

Retro-orbital alveolar soft-part sarcoma in a 76-year-old female: case report and review of the literature

Alveolar soft part sarcoma (ASPS) is a rare mesenchymal neoplasm that represents less than 1% of all sarcomas.¹ Orbital ASPS is extremely rare, representing only 5%–15% of all cases of ASPS.² The average age at presentation of orbital ASPS is 13.5–17.8 years. A recent review of the literature suggests there are fewer than 70 documented cases of primary orbital ASPS.¹

We present a case of a 76-year-old female with binocular diplopia secondary to a right retro-orbital ASPS. To the best of our knowledge, this is the oldest patient to date to present with a primary orbital ASPS. This study adhered to the Declaration of Helsinki and was compliant with the Health Insurance Portability and Accountability Act of 1996.

A 76-year-old White female with no past ocular history presented with complaints of worsening binocular diplopia in primary gaze and right gaze over 6 weeks. On presentation, the patient's visual acuity was 20/30-2 OD and 20/30-2 OS; intraocular pressure and pupils were normal. Extraocular motility testing demonstrated a moderate supraduction deficit and a mild abduction deficit OD. External examination was significant for right exophthalmos. Hertel measurements demonstrated 2 mm of proptosis OD. Fundus examination was significant for mild disc edema OD. Humphrey 24-2 visual field testing demonstrated scattered superior deficits OD. Optical coherence tomography of the retinal nerve fibre layer was significant for mild thickening of the optic disc OD.

Magnetic resonance imaging of the orbits was performed, which revealed a well-circumscribed 1.9 × 1.7 × 1.3 cm right retro-orbital mass with associated mass effect on the right optic nerve and right medial rectus (Fig. 1A, B). Given the concern for tumour vascularity on neuroimaging, the patient was seen by the interventional radiology service for preoperative cerebral and orbital angiogram with planned embolization of the tumour (Supplementary Fig. 2, available online). Despite multiple attempts, the IR service was unable to embolize the tumour. The patient was seen elsewhere for a second attempt at embolization. The patient underwent cerebral angiography and attempted transarterial embolization via the right ophthalmic artery, but this was only partially successful, and therefore, the procedure was completed via percutaneous direct stick embolization along the superomedial right orbit (Supplementary Fig. 3, available online). This procedure was complicated by a central retinal artery occlusion OD and ischemic optic neuropathy OD. Of note, the patient's visual acuity decreased to light perception OD after this procedure.

The following day, the patient underwent a right lateral orbitotomy with bone flap for excisional biopsy of the mass

(Supplementary Video 1, available online). Histopathologic analysis demonstrated large polygonal neoplastic-appearing cells arranged in sheets and smaller nests separated by fibrous connective tissue associated with sinusoidal vasculature. Tumour necrosis was noted. The nuclei of the cells were surrounded by eosinophilic granular cytoplasm with very prominent cell borders (Supplementary Fig. 4, available online). Isolated cells also demonstrated crystals within the cytoplasm that were arranged in stacks. By immunohistochemistry, the neoplastic cells were positive for antibodies to TFE3 (nuclear; Supplementary Fig. 2, available online), CD68 (cytoplasmic granular), and SDHB (granular, intact) while negative for CKpan, CAIX, SF1, PAX8, STAT6, SMA, desmin, myogenin, S100 protein, SOX10, Melan-A, HMB45, INSM1, chromogranin, and synaptophysin. The diagnosis of ASPS was rendered.

At her 1-month postoperative visit, the patient was noted to have a visual acuity of no light perception OD, a relative afferent pupillary defect OD; a-3 supra-, infra-, ab- and adduction deficits OD; significant ptosis OD with 5 mm of lagophthalmos OD and significant optic disc edema; peripapillary heme; and a cherry-red spot—appearing macula on fundus examination OD. Repeat magnetic resonance imaging demonstrated residual enhancing tissue measuring 1.5 × 2 × 1.6 cm. The patient was recently presented at the tumour board and was recommended to undergo exenteration of the right orbit; however, the patient declined this treatment and is considering radiation therapy.

To the authors' knowledge, this is the oldest patient presenting with orbital ASPS. Orbital ASPS usually presents during the first or second decade of life, with the oldest case thus far documented at 69 years of age.^{1,3} In addition, most patients complain of either proptosis or conjunctival edema on initial presentation.¹ The patient's age, coupled with her initial complaints of binocular diplopia, makes her a rarity in comparison with previously reported patients with orbital ASPS.

Of note, the patient first underwent cerebral angiography and embolization of the tumour to prevent retrobulbar bleeding during lateral orbitotomy. Unfortunately, the patient suffered a central retinal artery occlusion and ischemic optic neuropathy of the right eye during this procedure. Neurologic complications following cerebral angiography are extremely rare (0.1%–0.5%), and the patient's visual outcome after embolization was unexpected.⁴ It is unlikely, however, that a successful orbitotomy could have been performed without prior embolization given the location and vascularity of the tumour.²

By histopathology, ASPS is characterized as having a nested architecture with fibrous septae.⁵ The cells of the tumour can be large and contain abundant granular eosinophilic or clear cytoplasm.⁵ Interestingly, crystalline cytoplasmic inclusions are seen on histopathology, usually highlighted by a periodic acid–Schiff stain with diastase resistance.⁵ Moreover, the neoplastic cells will be immunoreactive with TFE3, which is considered a very sensitive

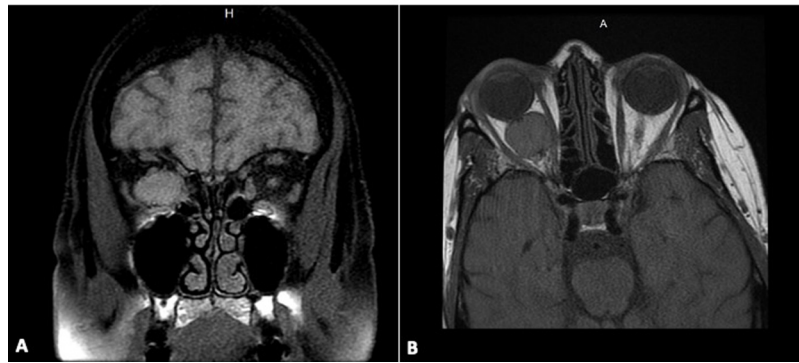


Fig. 1—(A) Coronal, T1-weighted fat-saturated magnetic resonance image demonstrating a well-circumscribed $1.9 \times 1.7 \times 1.3$ cm right retro-orbital mass with associated mass effect of the right optic nerve and right medial rectus. (B) Axial T1-weighted magnetic resonance image demonstrating a well-circumscribed $1.9 \times 1.7 \times 1.3$ cm right retro-orbital mass with associated mass effect of the right optic nerve and right medial rectus.

marker, although not entirely specific for ASPS.¹ Histologic analysis of our patient demonstrated similar findings.

The mainstay of treatment is surgical excision of the tumour with wide margins.⁶ Exenterative surgical resection portends a better survival rate.¹ As with our patient, this may be particularly difficult depending on the tumour location. The role of radiotherapy and chemotherapy in orbital ASPS remains unclear but is an option for unresectable or recurrent disease.¹ The overall prognosis of orbital ASPS is poor. The 5- and 20-year overall survival rates of ASPS are 60% and 15%, respectively.¹ Although our patient did not have metastatic disease at presentation, the size of the tumour, her advanced age, the presence of lymphovascular invasion, and tumour necrosis are considered negative risk factors for overall and disease-free survival.⁵

In conclusion, we present a case of a 76-year-old female with binocular diplopia secondary to a right retro-orbital ASPS. To the best of our knowledge, this is the oldest patient presentation of primary orbital ASPS to be documented. Orbital ASPS should be considered in the differential of primary orbital neoplasms, even in the elderly population.

Supplementary Materials

Supplementary material associated with this article can be found in the online version at doi:[10.1016/j.jcjo.2022.03.007](https://doi.org/10.1016/j.jcjo.2022.03.007).

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Footnotes and Disclosure

The authors have no financial disclosures or conflicts of interest to declare.