

## UNUSUAL INITIAL DEVELOPMENT OF AN ESTHESIONEUROBLASTOMA ABOVE THE CRIBRIFORM PLATE

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Esthesioneuroblastoma is a rare tumor localized in the superior part of the nasal cavity. Extension in the anterior cerebral fossa is sometimes observed. As we discovered in our patient, the location of the tumor above the cribriform plate was very unusual and meningioma was suspected. Features and prognosis are discussed below.

**Key-word:** Nose, neoplasms.

### Case report

A 59-year-old man was admitted in the emergency room after convulsions and loss of consciousness. Computed tomography revealed a hypodense basi-frontal lesion with a barely perceptible erosion of the cribriform plate (Fig. 1). No calcifications were noted. Complementary preoperative investigation by MRI showed a hyperintense T2-weighted solid mass above the cribriform plate, surrounded by edema. Administration of gadolinium indicated a moderate to intense enhancement and a small bud tissue developed through the cribriform plate (Fig. 2). The diagnosis of meningioma was then made. The patient underwent a surgical resection by supra-orbital approach. There were no post-operative complications. Histological examination demonstrated typical features of esthesioneuroblastoma. Treatment was supplemented by radiotherapy. Unfortunately, a local recurrence was detected 1 year later (Fig. 3). A new surgical resection was therefore undertaken. No evidence of local recurrence or metastases has been identified after 2 years of observation.

### Discussion

Esthesioneuroblastoma (ENB) or olfactory neuroblastoma was initially described by L. Berger in 1924 (1). This lesion represents 3% of all nasal tumors and occurs between ages 3 and 79, with a peak at age 10 and age 40 and occurring predominantly in males (1-2). The typical localization is under the cribriform plate. In fact, the lesion arises from sensory cells in the olfactory mucosa which is

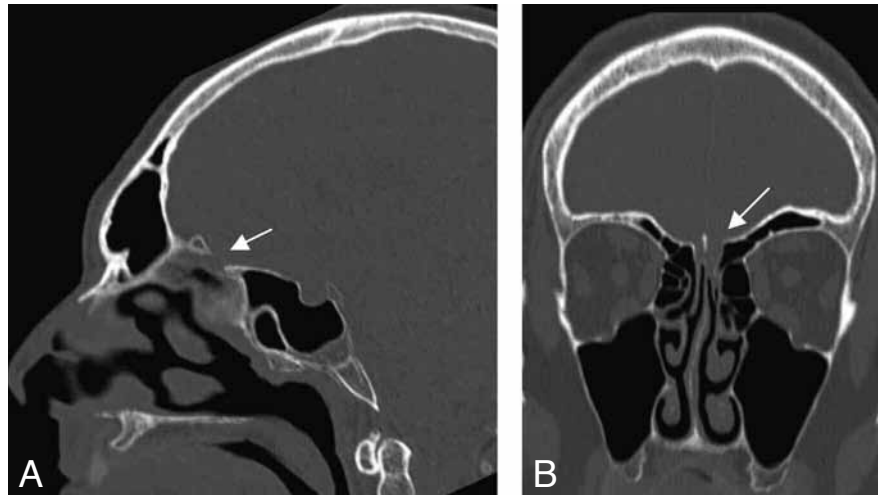


Fig. 1. — Sagittal (A) and coronal (B) CT slices showing a slight erosion of the cribriform plate and the integrity of the superior nasal cavity (white arrow).

located in the most superior part of the nasal cavity. These cells originate in the neural crest and differentiate into olfactory elements. Many articles in the literature present ENB as a "site specific" tumor developing first in the superior nasal cavity, involving the nasal septum, turbinates, ethmoid. Maxillary sinuses, orbital cavities, and anterior cerebral fossa are occasionally affected. In our patient, the location of the ENB was very unusual, causing initial diagnostic confusion.

By CT or MRI, the choice of sagittal and coronal planes is important to estimate the development of ENB into the superior nasal cavity and/or the anterior cerebral fossa. CT may show calcifications in the lesion. These are not constant nor are they specific. Bone erosions are also described, including the cribriform

plate. These erosions are not always easily visible. Enhancement is usually moderate after CA perfusion. Cystic component is sometimes reported. This may help to differentiate ENB and meningioma when the infringement is located above the cribriform plate. "Dural tail" is difficult to distinguish of meningeal infiltration. Finally, it is well known that meningiomas can cause hyperostosis by osteoblastic reaction. Bone erosion is rare with meningiomas, except in very aggressive cases.

Differential diagnosis must include other regional tumors like epidermoid carcinoma, adenocarcinoma, lymphoma, metastasis of melanoma or hemangiopericytoma. The final diagnosis is histological in most cases.

Hyams has proposed to differentiate low and high-grade ENB. He has shown in a retrospective series of 49 patients that the 5-year survival is influenced by the grade: 80% for the low grade and 40% for the high (3). However, this distinction is often difficult to appreciate, as reported in the literature (4-5). In 1992, Dulguerov

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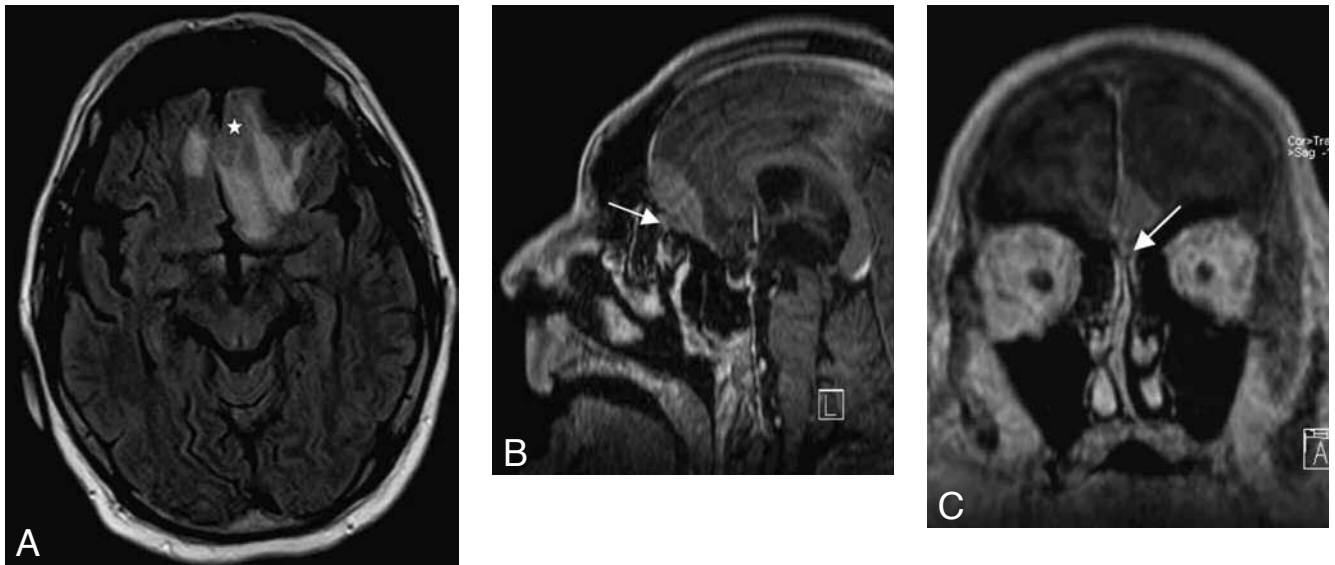


Fig. 2. — Axial FLAIR (A) and T1-weighted MR acquisition after gadolinium administration, with sagittal (B) and coronal (C) reconstructions, showing the ENB (asterisk) above the cribriform plate and surrounding edema. Enhancement is moderate to intense. A small extension along the nasal septum is present (white arrow).

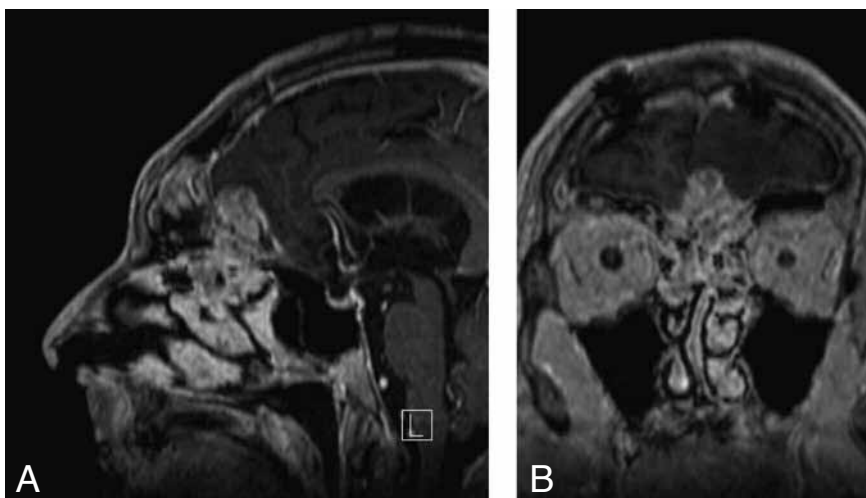


Fig. 3. — MR examination realized 1 year later, with sagittal (A) and coronal (B) T1-weighted slices, demonstrating a local recurrence of ENB.

and Calcaterra (6) proposed a TNM classification based on CT and MR imaging findings (Table I). This classification differs from that usually used for carcinomas of the nasal cavity and paranasal sinuses by the International Union Against Cancer (7). This is due to the great importance given to the extension of the tumor above the cribriform plate which involves difficulties in processing.

In most studies, the long term survival varies between 50% and 80% (8). The prognosis is usually determined by the initial stage. Surgical total resection is sometimes impossible. Local recurrences are frequent and mostly intra-cranial, by meningeal or parenchymal extension. The largest study which was

Table I. — TNM classification of Dulguerov and Calcaterra.

| Classification | Definition   |
|----------------|--|
| T1             | Tumor involving the nasal cavity and/or paranasal sinuses (excluding the sphenoid), sparing the most superior ethmoidal cells                |
| T2             | Tumor involving the nasal cavity and/or paranasal sinuses (including the sphenoid), with extension to – or erosion of – the cribriform plate |
| T3             | Tumor extending into the orbit or protruding into the anterior cranial fossa, with / without dural invasion                                  |
| T4             | Tumor involving the brain  |
| N0             | No cervical lymph node metastasis  |
| N1             | Any form of cervical lymph node metastasis   |
| M0             | No metastasis  |
| M1             | Distant metastasis   |

published by Duguerov and Allal shows a local rate of 29% (9). Distant recurrences are lung, liver, bone, but also meningeal or cerebro-spinal in 10%-25% of cases according to authors. The primary treatment for ENB is surgery followed by radiotherapy. A collaborative team of experienced medical and surgical specialists including neurosurgeons and otorhinolaryngologists is crucial to define the best treatment for each patient. To date, there is no evidence as to the effectiveness of chemotherapy (10,11).

In conclusion, an extra-axial tumor centered on the cribriform plate should include the diagnosis of ENB although this lesion is less frequent than meningioma, and knowing that most of ENB have an intranasal development. The diagnosis is often difficult and is histological in many cases.

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