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Clinical Image

14 Years Old Child, without Antecedent Who Presents Since 2 Months, a Right Maxillary Formation Having Increased in Volume with Fatigue, Bone Pain and Diplopia

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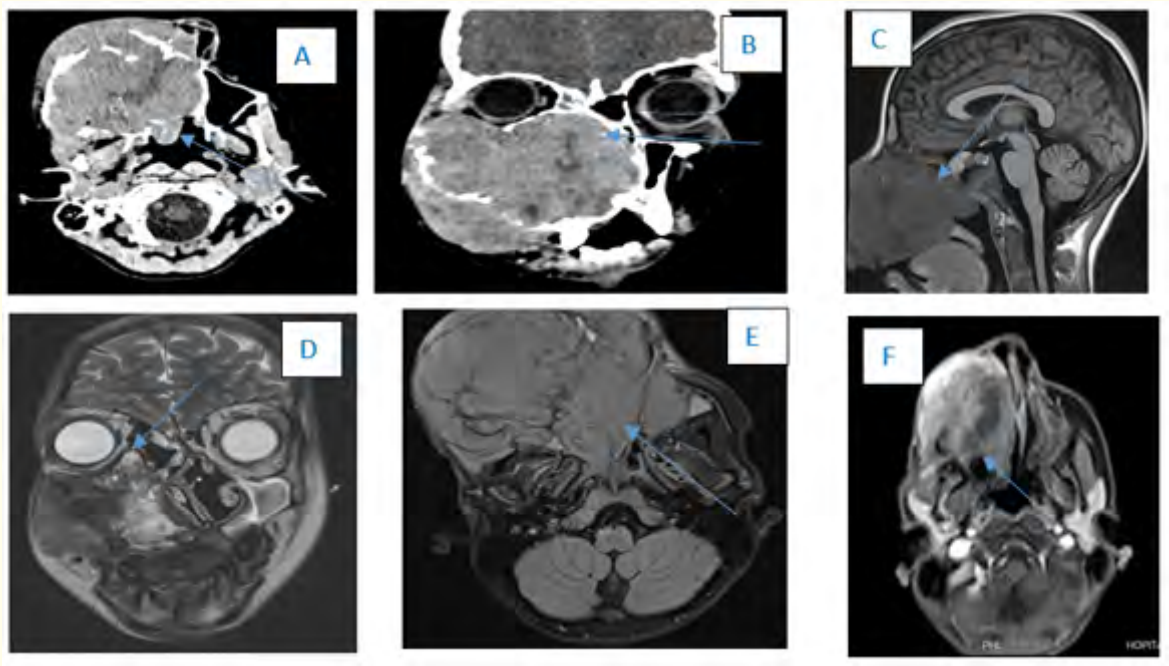

Figure: *Cerebral CT scan without and with injection (A-B) with cerebral MRI in sagittal T1 ©, coronal T2 (D), axial FLAIR (E)and T1 injected section (F) showing a tissue process of the right maxillary sinus in hyposignal T1 and intermediary signal T2, heterogeneously enhanced infiltrating the homolateral zygomatic bone, infiltrating the orbital floor with exophthalmos, infiltrating the choanae and bulging into the lumen of the rhinopharynx and nasal cavities and the homolateral ethmoidal cell, thus infiltrating the maxillary bone and extending to the homo and contralateral dental arch with lysis of the roots of the upper teeth bilaterally, without calcification or hemorrhagic areas.*

Background

- Rhabdomyosarcoma in the maxillary sinus is a malignant tumor proliferation originating from undifferentiated mesenchymal cells capable of producing striated muscle cells.
- It is a rare tumor affecting mostly children (70% of cases involve children under 12 years of age).
- The main localizations are cephalic, genitourinary and skeletal, especially the extremities.
- Three histological forms predominate:
  - Alveolar RMS with a poor prognosis.
  - Pleomorphic RMS with a poor prognosis.
  - Embryonal RMS with an intermediate prognosis, representing about 80% of these tumors, and botryoid RMS which constitute a particular form that is not very frequent but has a better prognosis.
- Some RMS are associated with a genetic predisposition to cancer (neurofibromatosis type 1, Li- Fraumeni syndrome).
- The radiological aspects are most often non-specific.

Clinical Presentation

- Symptoms vary according to extent of involvement.
- Common symptoms include facial mass, bone pain, weakness and fatigue, nasal obstruction, epistaxis and progressive respiratory failure with oropharyngeal expansion.
- Additionally, orbital involvement present: exophthalmos, retroorbital pain, extraocular muscle palsies, and progressive vision loss.
- RMS involvement often affects the hypothalamic-pituitary axis (HPA) and may present with diabetes insipidus.

Key diagnostic features:

- On CT, it presents as a voluminous tissue mass of similar density to that of the muscle occupying the sinus cavity.
- The starting point 'sinus' has a great orienting value.
- Extension to the other naso-sinus cavities and to the neighboring regions, notably the base of the skull and the orbit.
- Heterogeneous enhancement.
- Bone lysis, which is the most frequent sign and the most suggestive of malignancy of the lesion.
- However, a reactive bone condensation can be seen in some cases as well as intra-tumoral calcifications.
- On MRI, the lesion is in iso or discrete T1 hypersignal in relation to the muscles and in T2 hypersignal, enhancing after injection of gadolinium. A cluster of contrast called "botryoid sign" is strongly suggestive of the diagnosis.

Differential diagnoses:

- **Epidermoid carcinoma**: 50 to 60% of nasosinus tumors. The tumor presents as a large, poorly defined mass in T1 isosignal and T2 isosignal, often heterogeneous, with moderate enhancement after PDC injection. The CT appreciates the bone lyses (the tumor is often aggressive) and MRI better specifies the loco-regional extension essentially in the nerve or vascular sheaths towards the endo-cranium through the canal structures of the base.

- **Esthesioneuroblastoma**: 6% of nasal cavity tumors. Imaging shows a slowly erosive tumor that is homogeneous in small lesions and heterogeneous in large lesions with areas of necrosis. On CT, the lesion is isodense and clearly enhances after injection of PDC. Calcifications are frequent. On MRI, the lesion appears as a gadolinium-enhanced mass in T1 isosignal and T2 iso or hypersignal.
- **Non-hodgkin lymphone:** 8% of NSCTs. On imaging, NHL is characterized by a mass that differs from squamous cell carcinoma in that it is moderately involved in the bone, contrasting with a large tumor formation that fills the sinus of origin and extends into the surrounding spaces. On MRI, the lesion appears as a T1 isosignal with an iso or discrete T2 hypersignal and is moderately contrast-enhanced.
- **Malignant melanoma:** 0.5 to 2% of NST, on CT, melanoma often appears as a mass usually located in the nasal cavities, enhanced after injection of PDC and accompanied by lysis of adjacent bony structures. On MRI, melanotic melanoma is T1 hypersignal, related to the paramagnetic properties of melanin, and T2 hyposignal. On the other hand, amelanotic melanoma presents as T1 iso or hyposignal and T2 hyper or isosignal.
- **Chondrosarcoma:** 1% of TNS, is seen in CT as a lytic mass that erodes and destroys the surrounding bone walls, these limits are irregular and are not accompanied by a border of osteocondensation. MRI better assesses the tumor extension, the lesion appears in T1 iso signal and T2 iso or hypersignal, the tumor is relatively avascular, it enhances only slightly and late after injection of PDC.
- **Ameloblastic carcinoma:** Rare, it is seen on imaging as an aggressive, poorly defined tissue mass with heterogeneous enhancement after injection of PDC, associated with lysis of the opposite cortices and extension to the adjacent soft tissues.

Treatment:

- The management of RMS is multidisciplinary, including multidrug therapy, surgery and external radiotherapy.
- Despite improvements in therapeutic management, the prognosis of this tumor remains poor, especially for adults. The 5-year survival rate is 22% for adults compared to 55% for children [1-6].

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