Case report

Alveolar soft part sarcoma of the orbit, a case report of a rare tumor

Ángel Nava-Castañeda, José Luis Tovilla-Canales, Francisca Zuazo,
Lourdes Rodríguez-Cabrera, Lilia Garnica-Hayashi
Instituto de Oftalmología Fundación de Asistencia Privada Conde de Valenciana
Colonia Obrera, México DF. CP 06800

Abstract

Background: Alveolar Soft Part Sarcoma is a rare malignant tumor of uncertain histogenesis, representing 0.5-1% of all soft tissue tumors. It occurs predominantly in head and neck regions, especially the orbit and the tongue, in infants, causing no specific symptoms for an extended period of time. **Case:** A four year old girl was brought to consultation because of persistent swelling of her left upper and lower eyelid for the past six months. A 4 mm proptosis was noted on her left eye with significant upwards displacement of the globe. A firm, not pulsating and non tender mass was palpated, and motility examination revealed deficient abduction and infraduction. Visual acuity was 20/40 and 1/200 in the right and left eye, respectively. The computed tomography revealed an extraconal mass on the inferotemporal aspect of the left orbit, with no bony erosion or globe invasion. An excisional biopsy was made, finding a tumor with nests of clusters of large polygonal cells, separated by fibrous septa and a sinusoidal vascular channel. An Alveolar Soft Part Sarcoma was diagnosed. Our patient had a favorable post-operative follow up, and oncologic evaluations have not shown metastasis or local recurrences. **Observations and Conclusions:** Alveolar Soft Part Sarcoma is a rare malignant tumor of indolent course, but with propensity to distant metastases, making early diagnose and long term follow up necessary.

Keywords: Alveolar soft part sarcoma, proptosis, orbital tumor.

Introduction

Alveolar soft part sarcoma (ASPS) is a clinically and morphologically distinct soft-tissue sarcoma, first defined and named by Christopherson et al (1952) (Kashyap et al, 2004; Alkatan et al 2010). It is a rare and histologically distinct soft tissue malignancy of uncertain histogenesis (Kanhere et al, 2005; Wang et al, 2015). The tumor received its name due to its pseudo-alveolar histological appearance (Kayton et al, 2006).

Received on: 22/06/16 Accepted on: 25/12/16
Address for correspondence
Ángel Nava-Castañeda
Chimalpopoca #14 Colonia Obrera, México DF. CP 06800
Tel: 52 + 5554421700
Email: angellusnc@hotmail.com

It accounts for approximately 0.5% - 1% of all soft tissue sarcomas (Alkatan et al, 2010; Kim et al, 2013; Wang et al,2015; Jour et al, 2015). Most case, occur predominantly in the lower extremities, particularly the anterior portion of the thighs in adults, and in head and neck regions, especially the orbit and the tongue, in infants (Kashyap et al, 2004; Jour et al, 2015Wang et al, 2015).

We report a case of histologically proven ASPS localized in the left orbit.

Case Report

A four year old Hispanic girl was brought to consultation, because her mother noted a persistent swelling of the left upper and lower

eyelid for the last 6 months. Her past medical history and family history were noncontributory.

On examination, visual acuity was 20/40 on the right eye (OD) and 1/200 on the left eye (OS), with relative pupillary afferent defect on OS. The extraocular motility revealed a -1 abduction deficit and -3 infraduction deficit on OS. A 4 mm proptosis was noted on OS, with significant upward displacement of the globe. A mass extending approximately 3 cm below the lower eyelid margin was palpated. It was firm, not mobile, not pulsatile, and non-tender. The right eye was within normal limits. Dilated fundus exam was normal, revealing healthy optic discs on both eyes.

A computed tomography of the orbit with and without contrast showed an extraconal, hyperdense, well circumscribed mass of approximately 3.5 cm in diameter arising from the inferotemporal side of the left orbit. The mass enhanced homogenously after contrast injection. There was bone remodeling of the orbital floor, but no bone erosion or globe invasion was noted. Paranasal sinuses were free of tumor. (Figure 1)

These radiological features suggested a possible capillary hemangioma or hemangio-lymphangioma. Due to the anterior location of the tumor, an excisional biopsy by tranconjunctival approach was performed. The mass was entirely removed. Macroscopically, the excised tumor was soft in consistency and well delimitated with a brownish and homogeneous surface. When cut, the surface was yellow-brown and vessels were seen in the parenchyma.

Histologically, the lesion presented with fibrous septa and was composed of nests and clusters of large polygonal cells, separated by delicate sinusoidal vascular channels. The tumor cells had a distinct cell border and had round vesicular nuclei with prominent nucleoli and abundant eosinophilic cytoplasm. Mitotic figures were rare. The diagnostic of Orbital Alveolar Soft Part Sarcoma was made. (Figure 2)

The patient was referred to the Oncology department who did not recommend any additional treatment, but close surveillance. At her 6-month postoperative visit, the patient had healed well, with any evidence of metastases or local recurrence.

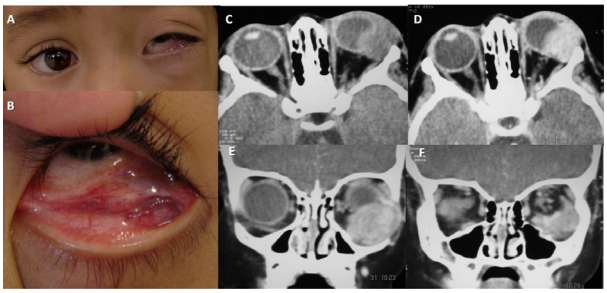

Figure 1: A: clinical picture of our patient with left eye globe displaced upwards. B: close up picture of the tumor affecting our patient. C: computed tomography revealing an extraconal mass. D: enhancement of tumor with contrast injection. E and F: coronal cuts of computed tomography showing the tumor extending far posterior and displacing the left eye globe upwards.

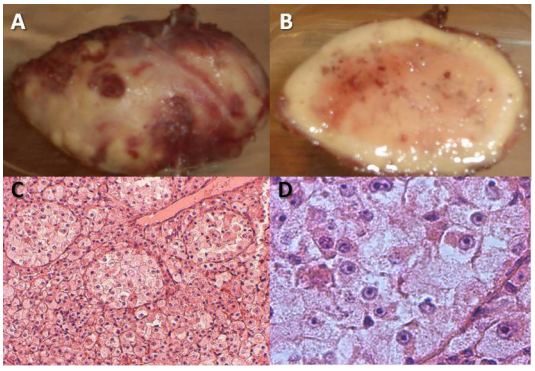

Figure 2: A: Macroscopic view of the excised tumor, showed a pink-brown capsule. B: cut surface of the tumor, yellow-brown in color with vessels within the parenchyma. C: Micrograph with medium magnification showing a neoplasia formed by groups of rounded cells with granular cytoplasm, with well defined borders, round to oval nuclei, with apparent nucleoli, without atypia. No mitosis were identified. Cells are surrounded by thin septa, giving an "alveolar" arrangement. D: Micrograph with high magnification showing groups of neoplastic cells with granular cytoplasm.

Discussion

Alveolar soft part sarcoma is clinically a rare malignancy and a tumor of uncertain histogenesis (Kashyap et al, 2004; Wang et al, 2015), it could be either neural crest or myogenic in origin (Kanhere et al, 2005). It has a strong predilection for adolescents and young women from the age of 15 to 35 (Kanhere et al, 2005; Kayton et al, 2006; Wang et al, 2015). Approximately 12-27% of ASPS involve the head and neck region, 41% of these involving the orbit (Kanhere et al, 2005; Wang et al, 2015). Orbital cases manifest as a slowly growing mass resulting in proptosis (Kim et al, 2013), conjunctival injection, globe displacement, ptosis, dilated episcleral vessels and decreased vision. It may also present with epibulbar mass or eyelid swelling (Morris et al, 2005; Alkatan et al, 2010).

Macroscopically the lesions are usually well circumscribed, encapsulated (Wang et al, 2015), red and vascularized (Alkatan et al, 2010); mostly located in the deep tissues, causing no specific symptoms for an extended period of time. Its late discovery is thought to be caused by its indolent growth (Wang et al, 2015).

Histologically the tumor is characterized by polygonal to round cells with abundant eosinophyllic cytoplasm (Kashyap et al, 2004), showing crystalline structures which are PAS-positive diastase-resistant approximately in two thirds of the cases (Alkatan et al, 2010). In the remaining cases, only PAS-positive granules, which are likely precursors of the crystals, are present (Kashyap et al, 2004). The tumor cells have pseudoalveolar, organoid arrangement (Kashyap et al, 2004; Alkatan et al, 2010) or uniform nests separated by fibrous septae (Kanhere et al, 2005; Alkatan et al, 2010), delicate vascular channels (Wang et al, 2015) or a network of reticulin fibers (Kashyap et al, 2004; Alkatan et al, 2010). Mitotic figures are uncommon (Morris et al, 2005; Alkatan et al, 2010). Large tumors may show areas of hemorrhage or necrosis and vascular invasion. There are no specific immunohistochemical markers for the tumor (Alkatan et al, 2010).

On imaging studies, ASPS may show moderate internal reflectivity on A-Scan, with minimal attenuation and good transmission (Grant et al, 1979). Computed tomography usually shows a well defined mass with smooth contour and possible bright enhancement (Morris et al, 2005).

The natural history of the ASPS is indolent, but although it is a slow growing tumor, it has propensity for distant metastases. These are common and associated with a high mortality rate (Kanhere et al, 2005). Orbital lesions may have a better prognosis than non orbital lesions on head and neck (Kanhere et al, 2005; Alkatan et al, 2010). Up to 30% of the ASPS cases present with metastasis at the initial diagnosis (Jour et al, 2015). The most common sites of metastasis are lung (42%), bones (19%), and brain (15%) (Kashyap et al, 2004; Kanhere et al, 2005). Poor prognostic factors include: increasing age, tumors larger than 5 cm and metastatic disease at initial presentation (Morris et al, 2005; Kayton et al, 2006).

Due to the rare occurrence of these tumors, the optimal management remains controversial (Kim et al, 2013). Radical surgical excision of primary or metastatic lesions with clear surgical margins seems to be essential for local control (Wang et al, 2015). Of the reported ASPS of the orbit, the mainstay for the initial management has been surgical, with either partial or total tumor resection or an orbital exenteration (Kim et al, 2013).

For cases with positive surgical margins, microscopic residual disease and palliation for metastasis, radiotherapy and chemotherapy had been added as a surgical adjunct; however, with limited benefit (Kashyap et al, 2004; Kayton et al, 2006; Wang et al, 2015). Because there is no

consensus in the efficacy of adjuvant therapy, no radiotherapy or chemotherapy was offer to our patient (Kayton et al, 2006).

The 5 year survival with adequate surgical excision is reported to be as high as 65% (Kanhere et al, 2005), but the tumor can metastasize late, with 38% of metastasis appearing 10 years after the diagnosis (Morris et al, 2005; Alkatan et al, 2010), making the close clinical long term follow up very important and necessary (Kim et al, 2013).

References

Alkatan H, Al-Shedoukhy AA, Chaudhry IA, Al-Ayoubi A, 2010. Orbital alveolar soft part sarcoma: Histopathologic report of two cases. *Saudi journal of ophthalmology,* [online].

Christopherson WM, Foote FW Jr, Stewart FW, 1952. Alveolar soft part sarcomas. Structurally characteristic tumours of uncertain histiogenesis. *Cancer,* [online]

Grant GD, Shields JA, Flanagan JC, Horowitz P,1979. The ultrasonographic and radiologic features of histologically proven case of alveolar soft-part sarcoma of the orbit. *American journal of ophthalmology,* [e-journal]; 87(6):773–7. Abstract only.

Jour G, Hoch BL, Oda D, 2015. Metastatic alveolar soft part sarcoma to the posterior mandible as initial presentation: A first report of an extremely uncommon scenario. *Journal of Oral and Maxillofacial Surgery, Medicine and Pathology* [e-journal]. 2015; 27(3):438-41.

Kanhere HA, Pai PS, Neeli SI, Kantheria R, Saoji RR, D´cruz AK, 2005. Alveolar soft part sarcoma of the head and neck. *International journal of oral and maxillofacial surgery* [e-journal]. 2005;34(3):268-72.

Kashyap S, Sen S, Sharma MC, Betharia SM, Bajaj MS, 2004. Alveolar soft-part sarcoma of the orbit: report of three cases. *Canadian Journal of Ophthalmology* [e-journal]. 2004;39(5):552-6.

Kayton ML, Meyers P, Wexler LH, Gerald WL, LaQuaglia MP, 2006. Clinical presentation, treatment, and outcome of alveolar soft part sarcoma in children, adolescents, and young adults. *Journal of pediatric surgery* [e-journal]. 2006;41(1):187-93.

Kim HJ, Wojno T, Grossniklaus HE, Shehata BM, 2013. Alveolar soft-part sarcoma of the orbit: report of 2 cases with review of the literature. *Ophthalmic plastic and reconstructive surgery* [e-journal]. 2013;29(6):138-42.

Morris WR, Padgett D, Osborn FD, Fleming JC, 2005. Pathologic quiz case: an orbital mass in a 45-year-old woman. *Archives of pathology and laboratory medicine* [online].

Wang HW, Qin XJ, Yang WJ, Xu LQ, Ji T, Zhang CP, 2015. Alveolar soft part sarcoma of the oral and maxillofacial region: clinical analysis in a series of 18 patients. *Oral surgery oral medicine oral pathology oral radiology* [e-journal]. 2015;119(4):396-401.

Source of support: nil. Conflict of interest: none