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About A Case Of Rhabdomyosarcoma Of The Ethmoidal Sinus: Rare Tumor With Guarded Prognosis

S.Hassina, Z.hazil L,Krichenne MA,Hasnaoui, Robbana L,tebbay,ELmajdoubi K,bekkar B,Jeribi A , Y.Akannour, L.Seghini, E.Abdallah.

Department of Ophthalmology B, Rabat Specialty Hospital, CHU ibn Sina, Mohammed V Souissi University Rabat

Abstract:

ethmoid sinus rhabdomyosarcoma is a rare tumor in adults, with a poor prognosis, its diagnosis is suspected by clinical and imaging, but confirmation remains histological, the therapeutic indication is based on the assessment of tumor extension.

we report the case of a young patient presenting with rhabdomyosarcoma of the ethmoidal sinus

Keywords: adult, ethmoidal sinus, metastases, rhabdomyosarcoma

Introduction:

Rhabdomyosarcoma (RMS) is a malignant tumor of mesenchymal origin with more or less marked striated muscle differentiation, of unknown etiology [1]. It is a tumor of infants and young children, and is rare in adults. It is a highly malignant tumor, distinguished from other sarcomas by its locoregional aggressiveness, metastatic course and poor prognosis regardless of treatment. Diagnosis requires histological examination. Treatment is multidisciplinary, combining surgery, radiotherapy and chemotherapy.

Observation:

we report the case of a 23-year-old boy, with no pathological antecedents, who presented with nasal obstruction with purulent rhinorrhea associated with pain in the left hemiface. clinical examination revealed subtotal ptosis of the left eye associated with non-axial, non-reducible, non-pulsatile exophthalmos pushing the eyeball backwards and forwards (figure 1), with limited ocular movement.

a complementary craniofacial CT scan showed a tissue lesion centred on the ethmoid with involvement straddling the midline at the level of the ethmoidal cells, accompanied by lysis of the ethmoidal roof, with the presence of adenopathy at the level of chain 2B, initially suggesting a tumoral origin (figure 2), which was confirmed by histological evidence after biopsy, in favour of a rhabdomyosarcoma of the ethmoidal sinus.

the patient benefited from multidisciplinary surgical and oncological care

Discussion:

Malignant tumors of the paranasal sinuses represent <1% of all malignant tumors, and paranasal sinus sarcomas account for around 7% of all head and neck sarcomas. (1), the maxillary sinus was the most common primary site (50%) of RMS among paranasal sinuses, followed by the ethmoidal sinus (35%).

Although the 5-year overall survival (OS) rate for localized MRS exceeds 70% in children (<18 years), the prognosis for localized MRS in adults is extremely poor. (2, 3, 4), Other important prognostic factors include tumor size, presence of metastases and histological subtypes. Patients with metastatic disease have a poorer prognosis and should be considered for a variety of therapeutic approaches, underlining the need for accurate staging. Initial comprehensive staging uses cross-sectional imaging of the primary tumor, with a general extension workup depending on orientation.

Management of RMS patients involves surgery and/or radiotherapy for local control and chemotherapy of varying intensity and duration, depending on the risk group assigned.

Conclusion :

Le SRM chez l'adulte est extrêmement rare et son pronostic est sombre, en particulier chez les patients présentant des métastases à distance.

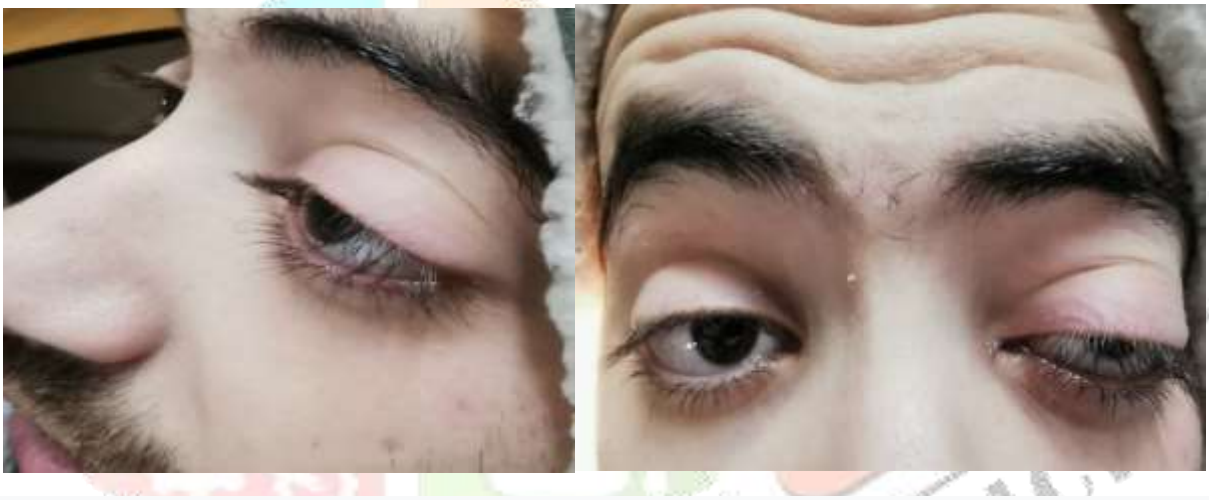

Figure1: photos in front and profile view showing ptosis and exophthalmos of the left eye

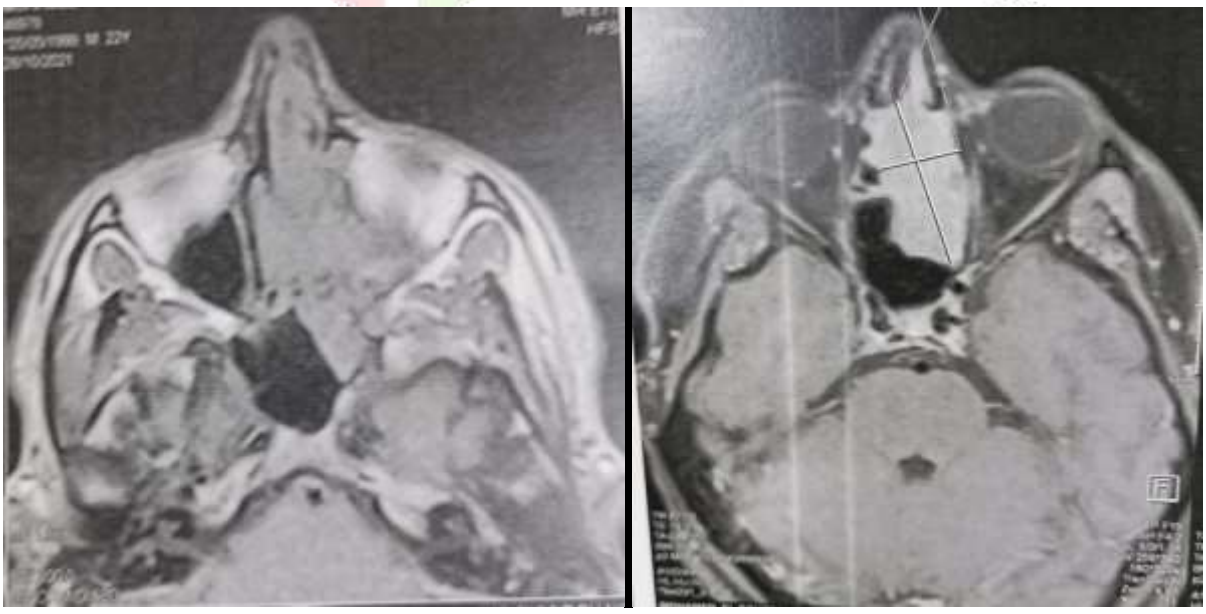

Figure2: transverse scan section showing the lesional process which depends on the ethmoidal sinus

Conflicts of interest:

All authors declare that they have no conflicts of interest.

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