Short communication

Orbital alveolar rhabdomyosarcoma masked by ethmoid sinusitis in a 25-year-old

E. Sanz-Marcoa,*, E. Españaa,b, A. Alamarc,d, J. Pérez-Rojasd,e, M.J. López-Pratsa, M. Díaz-Llopisa,fg

a Servicio de Oftalmología, Hospital Universitario La Fe, Valencia, Spain
b Facultad de Óptica, Universidad de Valencia, Valencia, Spain
c Servicio de Otorrinolaringología, Hospital Universitario La Fe, Valencia, Spain
d Servicio de Anatomía Patológica, Hospital Universitario La Fe, Valencia, Spain
e Departamento de Anatomía Patológica, Universidad Europea de Madrid (UEM), Madrid, Spain
f Departamento de Cirugía, Facultad de Medicina, Universidad de Valencia, Valencia, Spain
g Centro de Investigación de Enfermedades Raras (CIBERER), Valencia, Spain

ABSTRACT

Case report: A 25-year-old woman with right subacute sinusitis complained about discomfort in her right eye. Clinical manifestations and computed tomography were suggestive of subperiosteal orbital ethmoid wall abscess, for which the patient underwent urgent drainage. A solid tumor was found, with a positive biopsy for alveolar rhabdomyosarcoma. Complete remission and resolution of orbital symptoms were achieved with chemotherapy and radiation therapy.

Discussion: Alveolar orbital rhabdomyosarcoma in adults is uncommon. Rhabdomyosarcoma has a high risk of spreading. It can simulate a sinusitis, as in our patient, early diagnosis and early treatment being especially important in these patients.

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Rhabdomyosarcoma alveolar orbitario enmascarado por sinusitis etmoidal en un adulto de 25 años

RESUMEN

Caso clínico: Mujer de 25 años con sinusitis subaguda derecha acudió por molestias en ojo derecho. Ante clínica y tomografía urgente sugestiva de absceso subperióstico orbitario etmoidal, se realizó un drenaje urgente, hallándose una tumoración sólida con biopsia positiva para rhabdomyosarcoma alveolar. Se inició tratamiento con quimioterapia y radioterapia con remisión completa y resolución del cuadro clínico.

Palabras clave:
Rhabdomyosarcoma
Rhabdomyosarcoma orbitario enmascarado
Rhabdomyosarcoma alveolar


* Corresponding author.
E-mail address: empar19@hotmail.com (E. Sanz-Marco).

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Rhabdomyosarcoma is a small cell malign tumor with histological characteristics similar to those of striated muscles in various embryogenic stages. It is the most frequent orbital tumor in childhood, with a prevalence of 5% of pediatric cancer and 20% of soft tissue malign tumors.1 However, it is infrequent in adults and the alveolar subtype expression is rare, particularly in subjects over 45.2

Ethmoidal sinus alveolar rhabdomyosarcoma, in a 25-year-old adult, was diagnosed during the drainage of a presumed ethmoidal subperiosteal abscess. The literature includes one case of ethmoidal sinus alveolar rhabdomyosarcoma in adults which, in contrast with the presented case, exhibited typical debut with ocular proptosis.3

Rhabdomyosarcoma can appear in paranasal sinuses and secondarily affect the orbit, presenting with banal symptomatology simulating sinusitis. Early diagnosis and treatment is particularly important in these cases.

**Clinical case**

A female, 25, with subacute sinusitis (2 months evolution) treated without improvement of symptoms visited the emergency section due to headaches and discomfort in right eye (RE). Examination revealed pain to palpation with slight tumefaction in right orbit and discreet 6 mm exophthalmos in RE (Fig. 1), visual acuity (VA) 20/20 in both eyes, isochoric and normally reactive pupils and no diplopia. Funduscopy showed RE papillary diffusion.

An urgent CT (Fig. 1) revealed the presence of subperiosteal intra-orbital abscess in the ethmoid wall. During urgent superomedial external approach through the supratarsal skin fold a solid tumor was found, the biopsy of which led to the diagnosis of alveolar orbital rhabdomyosarcoma (Fig. 2). Post-surgery MR (Fig. 3) confirmed the presence of solid tumor in the right ethmoidal walls. The findings led to defining the case as high risk in subgroup G of the European Pediatric Soft Tissue Sarcoma Study Group and subgroup III of the Rhabdomyosarcoma Study Group. Negative extension study was carried out with chest and abdomen CT and liver echography. While awaiting the extraction of ova prior to chemotherapy, the rhabdomyosarcoma increased in size compromising the orbital apex (Fig. 4), with the appearance of ophthalmoplegia, RE areactive midriasis with clear afferent pupil defect and VA reduced to perception of light. Treatment was initiated with radio- and chemotherapy according to the IVADO scheme (4 cycles of ifosfamide 3 g/m², vincristine 1.5 mg/m², actinomycin D 1.5 mg/m², doxorubicin 30 mg/m²).

The treatment achieved remission and resolution of proptosis and ophthalmoplegia, with RE papillary atrophy and amaurosis which remains unchanged to this date (51 months follow-up) (Fig. 5).

**Discussion**

There are 4 histological types of rhabdomyosarcoma: pleomorphic, embryonary, alveolar and botryoid. The distribution of subtypes varies according to series being approximately as follows: 49% embryonary, 31% alveolar, 14% pleomorphic and 6% botryoid. Other authors report different proportions in adults, i.e., 43% pleomorphic, 34% embryonary and 23% alveolar.4 The embryonary subtype exhibits a preference for infantile orbits, and is most frequent in this location.3 The case reported

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Fig. 1 – (A) Discrete right eye exophthalmos over the left one. (B–E) CT cross-section (B) and coronal (C–E): partial right side ethmoidal scene is occupation with discontinued bone extension in the lamina papyracea and presence of well defined soft tissue 27 mm x 17 mm isodense image involving orbital fat and orbital vertex.
Fig. 2 – Histopathology: (A–C) the tissue comprises dense aggregates of small cells with hyperchromatic nuclei, with granular cytoplasm with periodic acid–Schiff stain (PAS), vascularized dense fibrous stroma without fibrillary or neuropil component, without mitosis, with remains of bone trabeculae and mucosa glands, no necrosis or epidermoid, glandular or epithelial differentiation (negative AE1-3 pankeratines). Ki 67 is intensely positive with a very high proliferation index approaching 70%. Negative immunostaining for lymphoid antigens, S-100, enolase, fibrillary glyal protein and synaptophysin, which discards neuroid or neuroendocrine tumor. Positive immunohistochemistry for: (D) desmin, (E) myoglobin, muscle specific actin, myosine and (F) vimentin, corresponding to an immunophenotype typical of rhabdomyosarcoma. In conclusion, immunohistochemistry is conclusive for rhabdomyosarcoma and arrangements in dense aggregates of small cells in sub classification as alveolar rhabdomyosarcoma.

herein is an alveolar rhabdomyosarcoma with the highest incidence between 10 and 25 years of age with preference for deep soft limb tissue, being infrequent in the orbit. Accordingly, this patient is an atypical case due to adult orititary rhabdomyosarcoma as well as alveolar rhabdomyosarcoma. On the other hand and considering the previous sinusitis symptoms, onset seemed ethmoidal instead of paranasal invasion from the orbit.

CT and MR are essential techniques for diagnosing this disease because they allow to determine location, size and infiltration. CT appears as a circumscribed, homogeneous and isodense mass vis-à-vis extraocular muscles. Generally, in T1 MR the tumor is isointense vis-à-vis extraocular muscles and hypointense vis-à-vis ocular fat. In T2 MR it can be hypo-, iso- or hyperintense vis-à-vis orbital structures. The CT of this case exhibited a half-moon shaped mass with partial occupation of the ethmoidal sinus and orbitary vertex, without signs of surrounding infiltration or information. The image is compatible with orbital phlegmon and rhabdomyosarcoma despite not exhibiting in CT the typical hypodensity of an abscess. However, previous sinusitis inclines the urgent diagnosis toward words subperiestale phlegmon. The urgent nature of the diagnosis excluded MR which would have discarded the phlegmon diagnosis and confirmed its solid mass (Fig. 3).

Rhabdomyosarcoma is a malign tumor with a tendency to local, lymphatic and hematogenous invasion. In addition, due

Fig. 3 – Magnetic resonance: right half-moon intraorbit tumor (35 mm × 28 mm × 18 mm) involving ethmoidal cells with implantation on the ethmoidal wall, displacing to optic nerve and internal rectus muscle.
Fig. 4 – Increased exophthalmos, with right eye proptosis, exotropia, a reactive midriasis and vascular congestion.

Fig. 5 – Post-surgery, 51 months without modification of post-surgery alterations: (A) right eye exophthalmos disappearance. (B–D) fibrosis image in control MR: (B) hyperintense image in STIR without gadolinium capture, (C) hypointense in T1 and (D) hyperintense in T2.

to its undifferentiated nature, the alveolar subtype of this case exhibits high risk of early dissemination. The most frequent clinic of orbital rhabdomyosarcoma is rapidly evolving proptosis, diagnosed at an early stage as a space-occupying lesion of probable tumor origin. For this reason early detection of paranasal rhabdomyosarcoma is important, as onset can be insidious and lost within a banal condition as in the case reported herein. In these cases, early diagnosis and treatment were crucial for the vital prognosis.

Conflict of interests

No conflict of interests has been declared by the authors.

REFERENCES