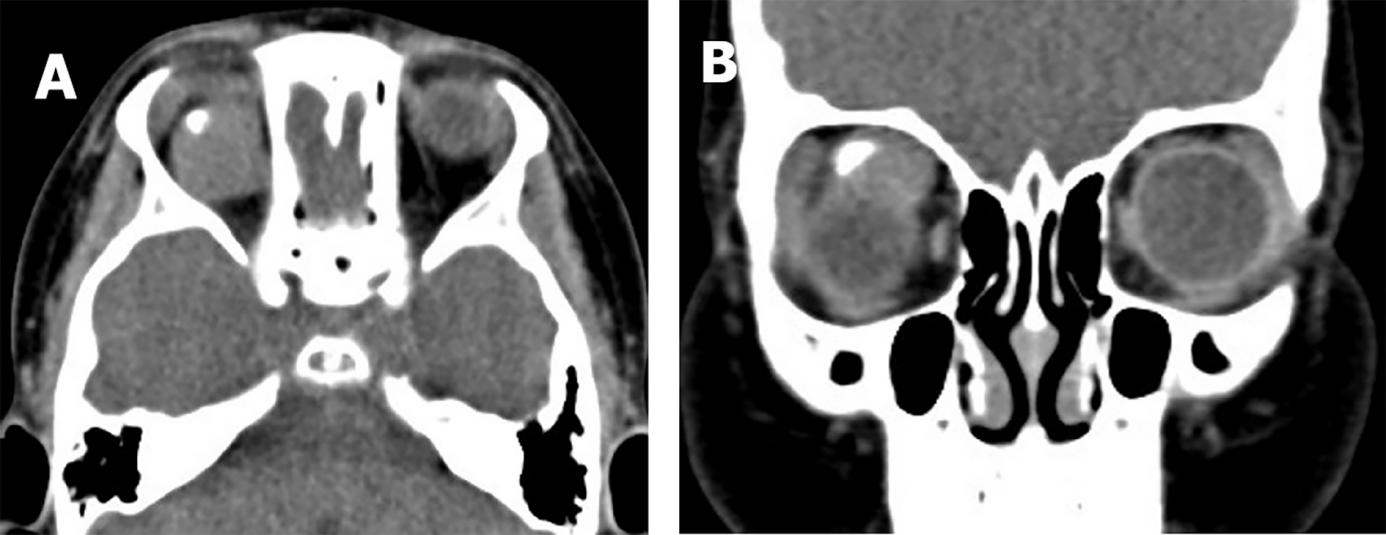
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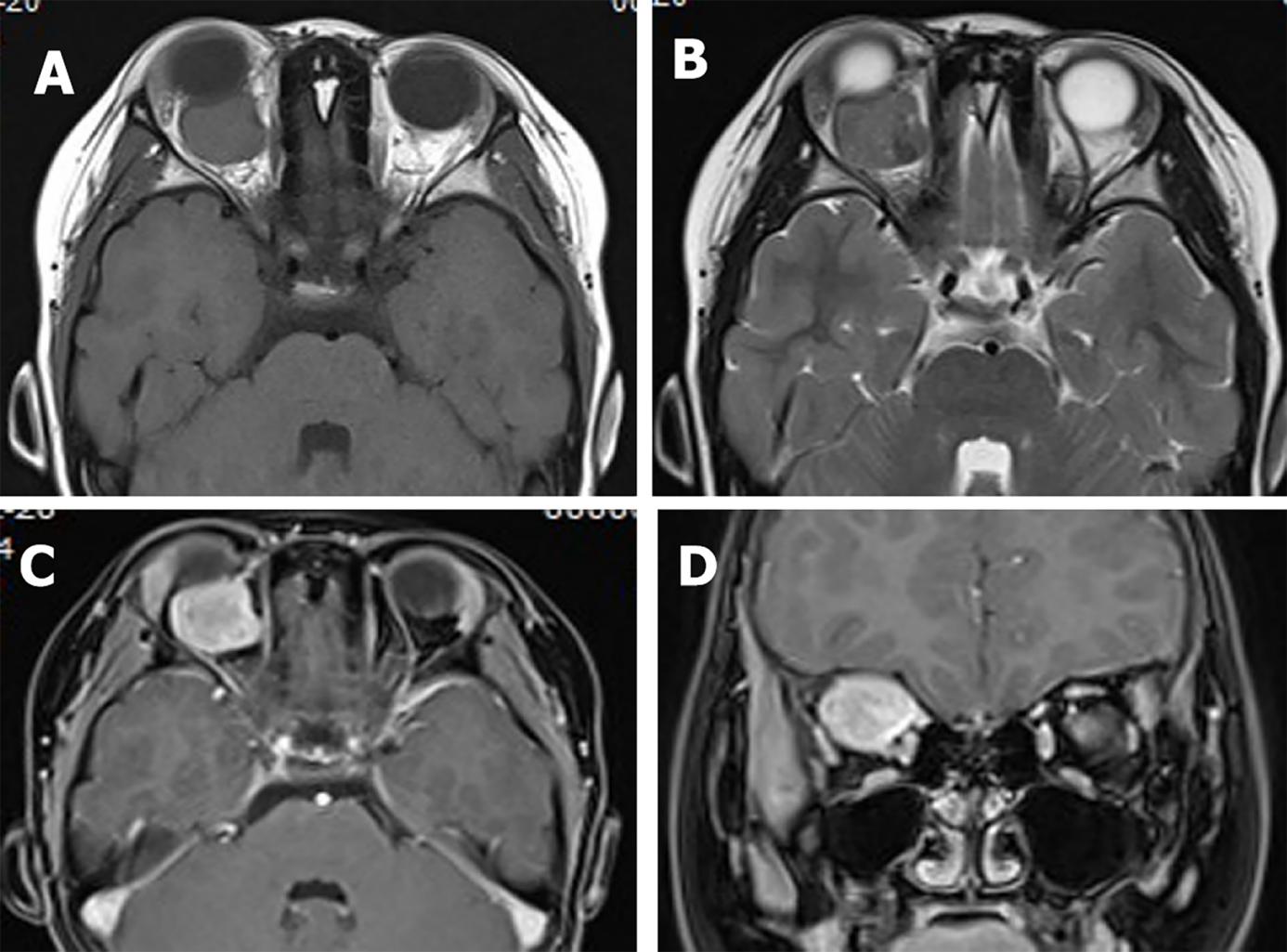
**Figure Legends**



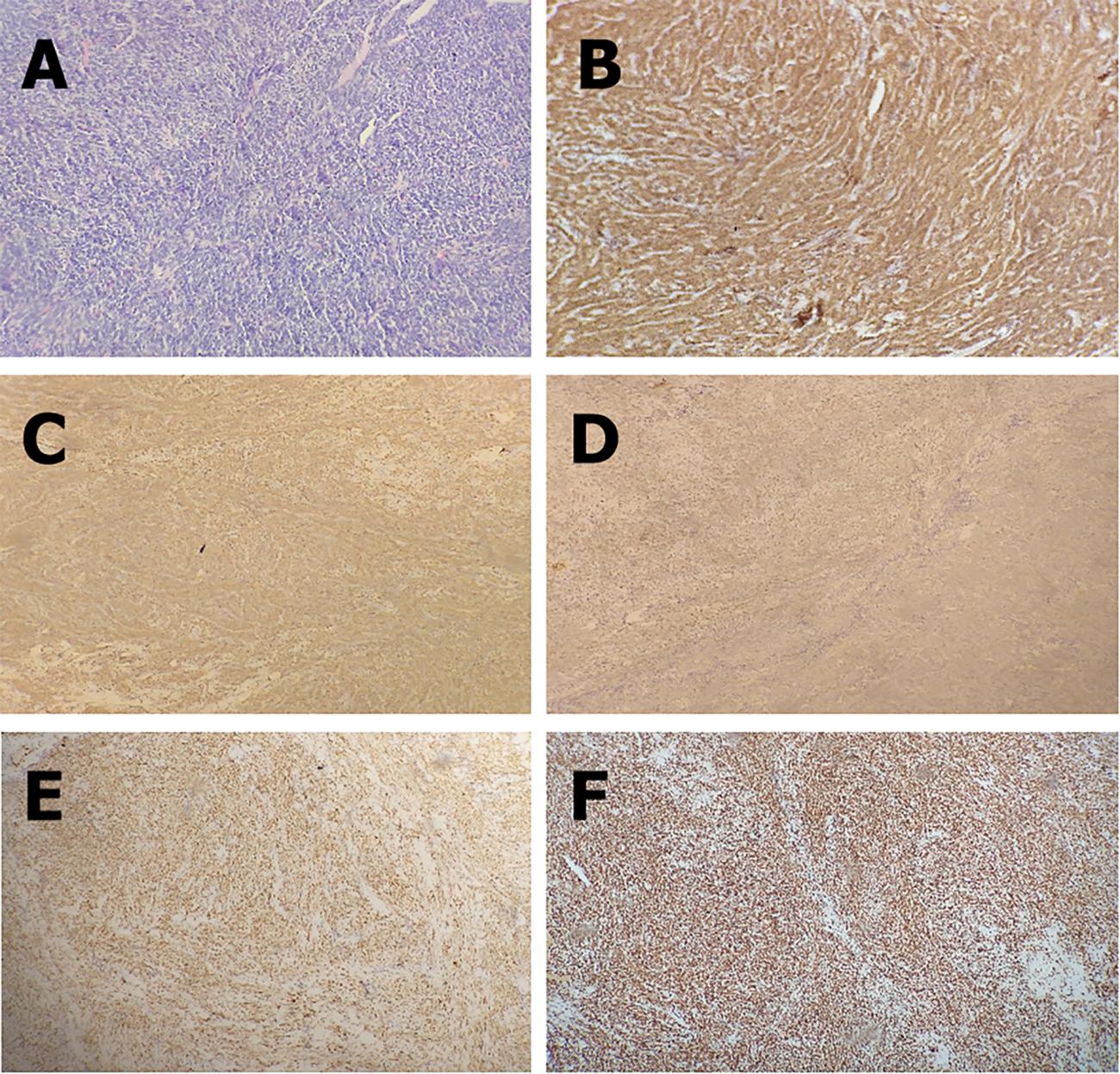
**Figure 1 A/B-scan showed moderate echogenic lesions in the right orbit.**



**Figure 2 Orbital computed tomography showed a well-defined soft tissue density mass in the right orbit, with a hyperdense speck suggestive of coarse calcification.** A: Axial computed tomography (CT); B: Coronal CT.



**Figure 3 Orbital magnetic resonance imaging showed a circular-like mass in the right orbit.** A: T1-weighted imaging showed moderate signals, mixed with low-signal regions; B: T2-weighted imaging showed mixed signals, with high number of moderately high signals, and mixed with low-signal regions; C and D: Most of the lesion was significantly and unevenly enhanced, whereas local lesions did not exhibit any enhancement.



**Figure 4 Histological and immunohistochemical examination.** A: The tumor was confirmed as monophasicsynovial sarcoma with calcification based on histological analysis (hematoxylin and eosin, 200×); B–F: Immunohistochemical study revealed positive staining for Bcl-2 (B), CD99 (C), CKpan (D), TLE1 (E), and INI-1 (F) (100×).



**Figure 5 Tumor presented as a well-defined, reddish lesion, with irregularly shaped soft tissue density mass.**



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