CASE REPORT

Paranasal sinus rhabdomyosarcoma: A rare tumor of poor prognosis

A. El Sanharawi, B. Coulibaly, J.-P. Bessede, K. Aubry

Introduction

Rhabdomyosarcoma (RMS) is a tumor of the conjunctive tissue, found more frequently and with better prognosis in children than adults. ENT forms are not unusual, but paranasal sinus locations are exceptional. We report a case of adult evolved ethmoid RMS, and discuss the characteristics, diagnosis and treatment of such tumors.

Case report

A 48-year-old man was referred to ENT for recurrent epistaxis. Brain and facial sinus CT found 30 × 25 mm tissular filling of the left anterior and medial ethmoid cells, with ethmoid and papyraceous lamina bone lysis without intra-orbital extension. Biological assessment found anemia and thrombopenia. A myelogram was therefore taken, which indicated massive metastatic medullary invasion. Biopsies, under general anesthesia, proved highly hemorrhagic. Subsequent recurrent epistaxis required platelet transfusion every two days; very rapid onset of left exophthalmia was associated with ptosis, superior palpebral edema and ophthalmoplegia. Contrast-enhanced cervico-thoraco-abdomino-pelvic CT showed no secondary pulmonary or hepatic locations. MRI found tumoral extension toward the frontal sinus, frontal cortex and left intra-orbital lysis.
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region (Fig. 1). 18-fluoro-desoxy-glucose positron emission tomography (18FDG-PET) found hypermetabolic cervical lymph-node sites and diffuse osteo-medullary infiltration (Fig. 2). Definitive anatomopathologic examination diagnosed grade-3 alveolar RMS on the Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) classification. Immunohistochemistry was positive for anti-desmin, anti-NCAM (neural cell adhesion molecule) and anti-myogenin antibodies, and negative for anti-caldesmon antibodies (Fig. 3). Fluorescence in situ hybridization (FISH) analysis found FKHR gene rearrangement. A multidisciplinary team meeting decided, given the tumoral extension, on initial adriamycin-based chemotherapy associated to regular transfusion and neutrophil growth factor injection. After initial improvement in general health status and disappearance of the epistaxis, there was rapid deterioration of consciousness, due to onset of left frontal intraparenchymatous cerebral hematoma; the patient died 5 weeks after initiation of chemotherapy.

Discussion

RMS is a rare malignant tumor, originating in the conjunctive and derivative tissue. It is especially rare in adults, whereas it is the most frequent form of soft-tissue tumor

Figure 1  Brain MRI (horizontal T1-weighted gadolinium-enhanced slices). A. Left ethmoid tumoral syndrome with left intra-orbital extension, exerting a mass effect on the right medial rectus muscle. B. Left frontal lobe tumoral invasion.

Figure 2  18FDG-PET. A. Horizontal, sagittal and frontal slices in CT paired to scintigraphy, showing infiltration via the skull-base lesion with bone lysis areas. B. Scintigraphic images, showing diffuse osteo-medullary infiltration. Left: corrected attenuation MIP; right: non-corrected attenuation MIP.
in children [1]. Histologically, three types are distinguished: embryonic, alveolar and pleomorphic. The embryonic type is the one most frequently encountered in ENT, mainly affecting children under 5 years of age; it is found in 25% of nasopharynx and sinonasal cavity RMS locations, and is the form associated with the best prognosis. The alveolar type mainly affects children over 5 years of age, adolescents and young adults; it may be located in the sinonasal cavities, and is of poorer prognosis. The pleomorphic form is rarer, mainly affects adults and is of poor prognosis [1].

RMS mainly develops in the thorax or extremities in adults, and in the ENT region or uro-genital tract in children [2]. Only 10–15% of adult ENT locations are in the paranasal sinuses [3,4].

At the outset of management, RMS does not present any specific clinical signs (except for the embryonic form, which may have the aspect of a gelatinous nasal polyp) [1,2]. Sinus involvement presents as early epistaxis associated with exophthalmia and ophthalmoplegia. Imaging does not determine diagnosis, but does assess locoregional extension. On MRI, the tumor shows a heterogeneous signal, equal to or more intense than the muscle [5]. Only histology with immunohistochemistry provides specific diagnosis, in the form of diffuse desmin and myogenin staining. Genetically, t(2;13)(q35;q14) or t(1;13)(q36;q14) translocation is specific, and is found in all alveolar forms, indicating rearrangement of the FKHR gene into either PAX3-FKHR or PAX7-FKHR gene [1].

RMS shows very rapid spontaneous evolution; diagnosis is very often at an advanced stage [3]. Prognosis is poor, with frequent lymph-node, lung or bone metastasis [1,6]. Extension assessment should include myelogram, brain and sinus MRI, contrast-enhanced cervico-thoraco-abdominal CT and 18FDG-PET.

Prognosis is poorer in adults than children, and ENT locations are usually difficult to resect due to extension. Genetic analysis provides complementary prognostic information, t(1;13)(q36;q14) translocation being associated with a poorer prognosis than t(2;13)(q35;q14) translocation in alveolar forms [1].

Treatment ideally associates surgery and radio-chemotherapy. Surgery should be the first-line attitude in operable ENT forms. Adjuvant radiation therapy may, depending on team experience, help optimize control [7]. Chemotherapy classically associates vincristine-dactinomycin-cyclophosphamide, with adriamycin monotherapy reserved for advanced forms [8]. Non-operable forms should be managed by neo-adjuvant chemotherapy then, depending on regression and general health status, surgery followed by radio-chemotherapy. Whatever the treatment, prognosis in adults is poor [6].

**Conclusion**

Paranasal sinus RMS is a rare tumor of poor prognosis in adults. Locoregional extension is frequently advanced at
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Diagnosis, limiting treatment options and worsening prognosis. Surgery followed by radio-chemotherapy is the attitude of choice in locally advanced and metastatic forms.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References


