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Alveolar soft part sarcoma of the lateral rectus muscle: Suture technique to prevent postoperative strabismus

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ABSTRACT

Purpose: We present a rare case of intramuscular alveolar soft part sarcoma (ASPS) of the lateral rectus (LR) muscle and the surgical technique used to maintain orthotropia after complete resection of the mass.

Observations: A 5-year-old boy presented with progressive proptosis of the left eye due to an orbital tumor. The patient was previously diagnosed with ASPS of the left LR muscle from an incisional biopsy, and the tumor size increased despite 5 cycles of chemotherapy prior to presenting to our center. Magnetic resonance imaging showed a 28x19x15mm-sized contrast-enhancing intramuscular mass of the left LR muscle, and there was no evidence of nodal or distant metastasis. The mass was excised en bloc, along with the insertion and the posterior normal part of LR muscle. To maintain proper eye alignment after resecting LR muscle, a 4-0 Prolene® hang-back suture was placed between the scleral insertion and the periorbita of the posterior orbit and the left medial rectus muscle was injected with botulinum toxin. During the follow-up of 51 months after surgery, the patient had no evidence of recurrence or metastasis and remained orthotropic in primary gaze, with a good cosmetic result.

Conclusions and Importance: ASPS of extraocular muscles is a rare tumor occurring mainly in children and young adults, and treatment may cause significant sequelae such as orbital exenteration, radiation-induced complications, and large-angle strabismus. Complete resection of tumor including the extraocular muscle is essential for treatment, and a subsequent reconstruction using a hang-back suture technique is useful to achieve proper eye alignment as well as a good cosmetic outcome.

1. Introduction

Alveolar Soft Part Sarcoma (ASPS) is a rare malignant tumor of children and young adults first described by Cristopherson in 1952.[1] It represents 0.5%–1% of all soft tissue sarcomas and commonly arises in the extremities and trunk,[2] along with cases reported in the orbit,[3] tongue,[4] and abdomen.[5] Difficulty in diagnosing ASPS lies in its ambiguous presentation, as symptoms mostly arise as a result of metastasis.[6] It is also radiologically similar on CT or MRI to hemangiomas resulting in some instances of misdiagnosis and delay in treatment.[7]

Although ASPS of the orbit are particularly rare, several case reports have been published focusing on presentation, diagnosis and outcome of treatment.[3] The mainstay of treatment in the literature remains to be surgical excision of the tumor along with safe margins, many times resulting in orbital exenteration and cosmetic disfigurement.[8] In this case report we present a patient with ASPS of the lateral rectus (LR) muscle, who underwent surgical excision of the tumor and reconstruction of the LR muscle with a suture method allowing him to maintain a disease-free follow-up and an excellent cosmetic result with orthotropia in primary gaze.

2. Case report

A 5-year-old Korean boy was referred to the ophthalmology service of our center with a complaint of left orbital mass. Three years prior to presenting to us, the patient first visited to another hospital with subconjunctival hemorrhage and progressive proptosis of the left eye. An orbital magnetic resonance imaging (MRI) revealed a 25x19x17mm-

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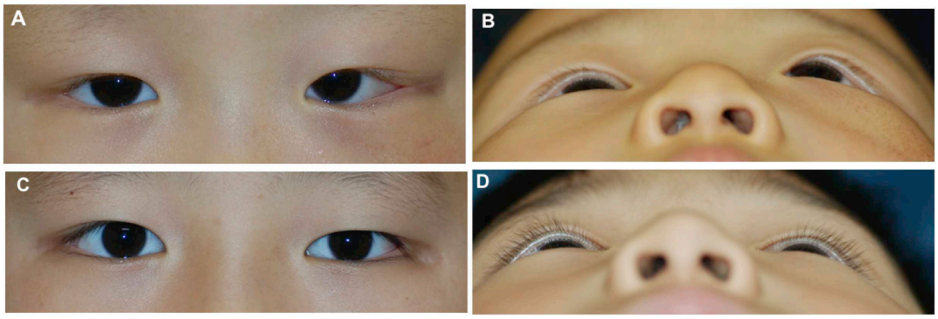

Fig. 1. Clinical photos.
(A and B) A 5-year-old boy with alveolar soft part sarcoma of the left lateral rectus muscle presented with progressive proptosis of the left eye despite chemotherapy. (C and D) The patient has shown orthotropia in primary gaze and complete resolution of proptosis for 51 months after surgery.

sized tumor of the left lateral orbit with involvement of LR muscle, and an incisional biopsy revealed ASPS. Systemic evaluations did not show evidence of metastasis. The patient underwent chemotherapy, but the tumor size increased despite 5 cycles of chemotherapy. The patient was recommended to undergo exenteration, and he visited our center for a second opinion.

External examination revealed left eye proptosis of 3 mm by Hertel exophthalmometry, orthotropia in primary gaze, and severe limitation of abduction of the left eye (Fig. 1). The patient had normal visual acuity in both eyes and +1.5D astigmatism in the left eye. An orbital MRI showed a 28x19x15mm-sized contrast-enhanced mass of the left LR muscle causing compression to left eyeball, but the tumor appeared well-demarcated within the muscle itself (Fig. 2). Positron emission tomography-computed tomography (PET-CT) revealed no evidence of metastasis.

A lateral orbitotomy with bone window via swinging eyelid approach was performed for a complete excision of the tumor. After a marginotomy of the lateral orbital rim, the periorbita was incised and the lateral rectus muscle was exposed. Dissection of the mass of the LR muscle was carried out until the mass was completely exposed (Fig. 3A). The mass was completely excised, including the insertion of the LR muscle and the posterior normal part of the LR muscle. To prevent a large esotropia, a hang-back suture of 4–0 Prolene® was made between the sclera at the original insertion of LR muscle and the periorbita of the posterior orbit. The length of the thread was adjusted so that the eyeball was positioned correctly. The medial rectus (MR) muscle of the left eye was injected with 15 units of botulinum toxin (Botox®) to ensure proper power balance and orthotropia. The bone was reinserted and the surgical site was closed. (Video 1).

Supplementary video related to this article can be found at https://doi.org/10.1016/j.ajoc.2020.100668

Histopathologic examination showed an organoid nests of polygonal eosinophilic cells arranged in an alveolar pattern separated by fibrovascular septa. There was no lymphovascular or perineural invasion of tumor cells, and the resection margins were free from tumor cells (Fig. 3B and C). Granular cytoplasm was positive for periodic acid-Schiff (PAS). Immunohistochemical staining revealed the tumor is positive for an antibody that detects the carboxyl terminal portion of the transcription factor E3 (TFE3) gene. These results were supportive for a diagnosis of ASPS.

There were no significant postoperative complications, and the patient had no adjuvant irradiation or chemotherapy. He has been regularly followed-up for 51 months after surgery, and there have been no signs of recurrence or metastasis based on ophthalmic examination, orbital MRIs, and PET-CTs. On the last follow-up, the patient was orthotropic with limited horizontal movement of the left eye (Fig. 1C), but he did not complain of any subjective diplopia in his daily life.

3. Discussion

ASPS is a rare tumor with ambiguous histological origins and clinical presentation.[9] De Barros GF et al.[10] reviewed the literature in 2018 and found 64 cases of primary orbital ASPS, with 60% occurring in females. To our knowledge, there have been only 26 reported patients with orbital ASPS of extraocular muscles in the literature, with 56% of orbital ASPS occurring in the left orbit, and the majority of affected patients being below 20 years of age.[3] Survival rates were reported to be 60% and 15% at 5 and 20 years, respectively, with metastases occurring in the lungs, bones, and brain.[2] ASPS is a highly vascular tumor, which lends difficulty in diagnosis based on imaging alone.[7] Genetics, histopathology and immunohistochemistry are more definitive methods to diagnose ASPS as the tumor arises due to a translocation of t (17)t (X:17) (p11; q25), leading to the high expression of TEF3 gene apparent on immunohistochemistry.[11] We also found that detection of TFE3 gene expression is useful to confirm the diagnosis of ASPS in the current case.

Radical surgical resection is the most advisable treatment for localized ASPS, and irradiation can be added when the surgical margin is questionable.[12] Due to the resistance to chemotherapy and irradiation, however, it is often needed to perform more aggressive surgical resection, including exenteration, for local control of orbital ASPS.[13,14] Neoadjuvant chemotherapy could be considered as an attempt to avoid exenteration, it is of note that chemotherapy is largely inactive, with response criteria in solid tumors (RECIST) rates lower than 10%.[14,15] Given the young age in which orbital ASPS presents, and the social and psychological effects that external appearance has on children and adolescents, particular attention should be also paid to cosmesis

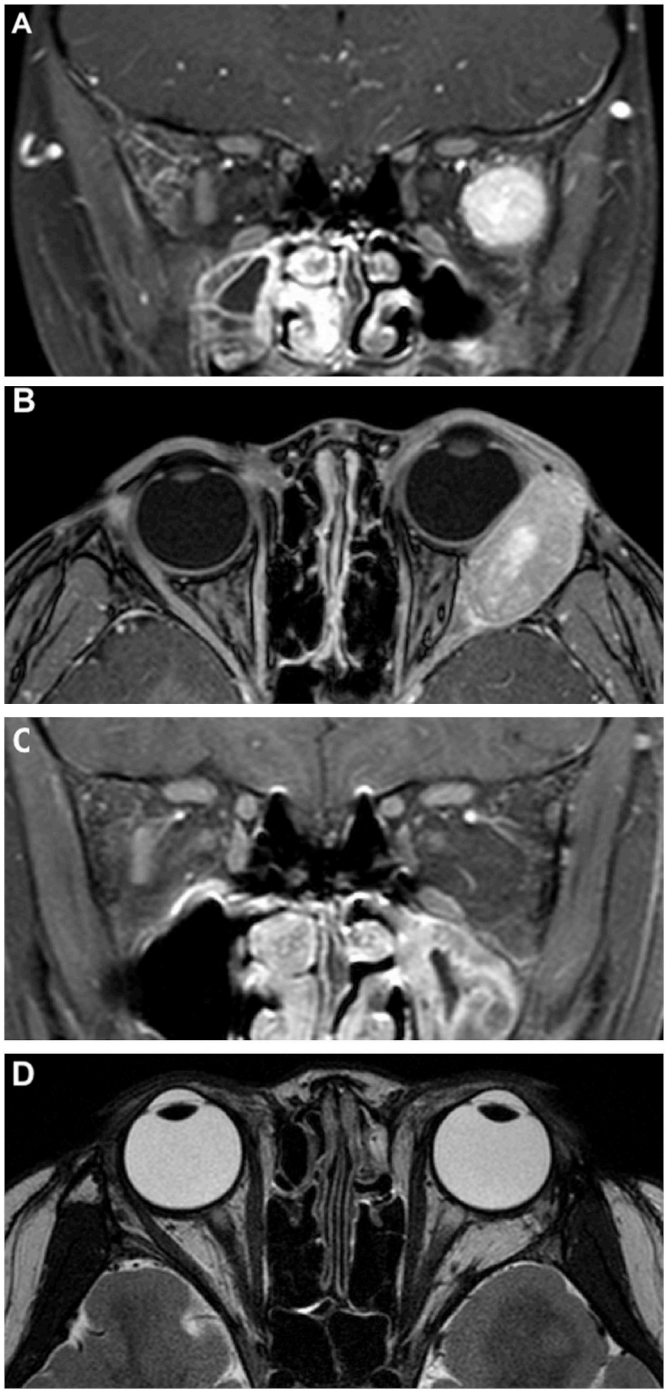

Fig. 2. Radiological images.
(A) Preoperative coronal and (B) axial MRIs with Gadolinium enhancement revealed a contrast-enhancing lesion of the left lateral rectus muscle, compressing the left eyeball. (C) Postoperative coronal and (D) axial MRIs taken 51 months after surgery showed no evidence of remnants or recurrence of the tumor.

following removal of the tumor. In the current case, tumor was resistant to chemotherapy conducted elsewhere and an imaging study showed a well-demarcated mass in the LR muscle, and we were able to achieve an en bloc resection of the tumor. We did not evaluate intraoperative resection margin status in the current case because there was no clinical suspicion of tumor involvement during surgery, and clear resection margins were confirmed on postoperative histopathologic examination. However, intraoperative control of surgical margins may be appropriate for tumors with findings suggestive of tumor infiltration to guide further resection or adjuvant treatments. Adjuvant irradiation was not performed because of postoperative clear resection margins as well as concerns about radiation-induced complications such as second malignancy and orbital bone deformation.[16] However, adjuvant irradiation should be considered to improve local control of ASPS, especially in patients with positive or uncertain resection margins after surgery.[13,15]

Regarding ASPS of extraocular muscles, surgical resection of the tumor may result in large strabismus. In the current case, after complete resection of the tumor of the LR muscle, we used a hang-back suture technique to maintain orthotropia in primary gaze. Prolene® suture was used in the current case, and a coated and braided non-absorbable suture such as Surgidec® would be also beneficial. We also injected a botulinum toxin into the antagonist muscle which might be useful to prevent esotropia until the stabilization of hang-back suture. This surgical technique reserves the tension that was placed on the eyeball prior to surgery and keeps the eye in an orthotropic position while in primary gaze. The patient has been maintaining orthotropia without recurrence or metastasis for 51 months after surgery and was pleased with the cosmetic result as well.

4. Conclusion

We present a rare case of ASPS of the LR muscle in 5-year-old boy who underwent successful surgical treatment with tumor resection followed by muscle reconstruction using the suture technique.

Patient consent

Written consent was obtained from the patient's parents/guardians.

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Authorship

All authors attest they meet the current ICMJE criteria for authorship.

Declaration of competing interest

All authors have no financial disclosures.

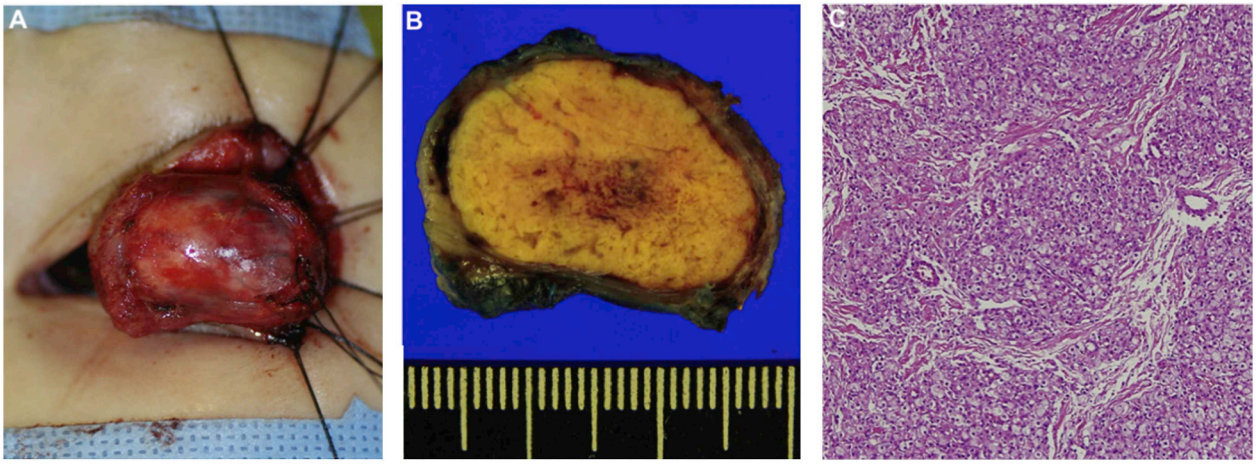

Fig. 3. Intraoperative and histopathologic images.
(A) A lateral orbitotomy with bone window was performed to excise alveolar soft part sarcoma of the lateral rectus muscle. (B) A longitudinal section of the specimen showed a completely excised and well-demarcated mass. (C) Histopathologic examination (H&E, magnification ×200) revealed an alveolar pattern of organoid nests of polygonal eosinophilic cells separated by fibrovascular septa, supporting the diagnosis of alveolar soft part sarcoma.

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None.

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