Orbital Rhabdomyosarcoma with a good life prognosis after surgical treatment in a 14-year-old patient

Case Report

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Abstract: The orbital rhabdomyosarcoma is one of the most frequent malignant orbital tumours in children. At this age, the common histological types are the embryonal and alveolar type. The onset is mainly under the age of 16. Without a recent and correct treatment it can give metastasis in lung and bone. The hereby paper presents one clinical case of a teenager presented at the ophthalmological consultation for a small tumor located in the superomedial part of the orbit. Computed tomography (CT) and magnetic resonance imaging (MRI) supported the diagnosis revealing the location and extension of the tumor. During the surgery, we discovered two small tumors and the histological examination revealed an embryonal type of orbital rhabdomyosarcoma. After the surgery, the patient followed an oncological treatment consisting of chemotherapy and local radiotherapy. The prognosis for life was favorable, linked with the recent diagnosis and treatment, the histological type and the good response at the oncological treatment.

Keywords: Orbital rhabdomyosarcoma • Local radiotherapy • Chemotherapy

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1. Introduction

Orbital rhabdomyosarcoma is the most common primary orbital malignant tumor in children and the most common soft tissue malignancy of childhood [1], with 10% of all cases occurring in the orbit [2]. There are three histological types: embryonal (the most common type in children), alveolar (also in children) and pleomorphic (in adults). Presentation is in the first decade of life, 90% of patients presenting before age 16 [3]. Diagnosis is based on clinical signs. The tumor may involve any part of the orbit; the common location is retrobulbar, followed by superior and inferior one. Most often the first sign is the presence of the tumor with a rapidly progressive proptosis. In other cases, a palpable mass and ptosis are present in about one third of cases. Swelling and injection of overlying skin develop later, but the skin is not hot. For the positive diagnosis, we need orbital ultrasound examination, cranial tomography and magnetic resonance imaging (MRI) in order to establish
the presence, location, extension of the tumor and for differential diagnosis [4]. Biopsy should be performed if rhabdomyosarcoma is suspected based on clinical and radiological exams [2].

Also, for evidence of metastatic spread we need chest X-ray, bone biopsy, liver function tests, lumbar puncture and skeletal survey. The most common sites for metastasis are lung and bone [2]. The treatment consists of surgery, associated with local radiotherapy and chemotherapy [5]. The prognosis for life is dependent on the stage and location of the tumor, and the moment of the diagnosis and treatment.

2. Patient and methods

We report the case of a 14 years old girl presented for a small tumor in the superior and internal angle of the left orbit. The onset of the illness was 3 months before the presentation, because of the appearance of a small tumor localized in the superior and internal part of the orbit, which grow in dimension in time.

From the personal pathological data we retain a childhood infectious diseases.

The heredocolateral pathological data revealed healthy parents.

The ocular examination at both eyes was normal regarding the anterior and posterior segment of the eye. Ocular refraction revealed a small hyperopia at both eyes. Examination of the right orbit didn’t show any pathological aspect, but at the left orbit we visualized and palpated a small tumor, in the superior and internal part of the orbit, painless and smooth. Ocular motility was normal in both eyes.

Exophthalmometry measured 12mm at both orbits. She had a normal fundus at both eyes.

Left orbital CT scan showed no bone destruction, but revealed a small space occupying lesion in the superointernal part of the orbit, with homogenous density, without bone destruction.

Blood investigations showed a moderate anemia (haematocrit=32.9%, Hb=10.8 g/dl) with a high Erythrocyte sedimentation rate (ESR) (60mm/h) and lymphocytes=16.1%.

The pediatric and Ear, Nose and Throat examination (ETN) was normal, including the clinical examination of the patient.

After the clinical ophtalmological examination, the paraclinical and laboratory examinations the positive diagnosis was: Left eye – Superior and internal orbital tumor, Both eyes – Small hyperopia.

At this moment it was necessary to make the differential diagnosis between the following tumors, which appears in childhood and with an internal and superior location of the orbit: deep orbital dermoid cyst (nonaxial proptosis, CT scan reveals heterogeneous well circumscribed lesion); anterior encephalocele (fronto-ethmoidal); anterior orbit capillary haemangioma (dark blue or purple through the overlying skin and displaces the glob); anterior orbital lymphangioma (soft bluish mass in the upper nasal quadrant and cystic conjunctival component) [6]; rhabdomyosarcoma (located retrobulbar superior and inferior, CT scan reveals poorly defined mass of homogeneous density; adjacent bony destruction); choroma (a form of acute myeloid leukae mia); neuroblastoma (malignant tumor, originate from primitive neuroblasts in the abdomen).

The final diagnosis may be put after the extirpation of the tumor, accomplished with the histological exam.

Evolution of this case depends on the histological type of the tumor.

The treatment began with the surgery of the orbit which had as a goal the total extirpation of the tumor. The steps of the treatment were: cutaneous incision of 10mm, at 4mm from the orbital margin, the evidence of the tumor which was grey, cartilaginous consistency and we discovered another tumor behind the first one, tumors which were histopathologically examined.

The macroscopic examination revealed two tumors, grey, cartilaginous consistency, about 3/2 cm each.

The microscopic examination of both tumors showed: dense areas with elongated cells with a centrally located, hyperchromatic nucleus surrounded by a considerable amount of eosinophilic cytoplasm, with perivascular location, alternative with paucicellular areas with small cells with hyperchromic nuclei (Hematoxilin-Eosine stain) The tumor cells differentiate a long rhabdomyoblastic line forming elongated spindle cell types (“strap cells”) (Figure 1).

Figure 1. Histopathological exam (HE stain, ob.10x); the electron microscopic appearance of the embryonal type of rhabdomyosarcoma.