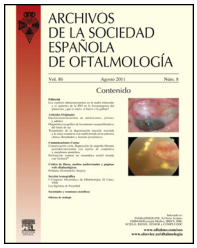
ARCHIVOS DE LA SOCIEDAD ESPAÑOLA DE OFTALMOLOGÍA

www.elsevier.es/oftalmologia



Short communication

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

E. Sanz-Marco[a,*], *E. España*[a,b], *A. Alamar*[c,d], *J. Pérez-Rojas*[d,e], *M.J. López-Prats*[a], *M. Díaz-Llopis*[a,f,g]

[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*

ARTICLE INFO

Article history:
Received 12 December 2010
Accepted 30 October 2012
Available online 2 August 2014

Keywords:
Rhabdomyosarcoma
Masked orbital rhabdomyosarcoma
Rhabdomyosarcoma alveolar
Apex
Sinusitis

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.

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

Palabras clave:
Rabdomiosarcoma
Rabdomiosarcoma orbitario enmascarado
Rabdomiosarcoma alveolar

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 rabdomiosarcoma alveolar. Se inició tratamiento con quimioterapia y radioterapia con remisión completa y resolución del cuadro clínico.

☆ Please cite this article as: Sanz-Marco E, España E, Alamar A, Pérez-Rojas J, López-Prats MJ, Díaz-Llopis M. Rabdomiosarcoma alveolar orbitario enmascarado por sinusitis etmoidal en un adulto de 25 años. Arch Soc Esp Oftalmol. 2014;89:182–185.

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

2173-5794/$ – see front matter

Ápex
Sinusitis

Discusión: El rabdomiosarcoma orbitario alveolar en adultos es infrecuente. Presenta un alto riesgo de diseminación y puede debutar simulando una sinusitis, como en este caso, por lo que un diagnóstico y tratamiento precoces son de especial importancia en estos pacientes.

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 orbitary 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-orbitary 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 orbitary 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 orbitary 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/m^2, vincristine 1.5 mg/m^2, actinomycin D 1.5 mg/m^2, doxorubicin 30 mg/m^2).

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: pleomorphism, 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.[1] The case reported

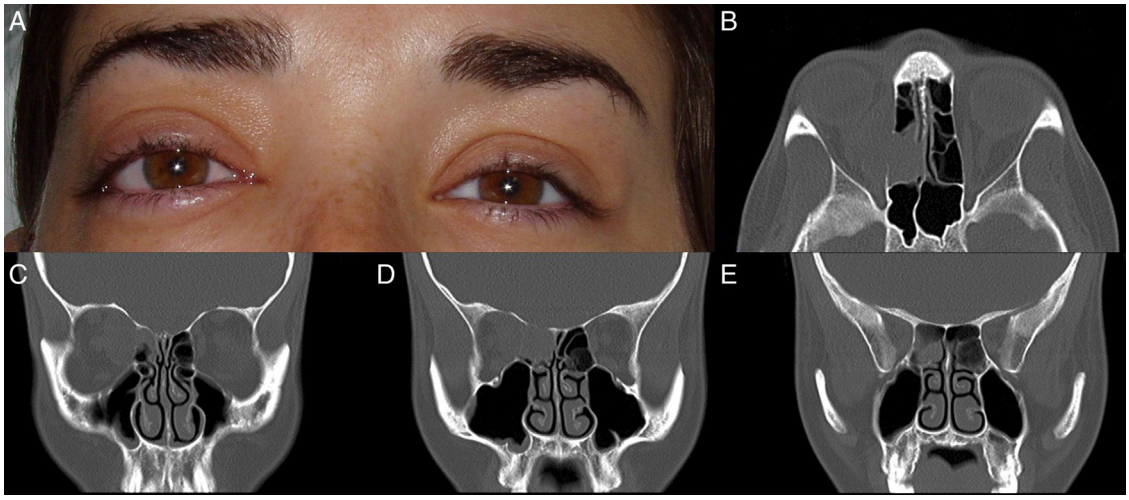


Fig. 1 – (A) Discrete right eye exophthalmos over the left one. (B–E) CT cross-section (B) and coronal (C–E): partial right side ethmoidal scene is occupation with discontinued bone extension in the lamina papyracea and presence of well defined soft tissue 27 mm × 17 mm isodense image involving orbitary fat and orbital vertex.