HISTIOCYTOSE LANGERHANSIENNE
Atteinte encéphalique, du massif facial et de la base du crâne

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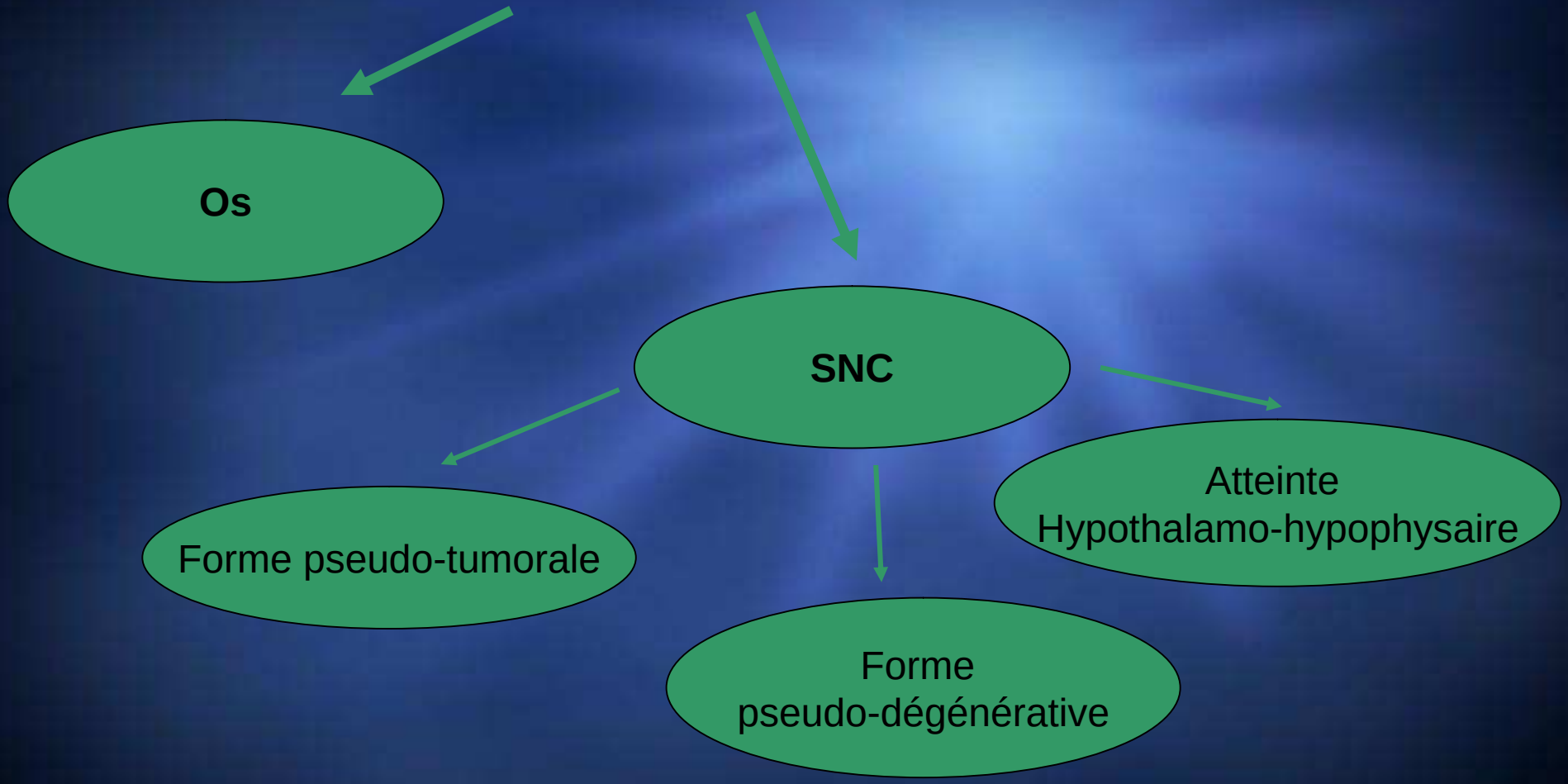
Histiocytose LANGERHANSIENNE

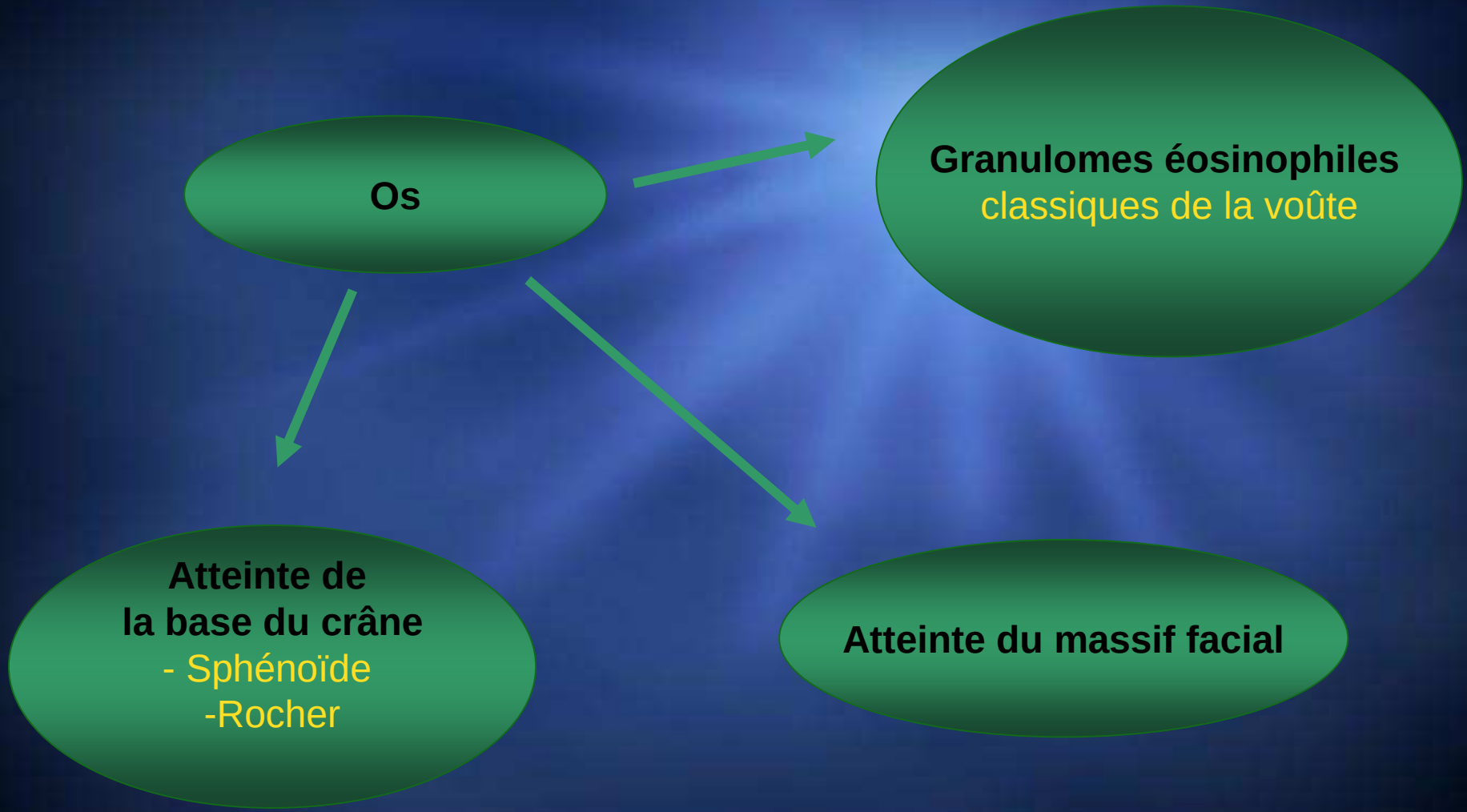
- Anciennement HX
  - Granulome éosinophile
  - M. Hans-Schüller-Christian
  - M. Abt-Letterer-Siwe
  - M.d'Hashimoto-Pritzker

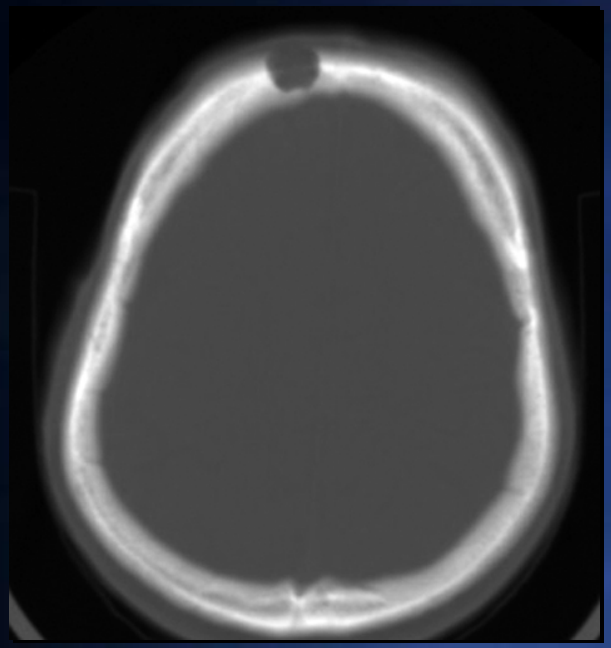
-Variabilité clinique
  - Lésion isolée
  → multi systémique sévère

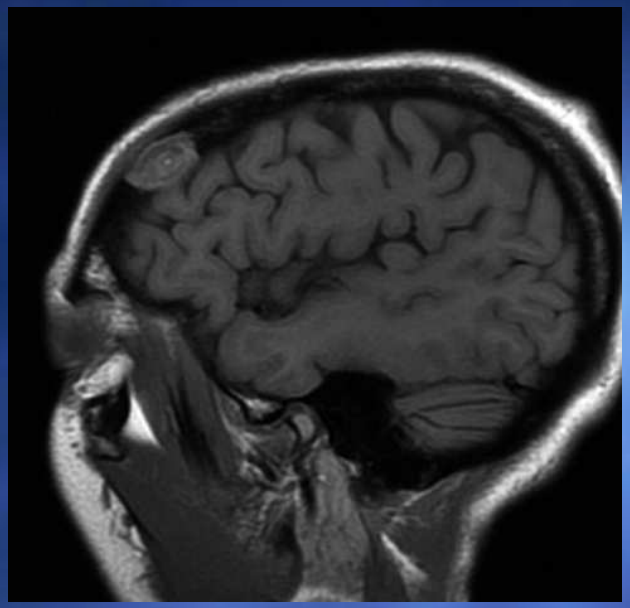
Histologie

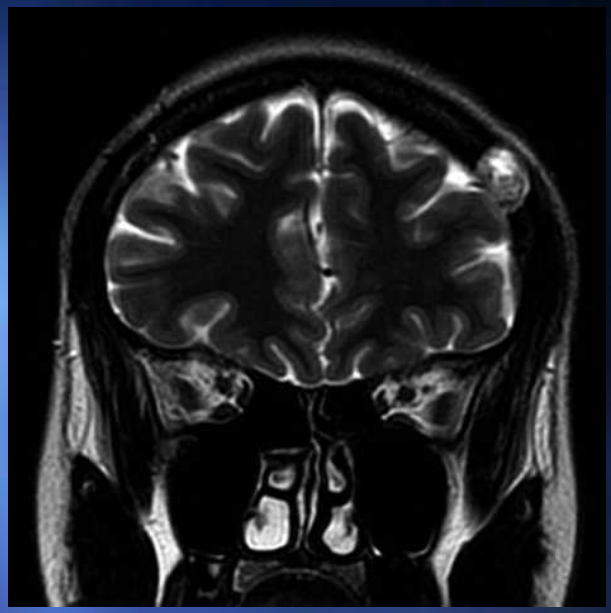
- Prolifération de nature clonale,
- d'histiocytes de type Langerhans
- S100+, CD1a+ et langerine+
- Granules de Birbeck (μ . électronique)

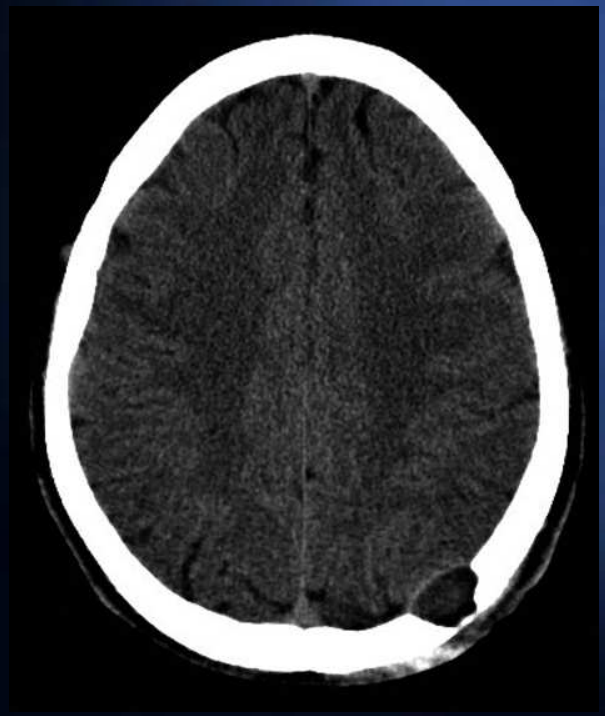

Os
SNC
Forme pseudo-tumorale
Atteinte
Hypothalamo-hypophysaire
Forme
pseudo-dégénérative

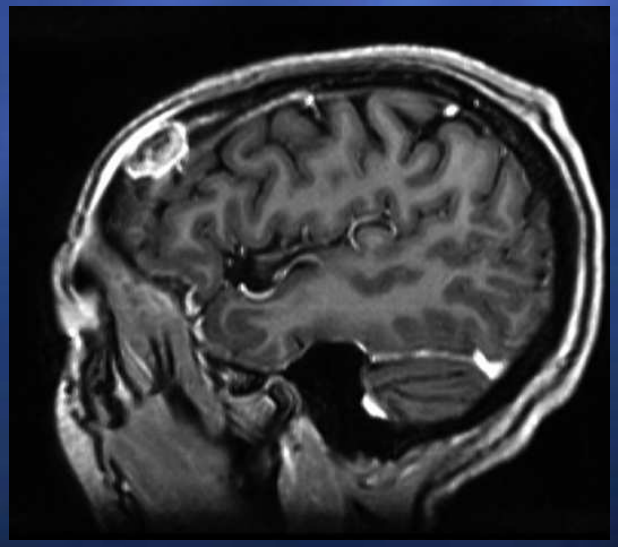

Os
Granulomes éosinophiles
classiques de la voûte
Atteinte de
la base du crâne
- Sphénoïde
-Rocher
Atteinte du massif facial

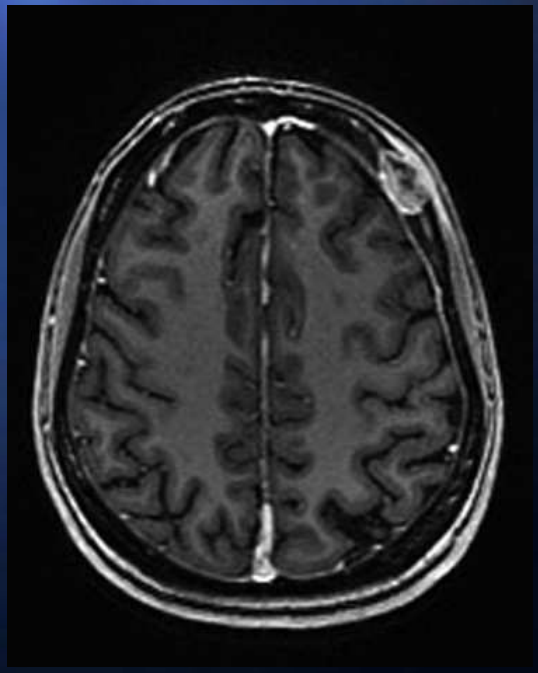


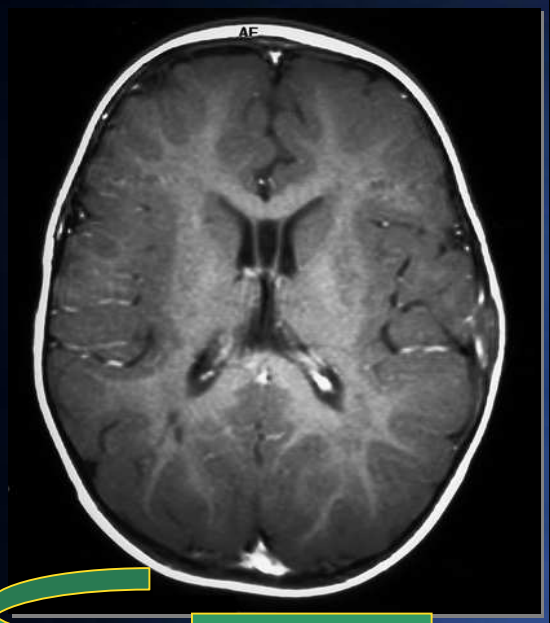


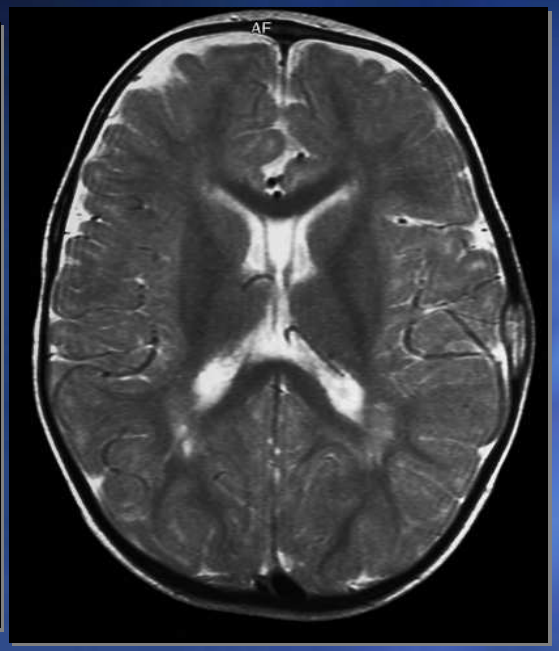








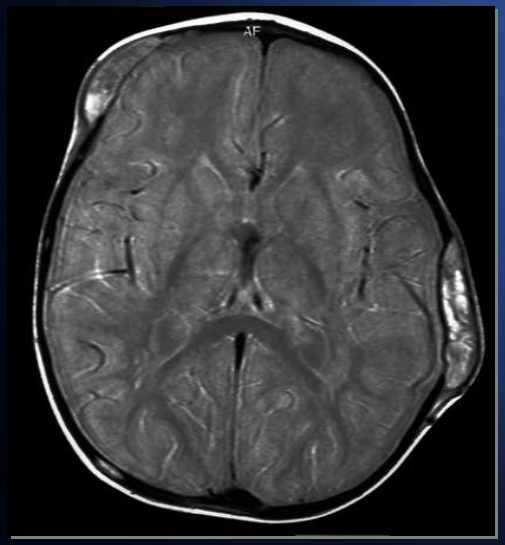


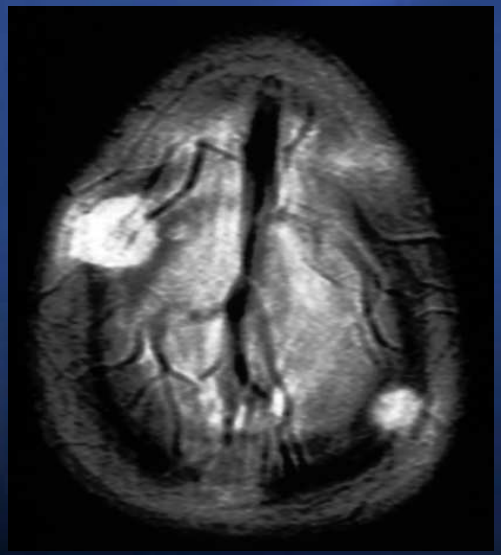


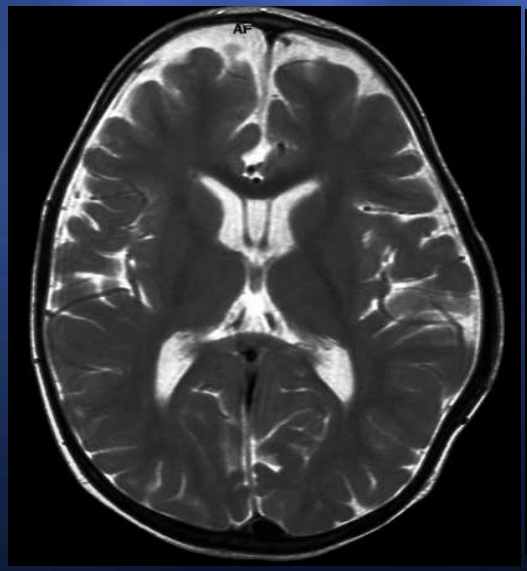
Granulomes éosinophiles
classiques de la voûte

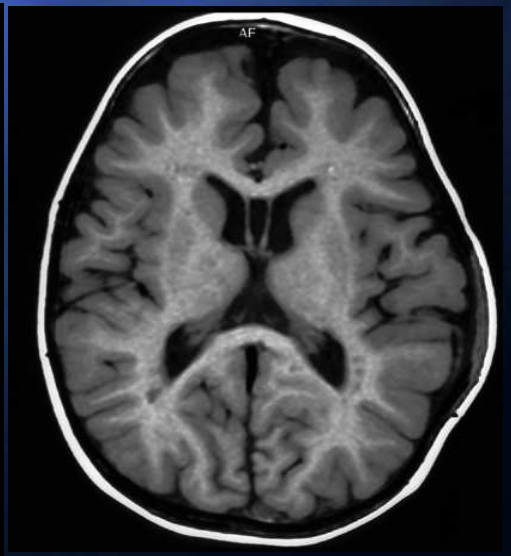
+ 6 mois

+ 1 an

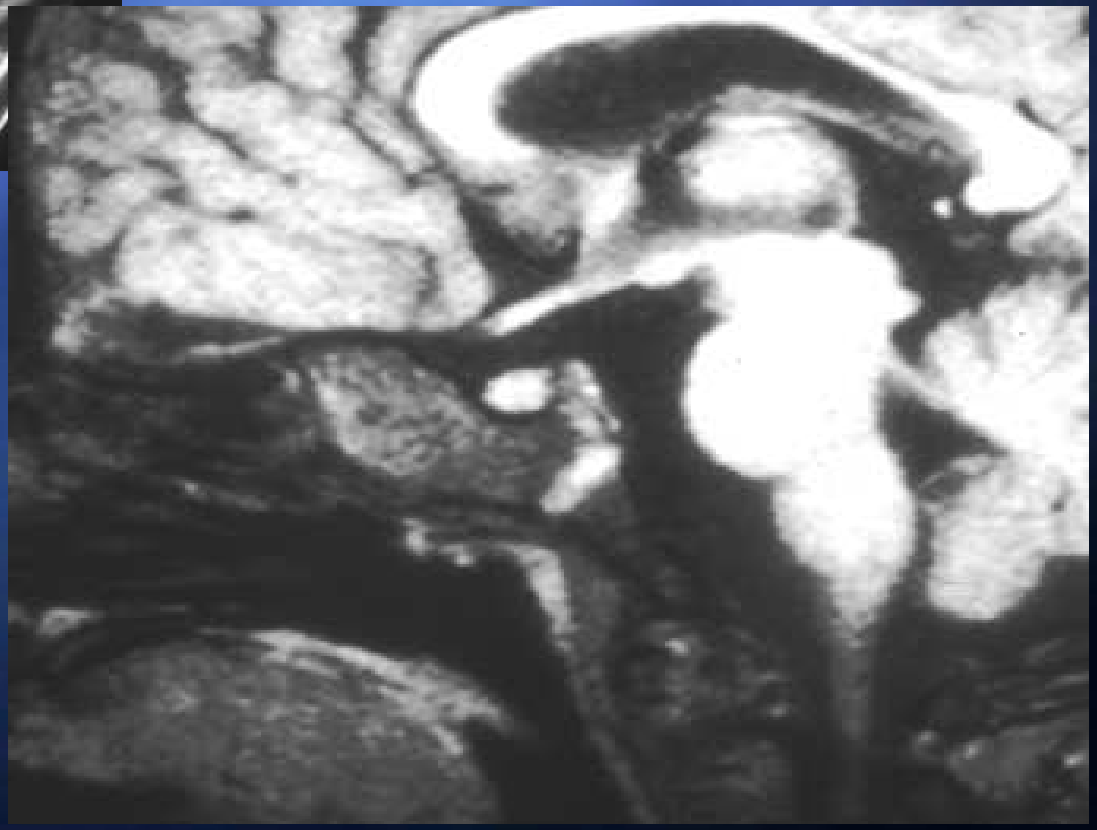


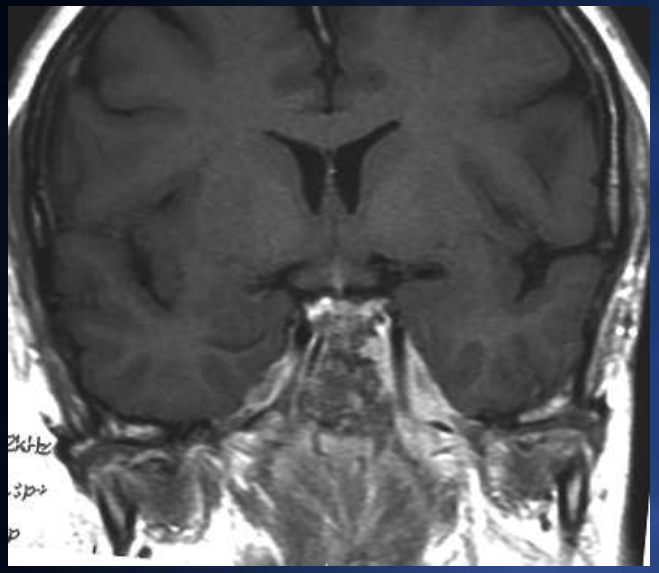


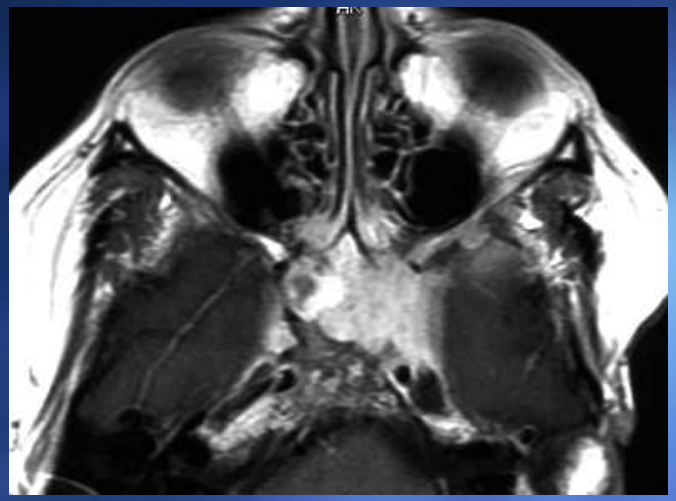


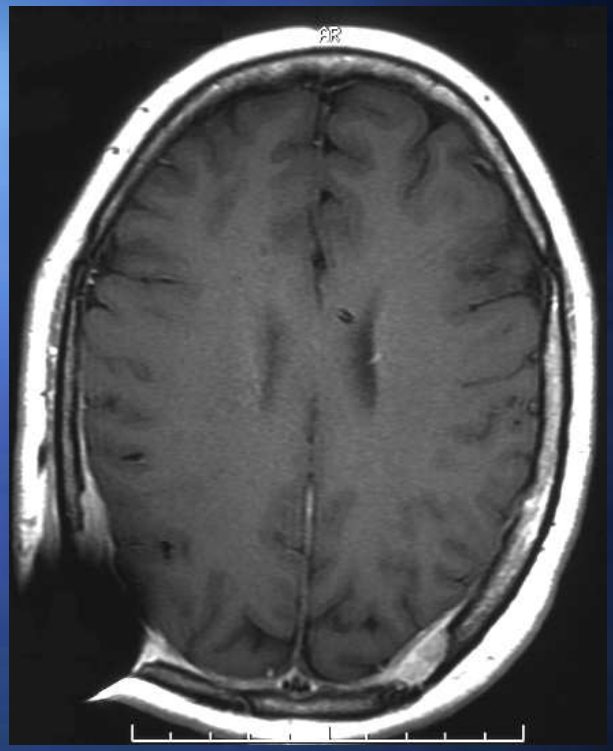


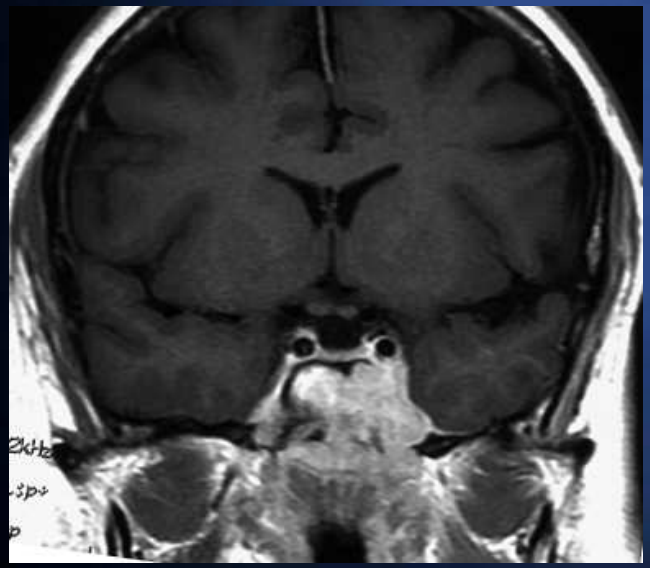
Atteinte de
la base du crâne
- Sphénoïde
-Rocher

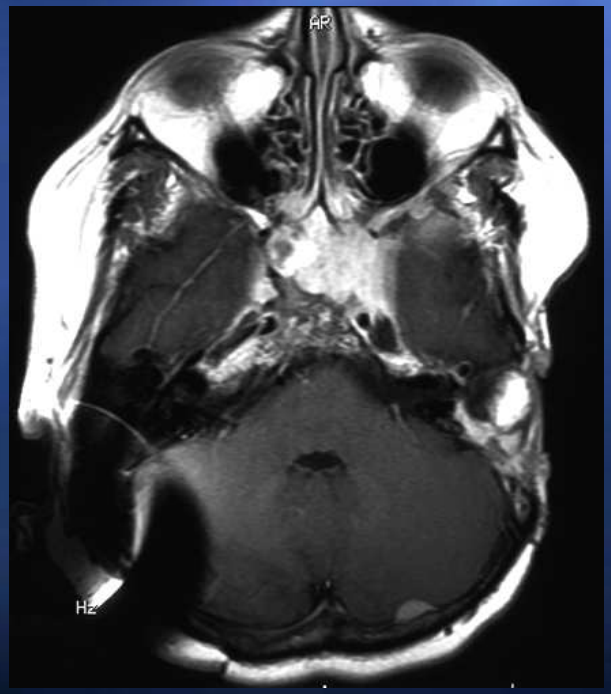


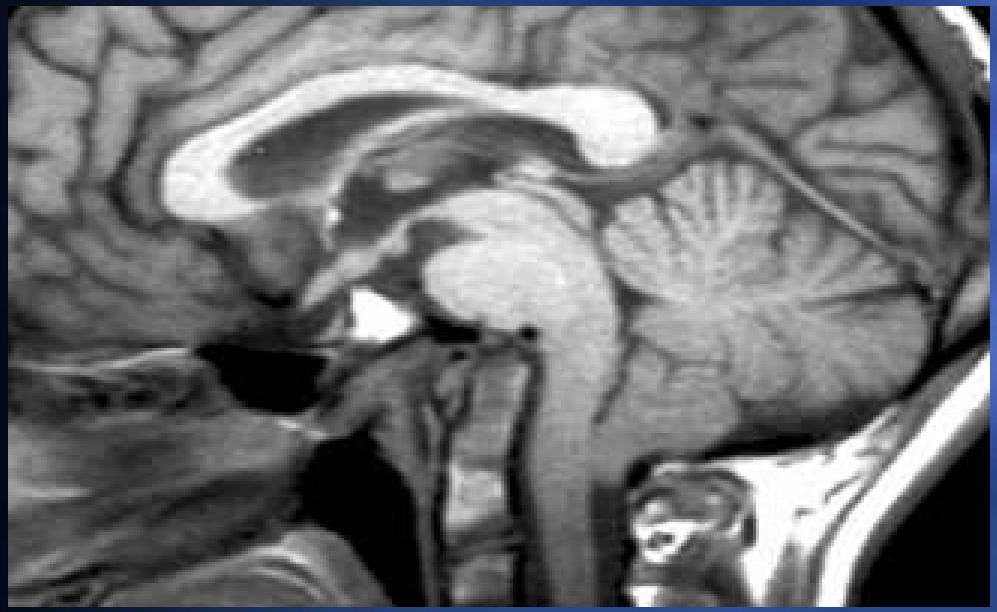


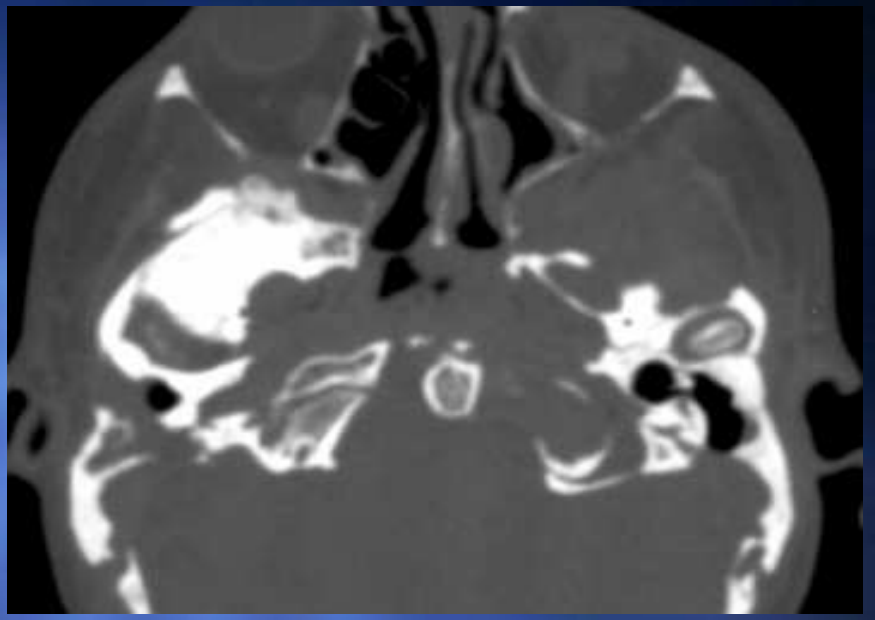


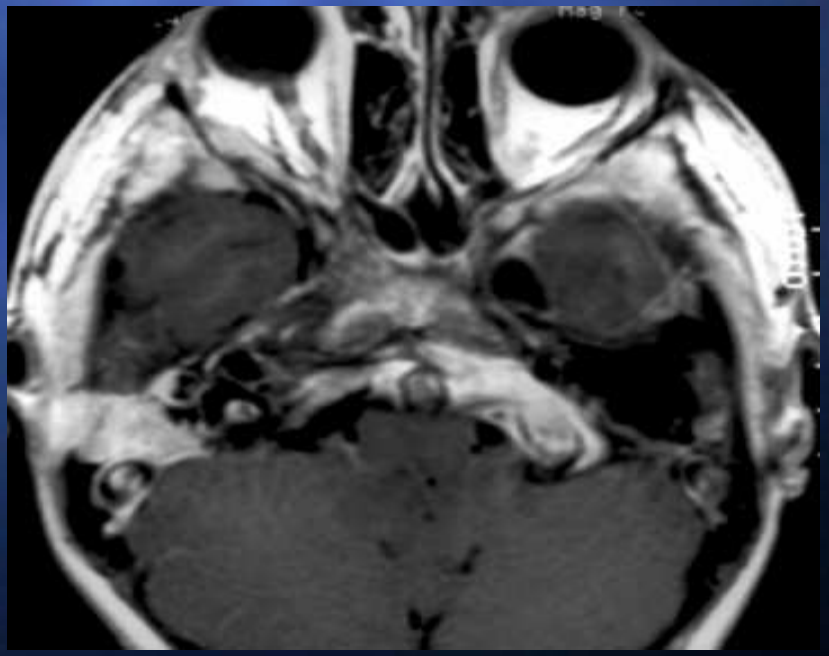


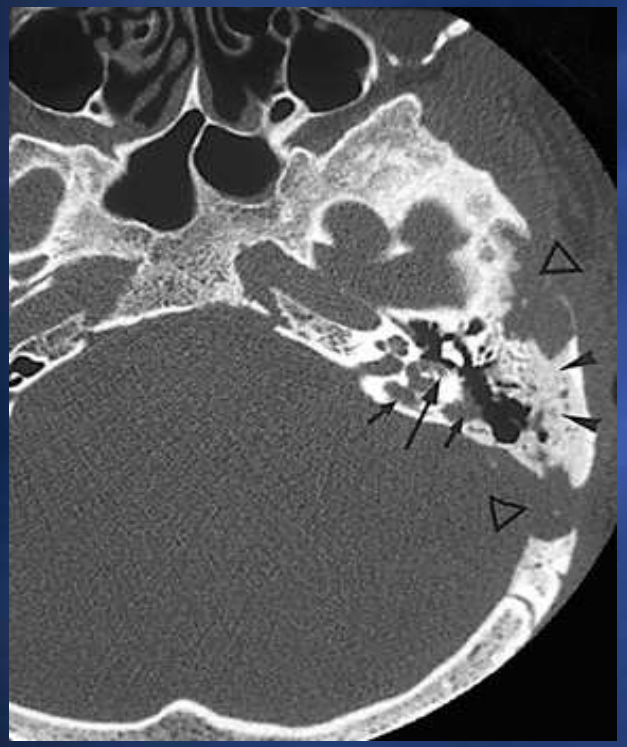


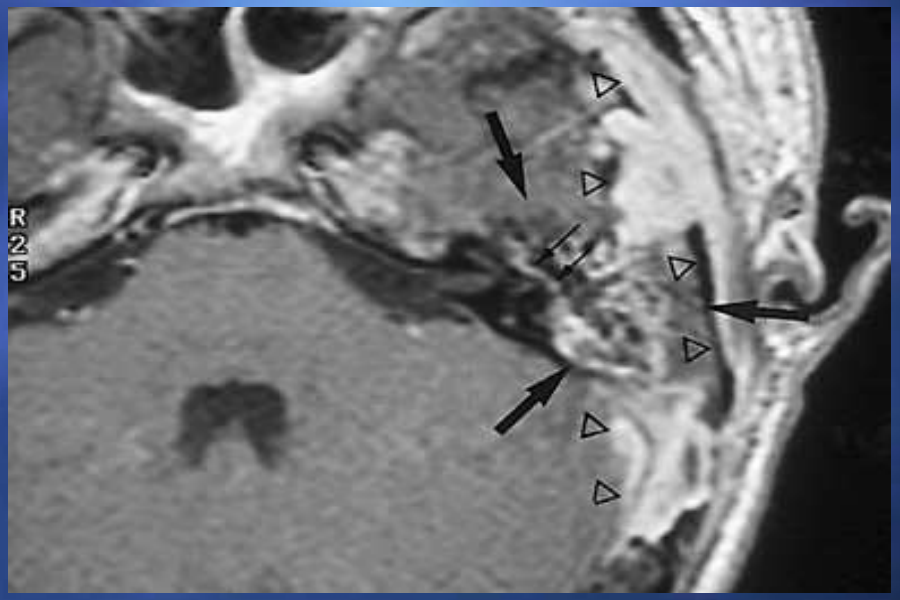











R
2
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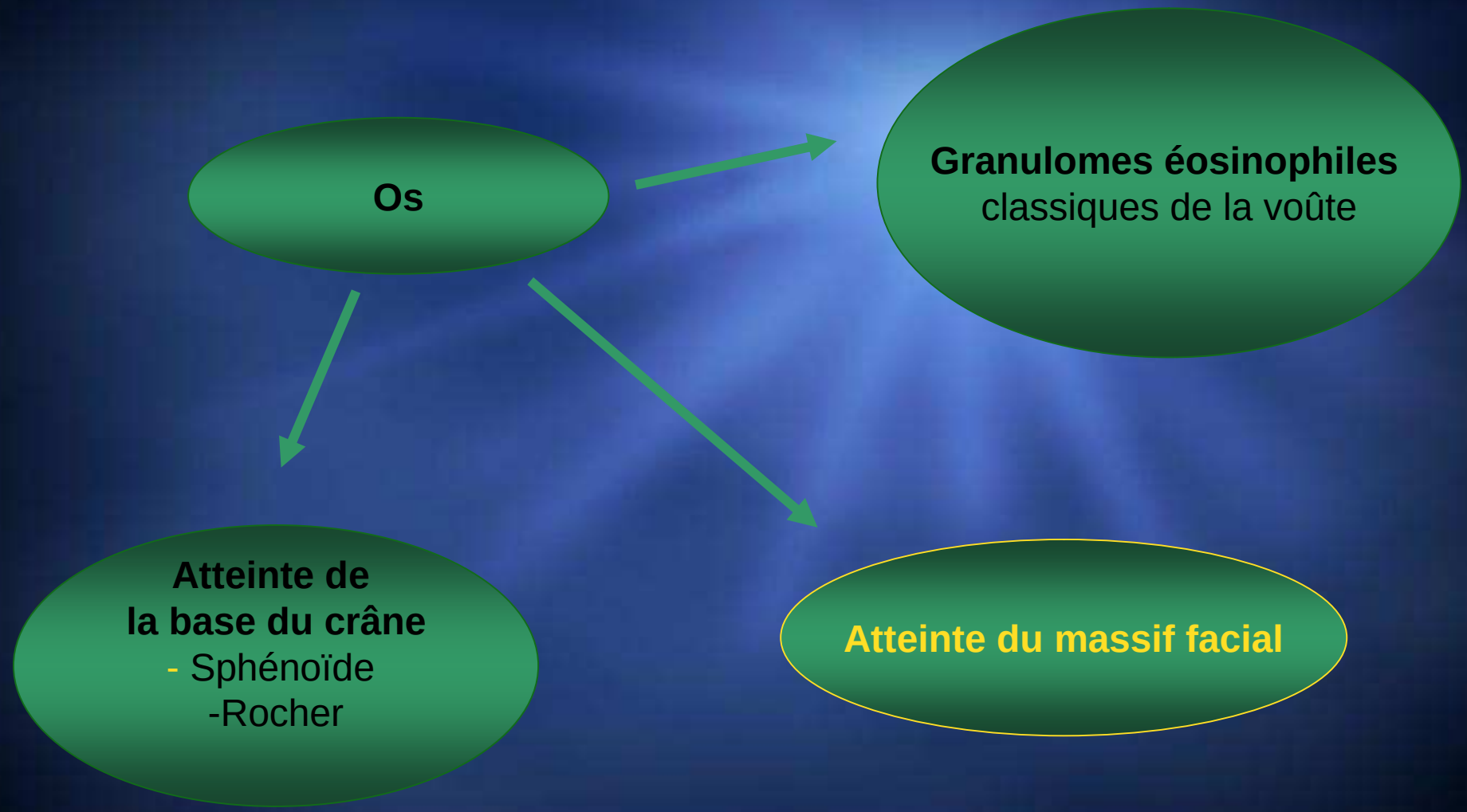
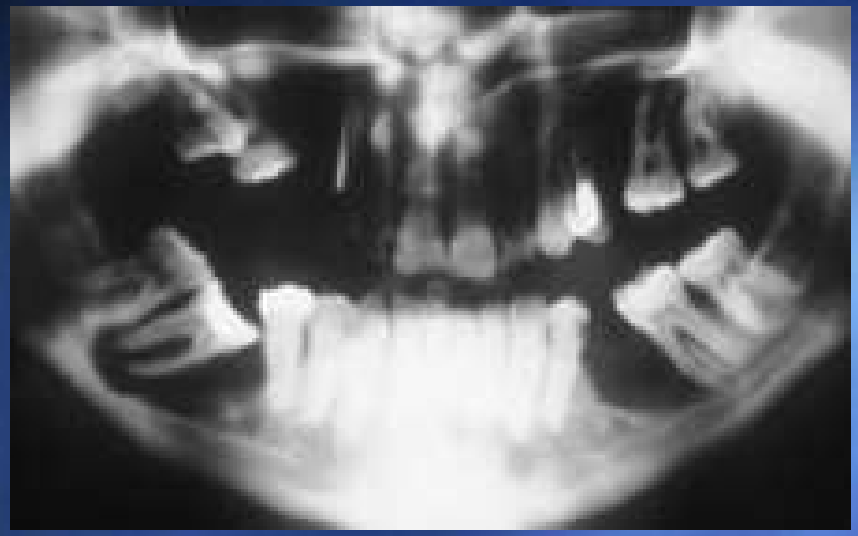
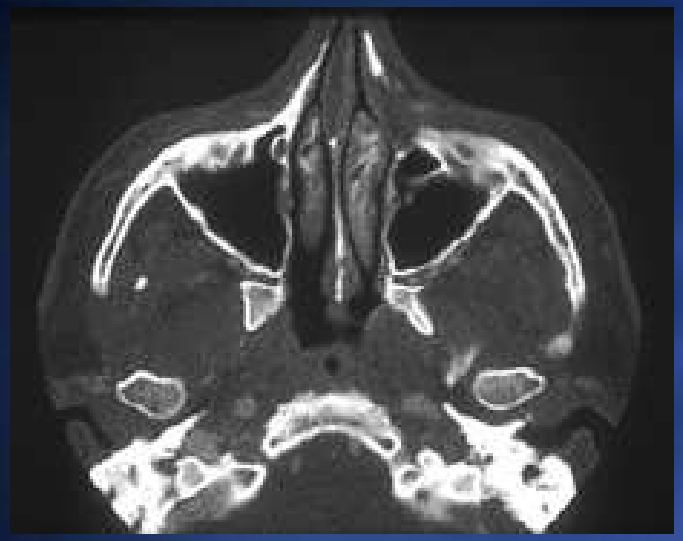
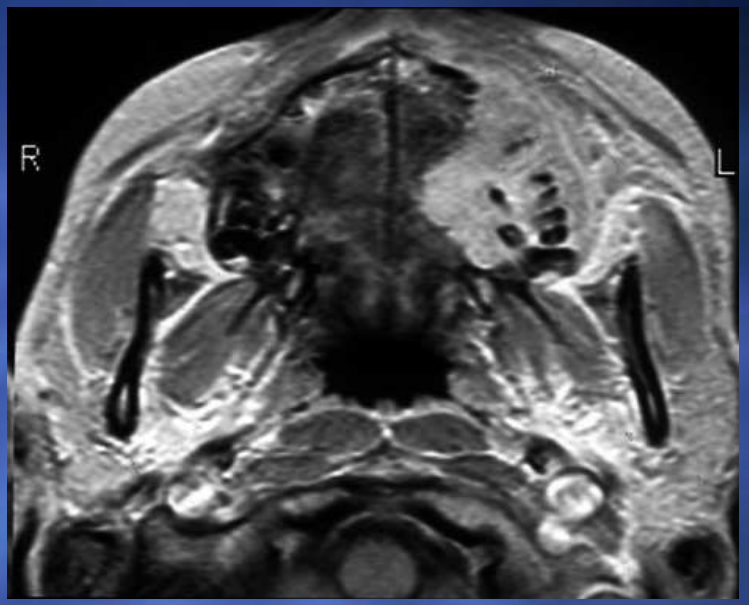
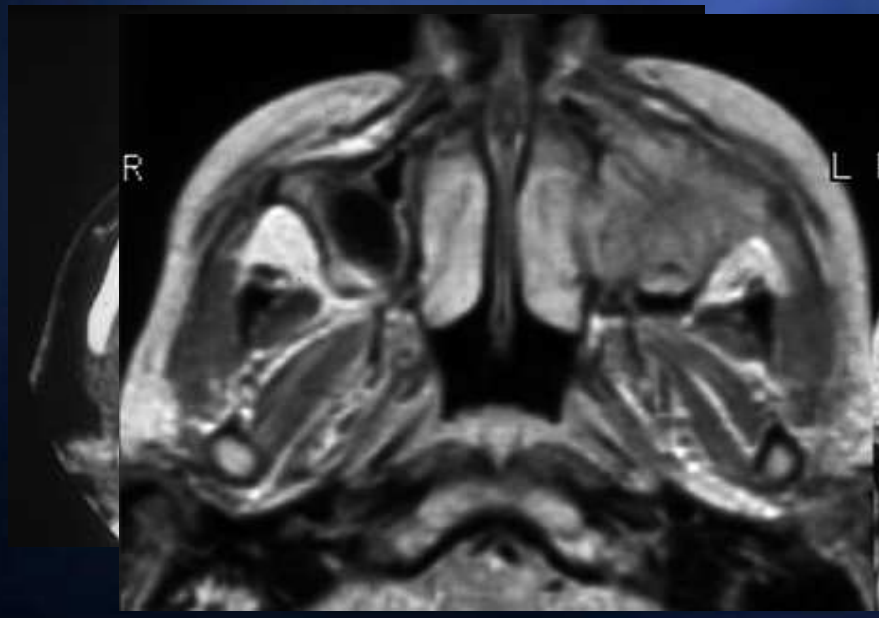
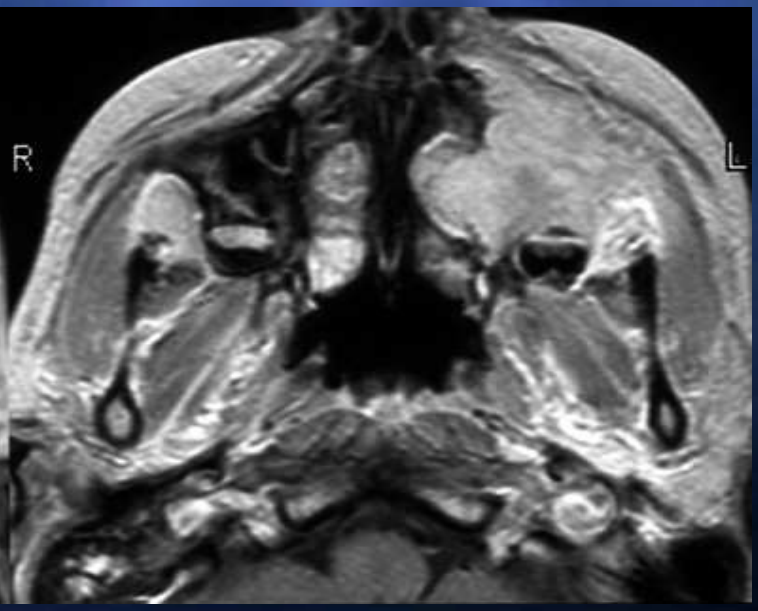

Os
Granulomes éosinophiles
classiques de la voûte
Atteinte de
la base du crâne
- Sphénoïde
-Rocher
Atteinte du massif facial






R
L


R
L


R
L

Formes pseudo-tumorales

Os

Méninges

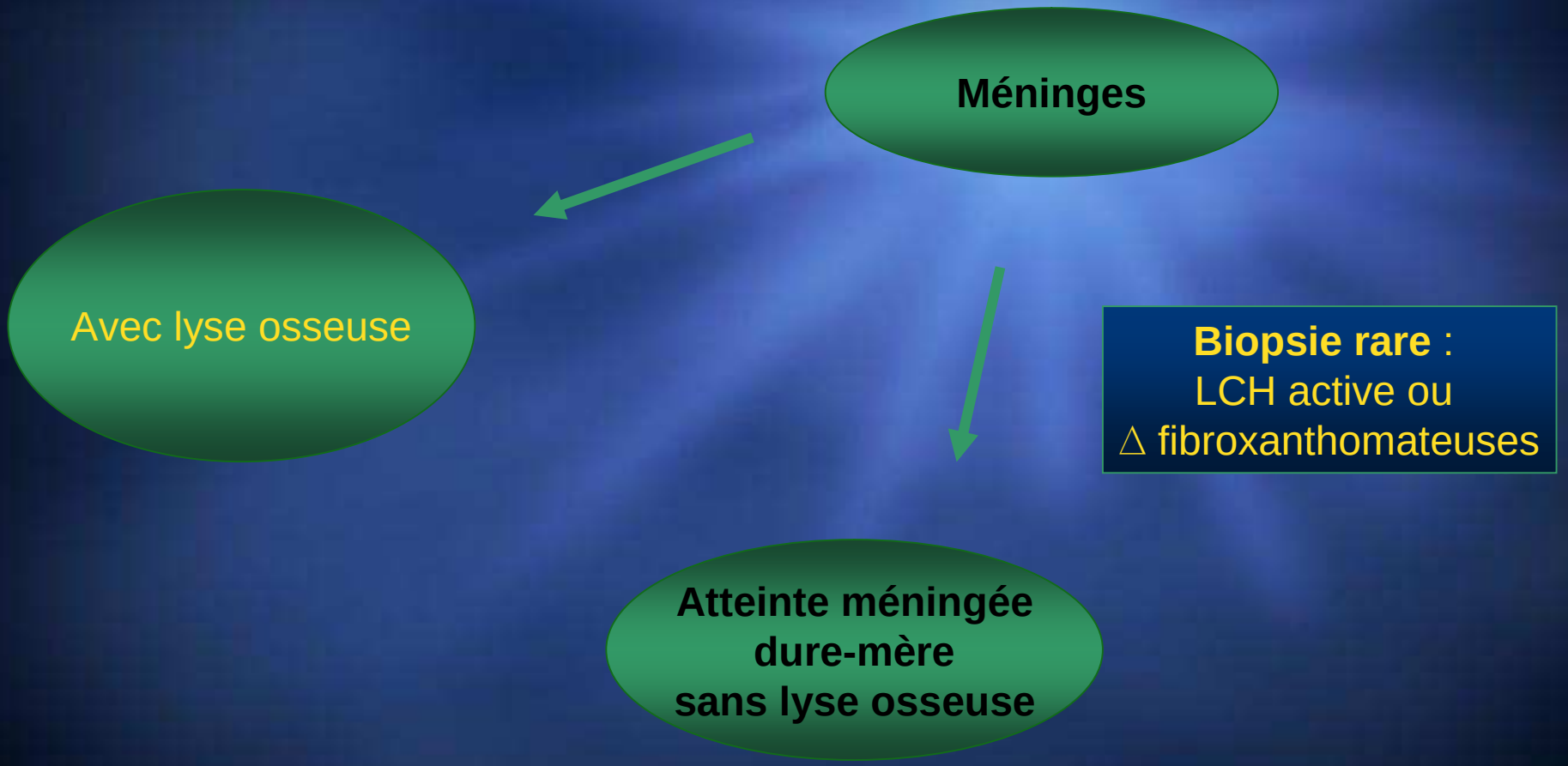
Axe
Hypothalamo-hypophysaire

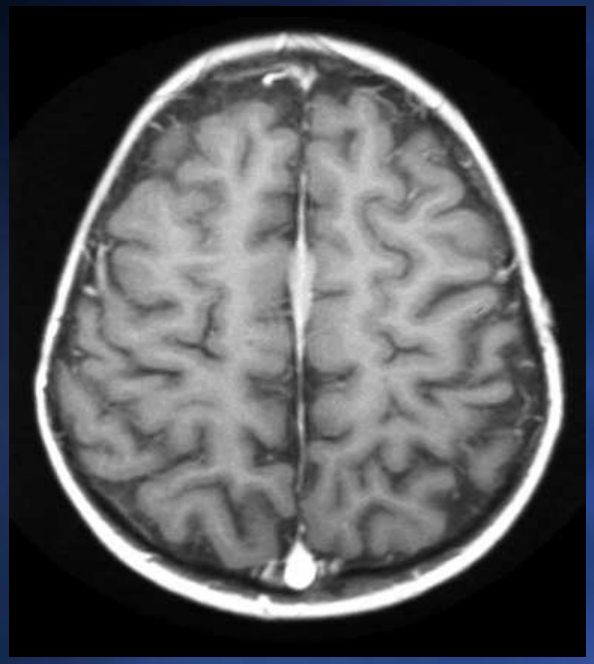
Diversité
Association lésionnelle

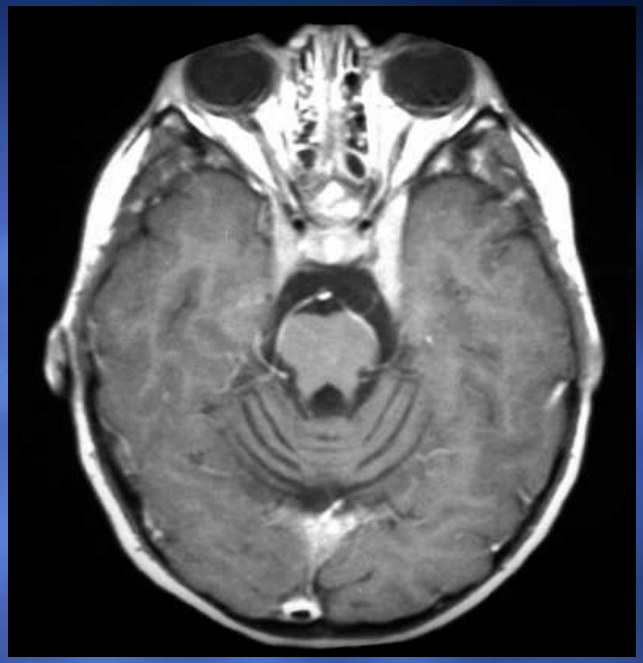
Plexus Choroïdes

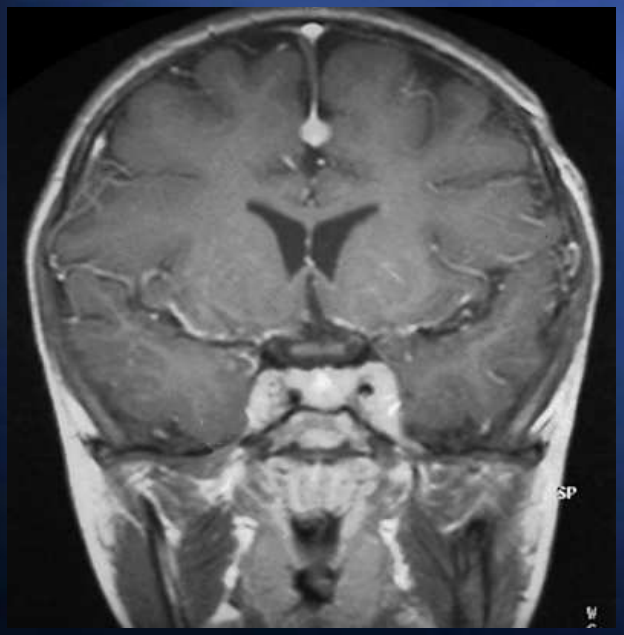
Masses
intra-parenchymateuses

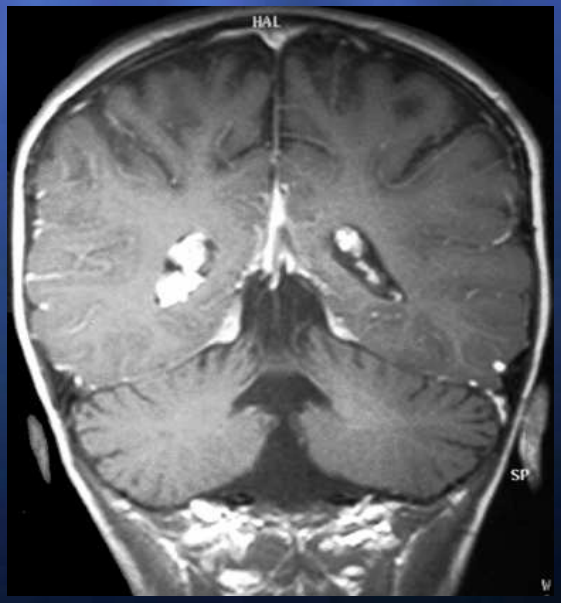
Pinéale


Méninges
Avec lyse osseuse
Biopsie rare :
LCH active ou
△ fibroxanthomateuses
Atteinte méningée
dure-mère
sans lyse osseuse

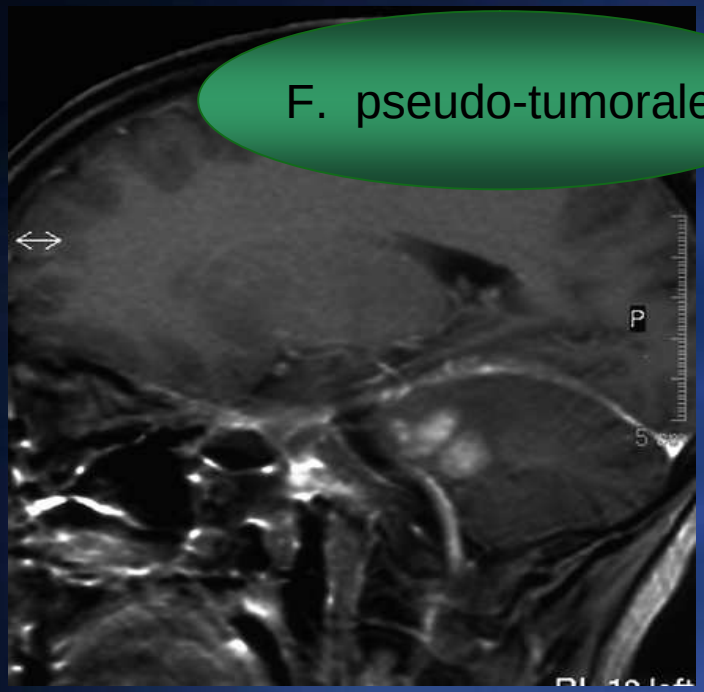


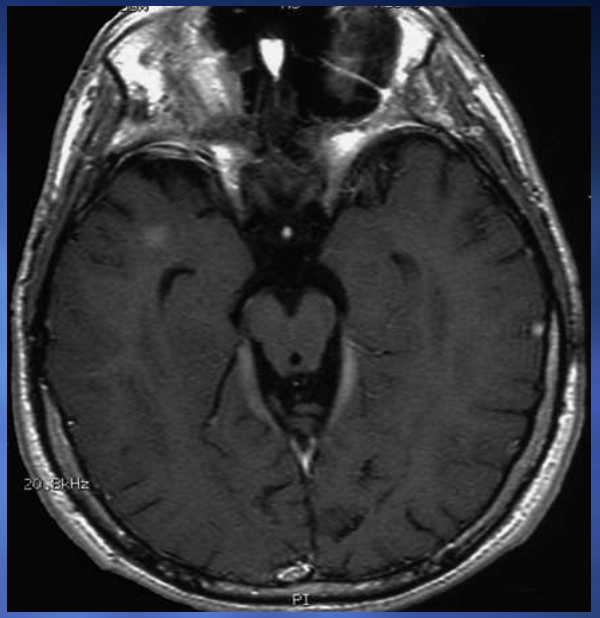


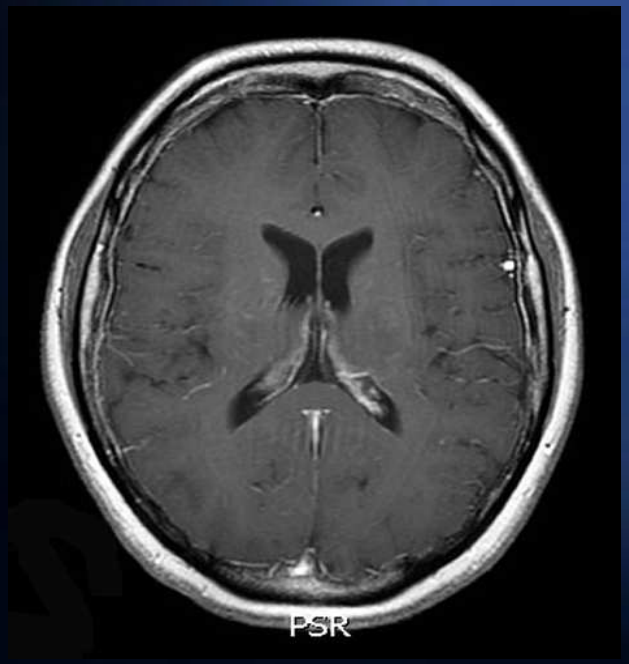


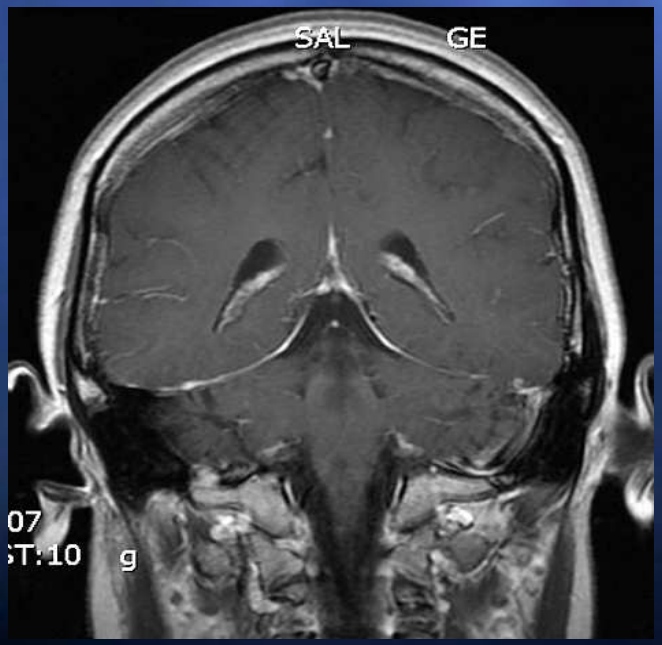

HAL
SP

F. pseudo-tumorale

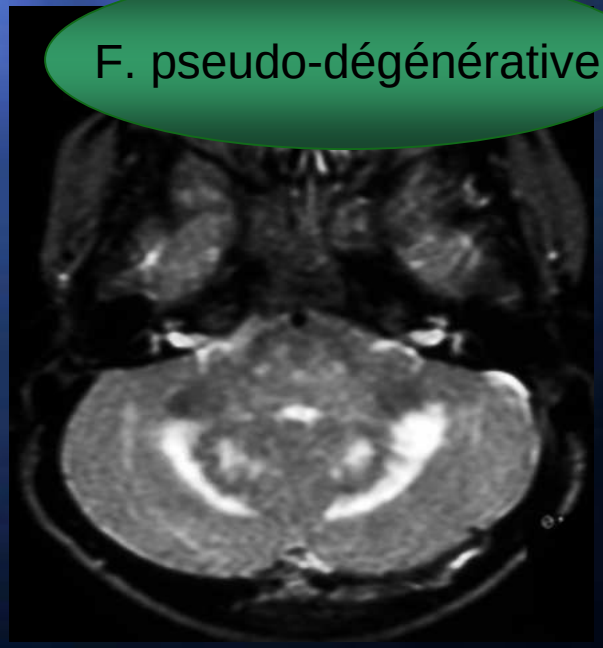


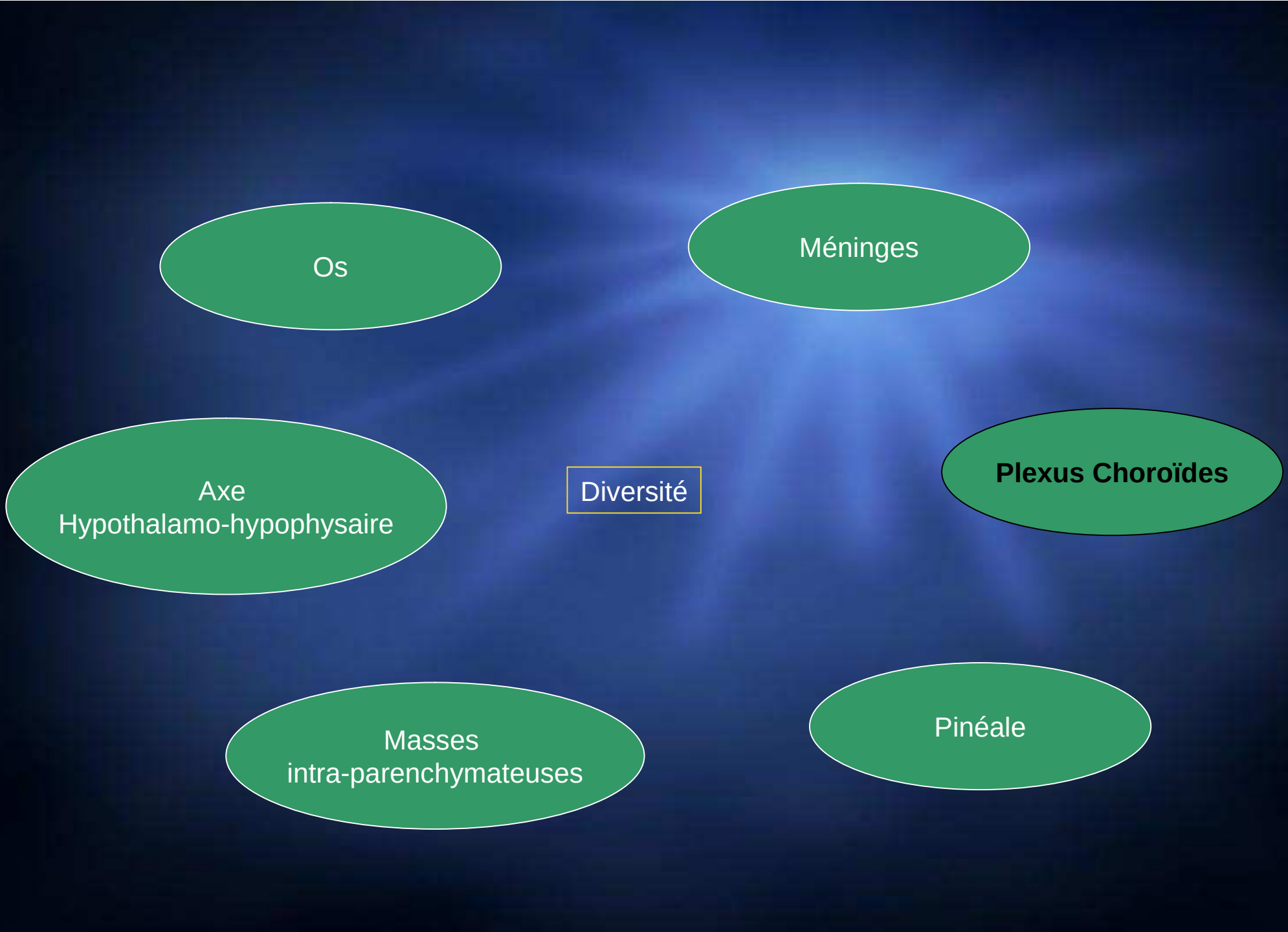






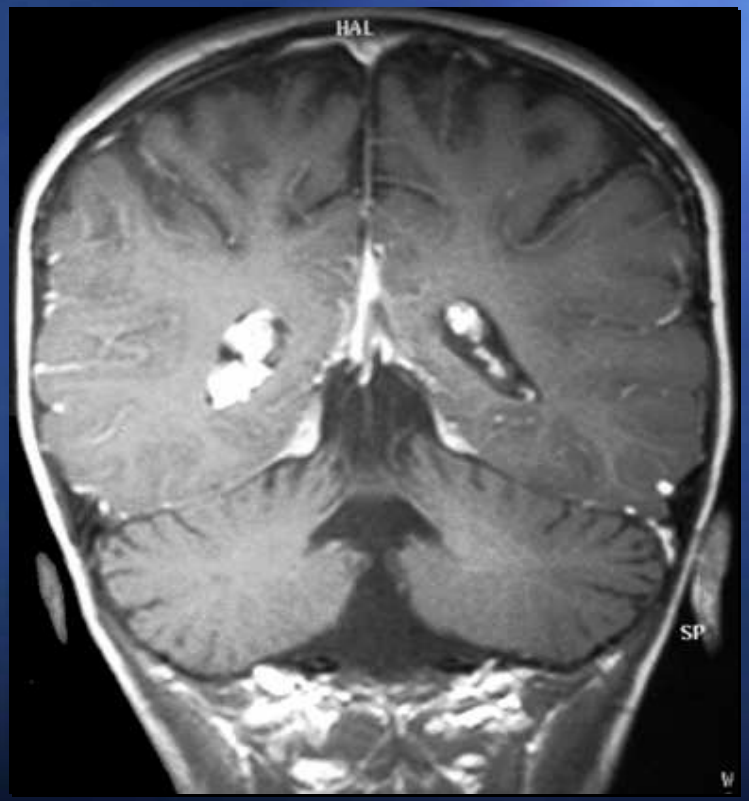
F. pseudo-dégénérative



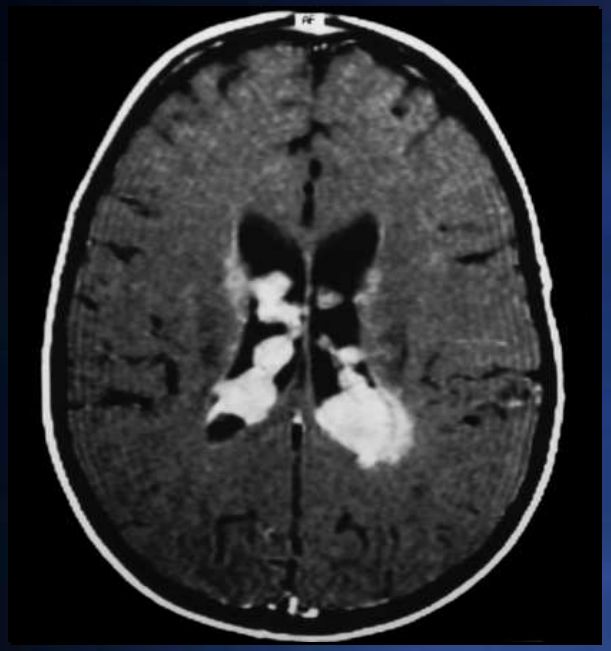

Os
Méninges
Axe
Hypothalamo-hypophysaire
Diversité
Plexus Choroïdes
Masses
intra-parenchymateuses
Pinéale

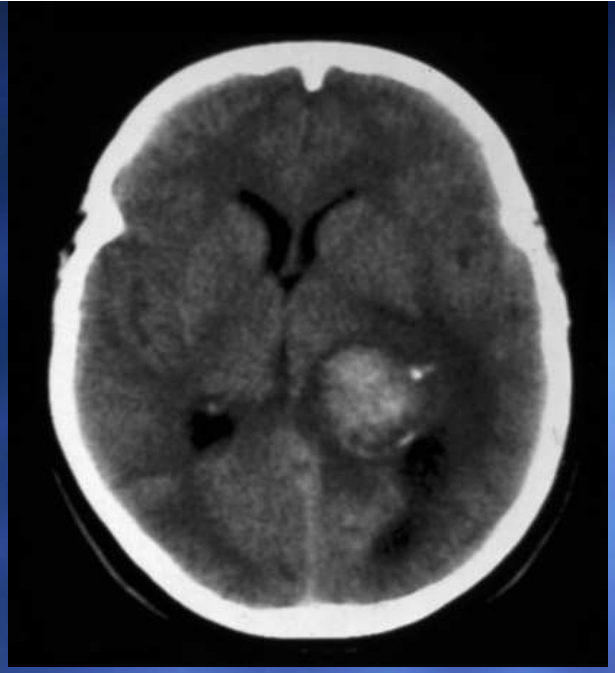
Critères diagnostiques:

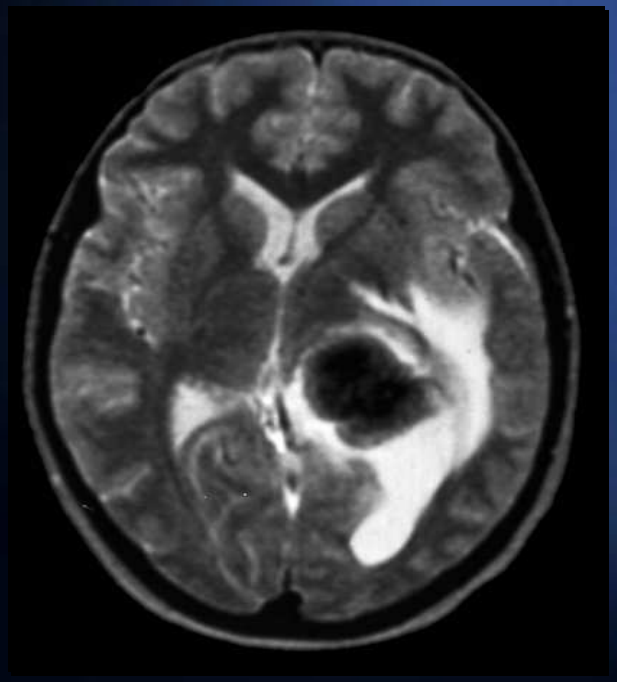
- Masses bilatérales
- Taille > 1/3 ep trigone
- Calcifications ≠ normale
- Cawliflowerlike
- ↑ taille



Plexus Choroïdes



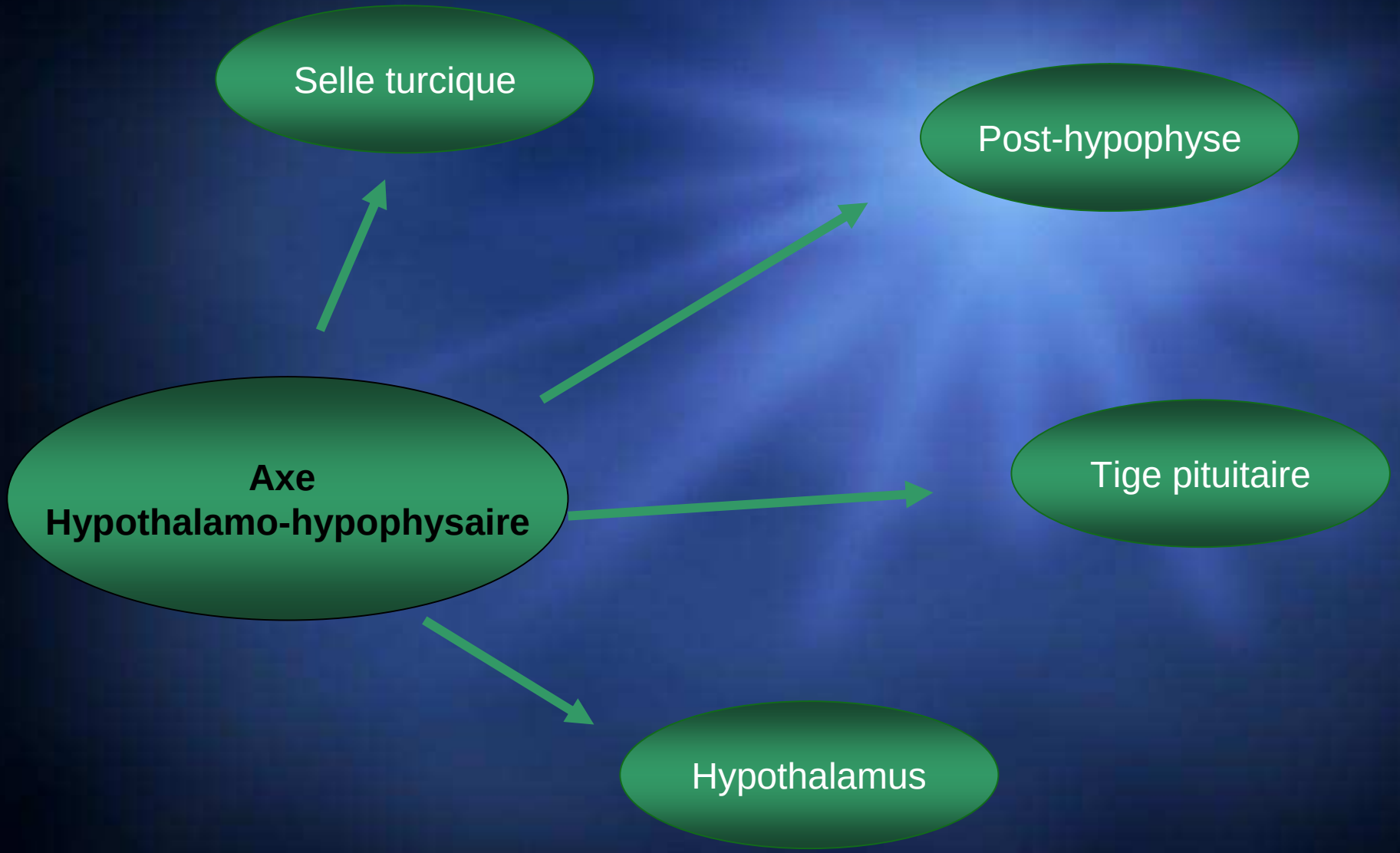




Plexus Choroïdes

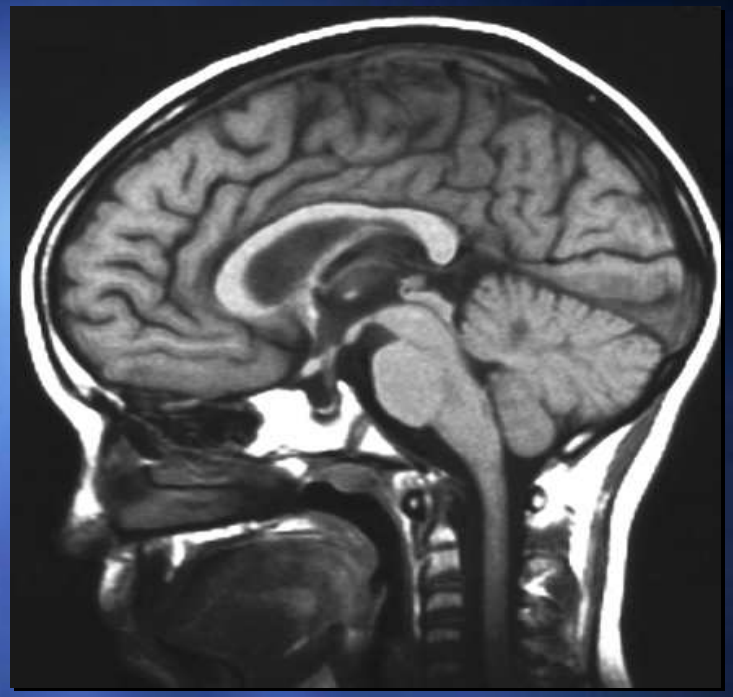
- △ kystiques 28%
- ↑ taille > 10mm 14%

Pinéale

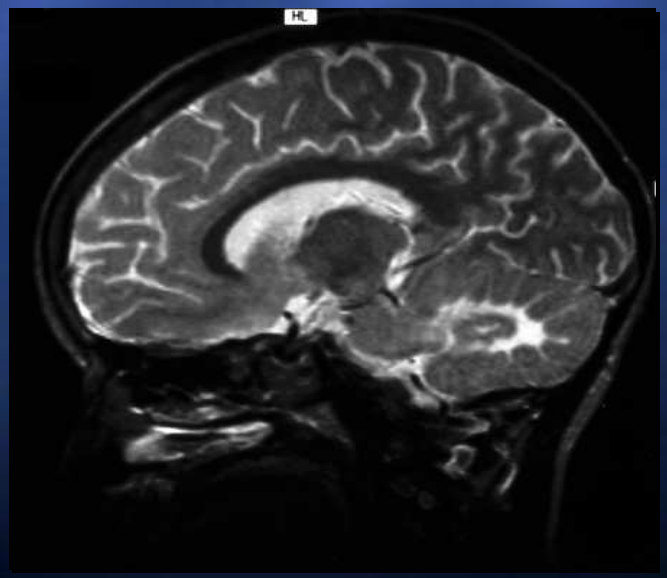

Selle turcique
Post-hypophyse
Axe
Hypothalamo-hypophysaire
Tige pituitaire
Hypothalamus

Selle turcique

- Normale
- Petite
- Vide
- ↑ taille



Axe
Hypothalamo-hypophysaire



+ F. dégénérative

Selle turcique

Evolution

Axe
Hypothalamo-hypophysaire

+ 4 ans

Selle turcique

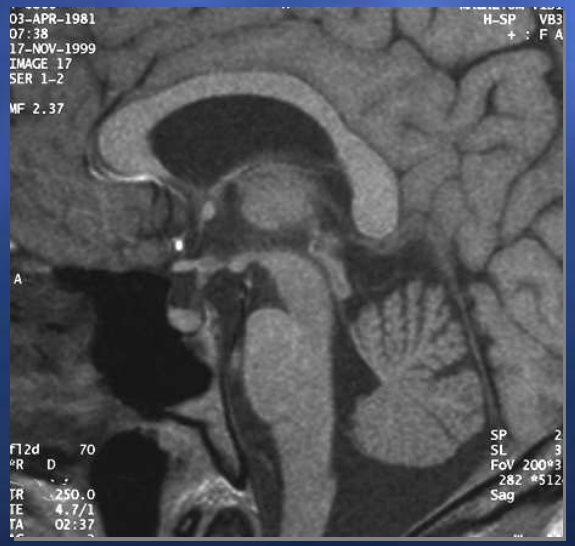
- Normale
- Petite
- Vide
- ↑ taille

Axe
Hypothalamo-hypophysaire

- He T1 présent
- He T1 absent
- Ectopique

Post-hypophyse

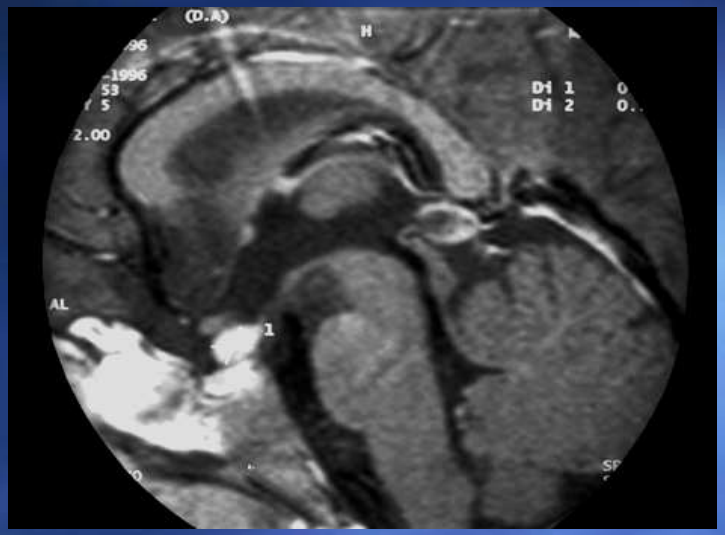
Axe
Hypothalamo-hypophysaire

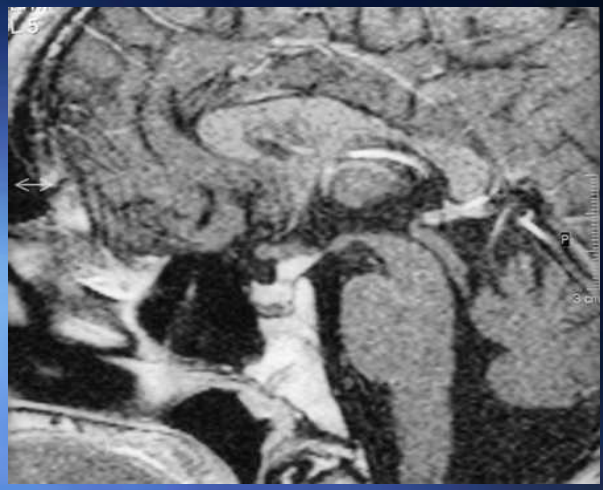


- **Nle = n'élimine pas le dg**
- Ep > 2 ou 3 mm

dans au – une des 3 portions

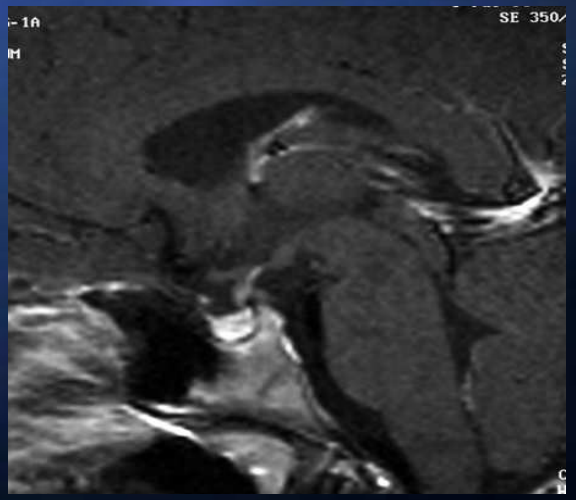
- Threadlike < 1 mm



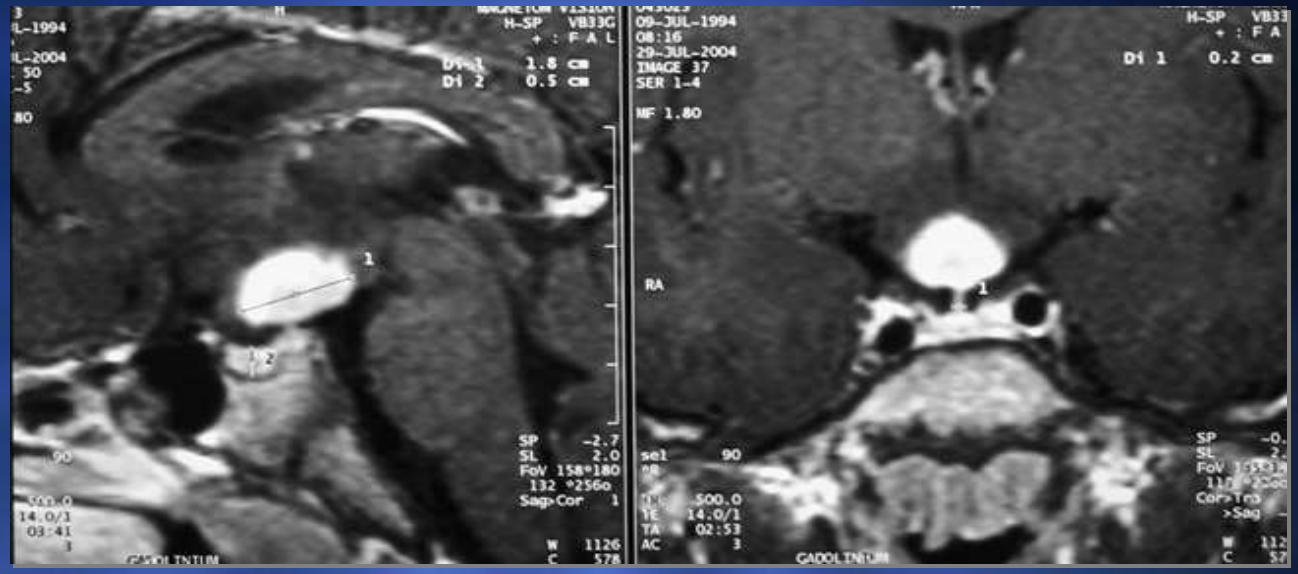


Axe
Hypothalamo-hypophysaire

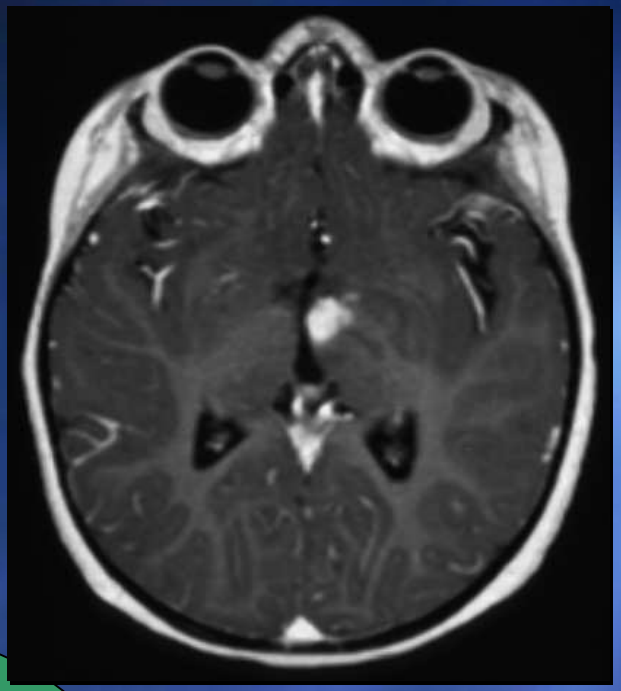
Tige pituitaire

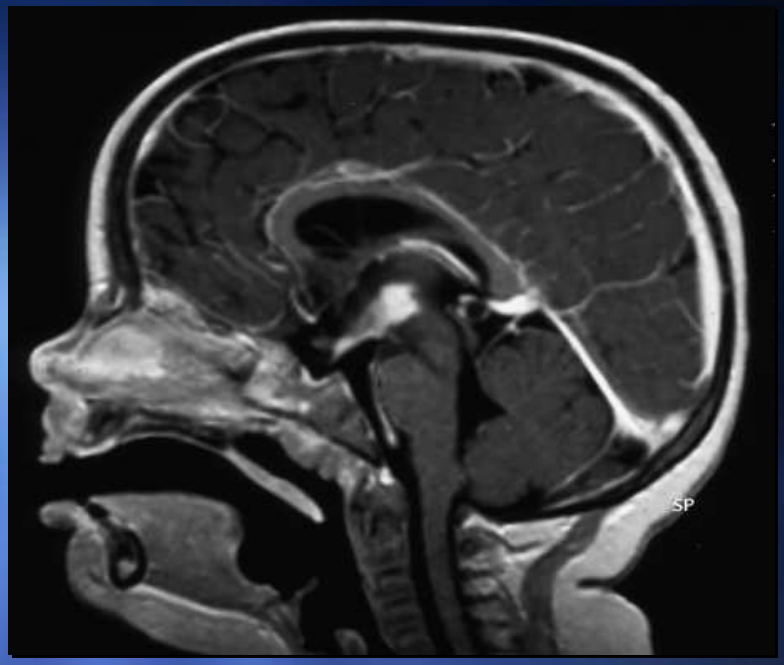


Axe
Hypothalamo-hypophysaire



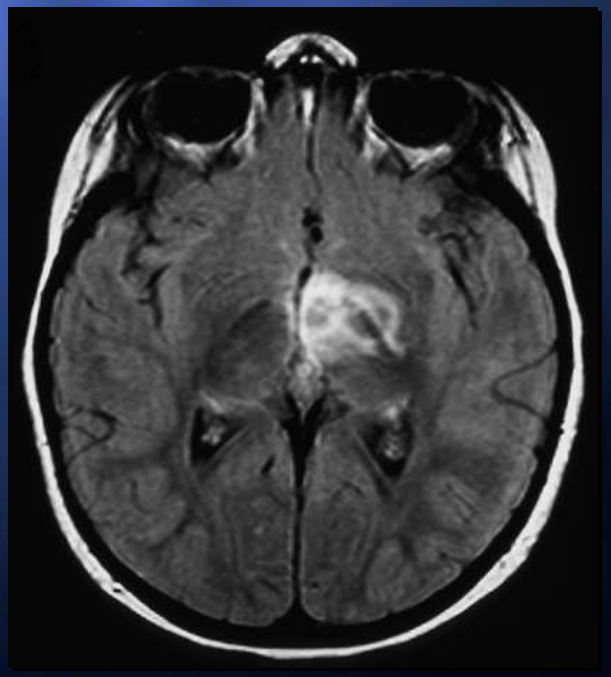
Hypothalamus

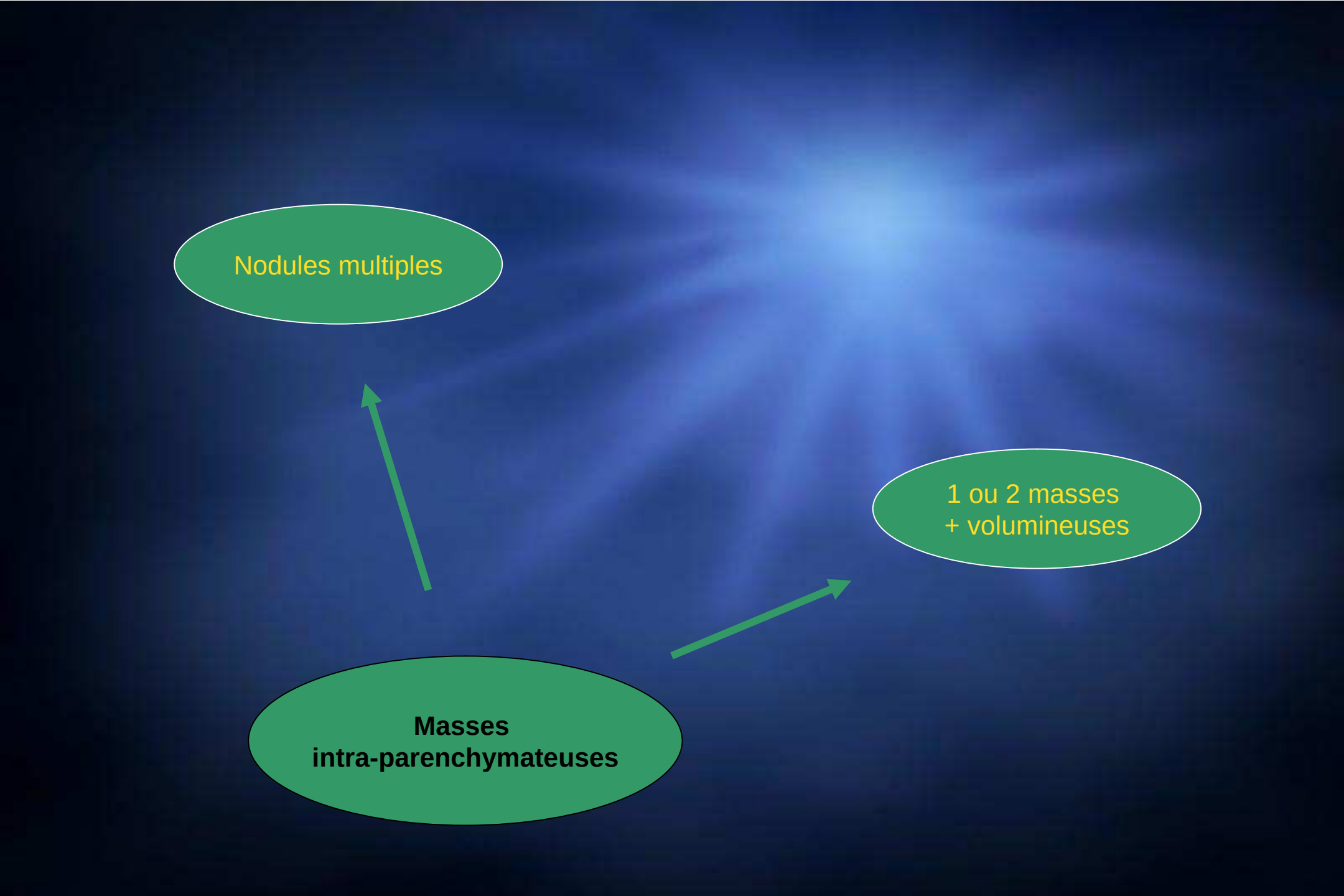




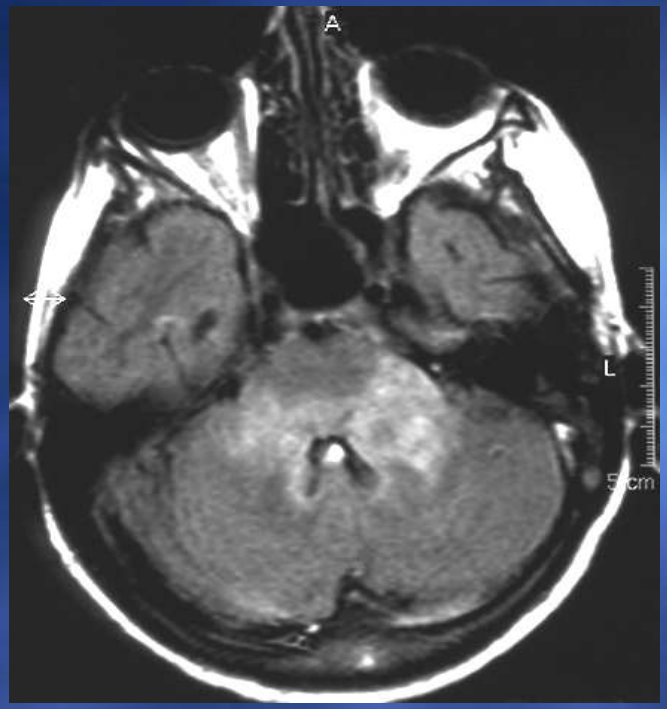
Axe
Hypothalamo-hypophysaire

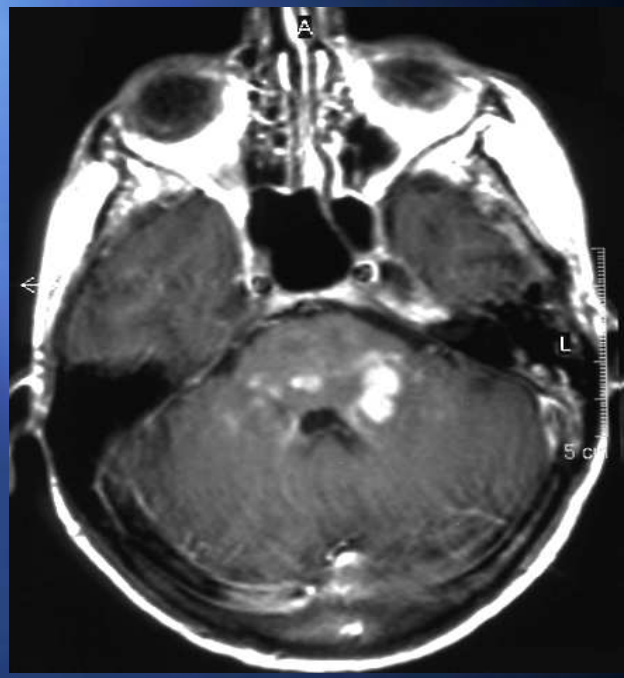
Hypothalamus




Nodules multiples
1 ou 2 masses
+ volumineuses
Masses
intra-parenchymateuses

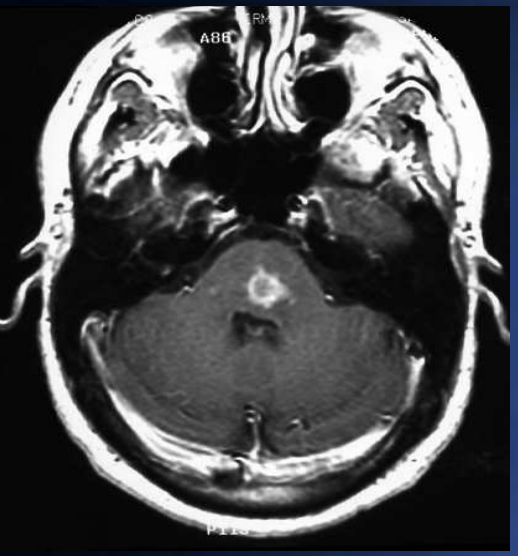
Nodules multiples

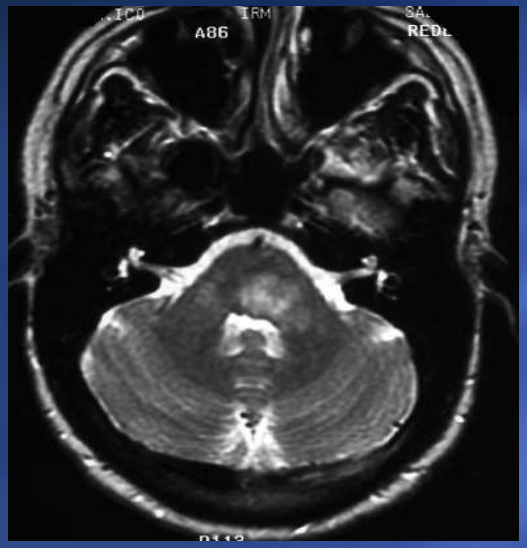


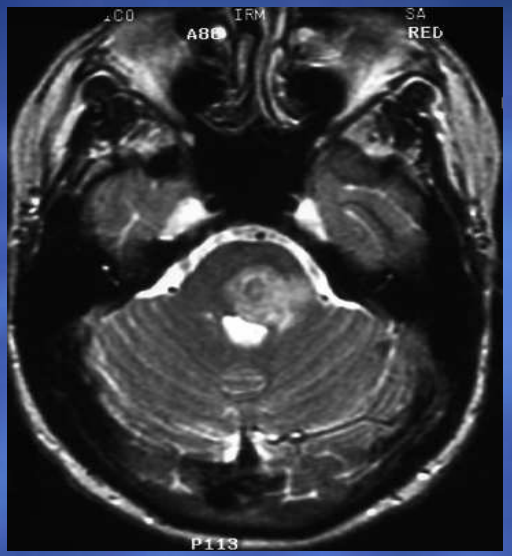


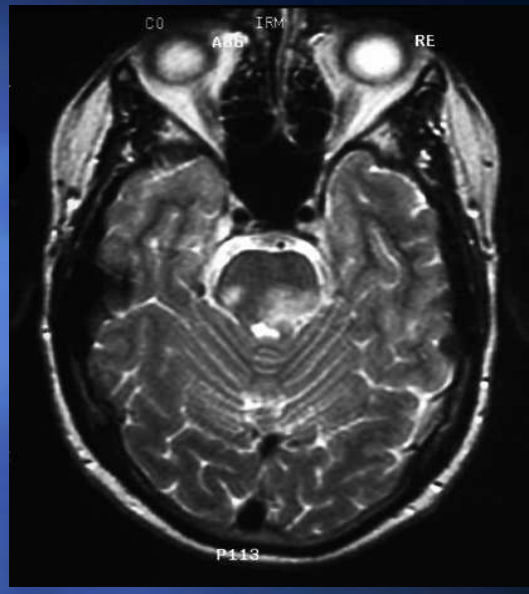
Masses
intra-parenchymateuses

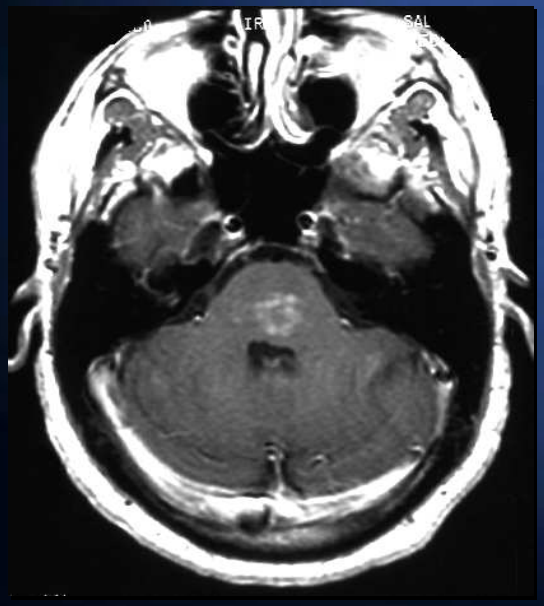
EVOLUTION +++

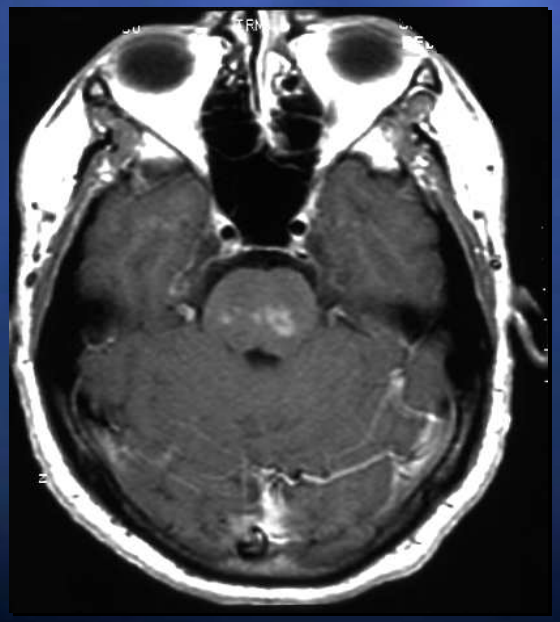


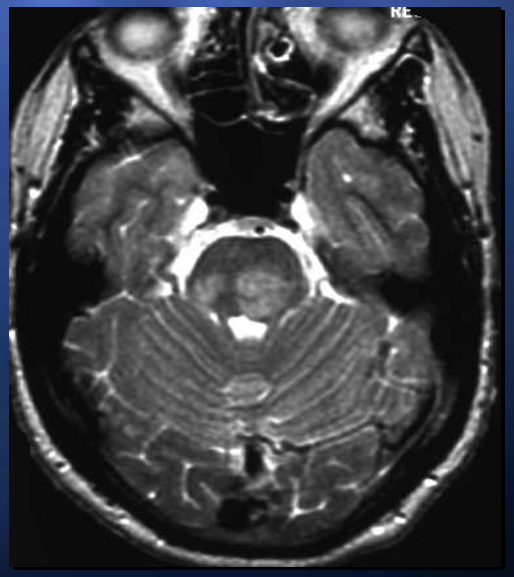




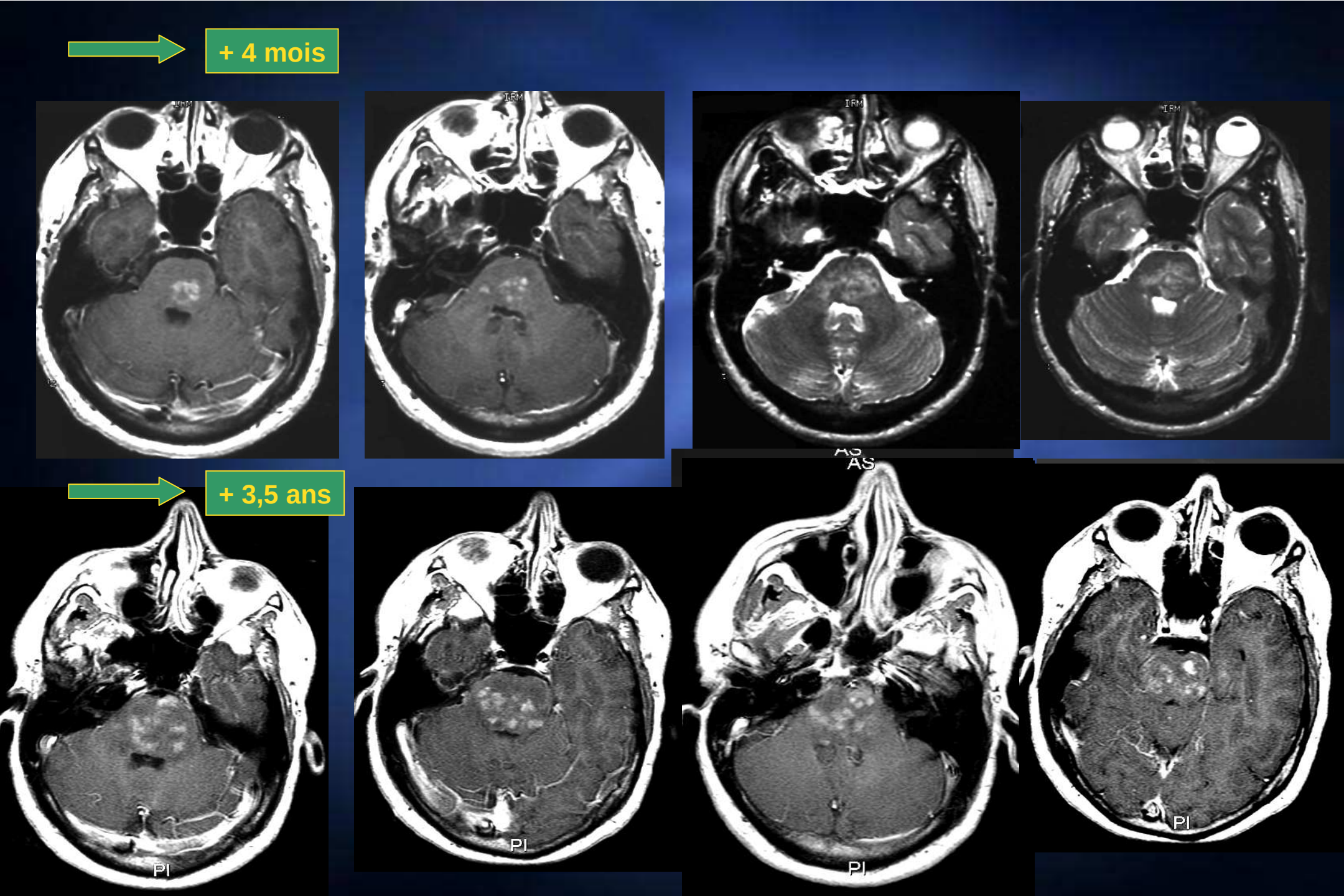




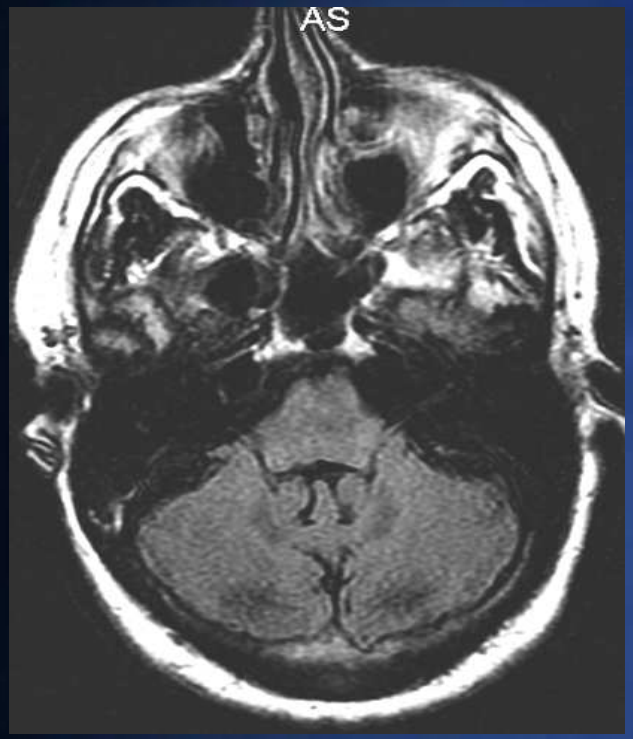


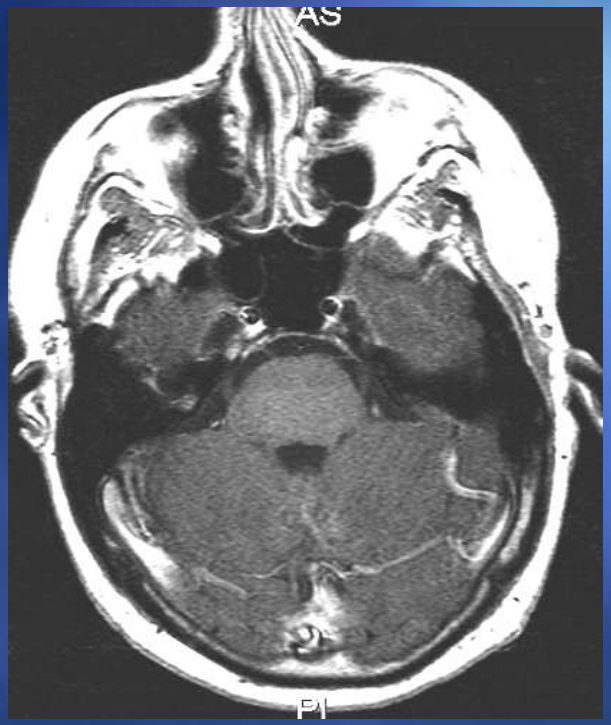


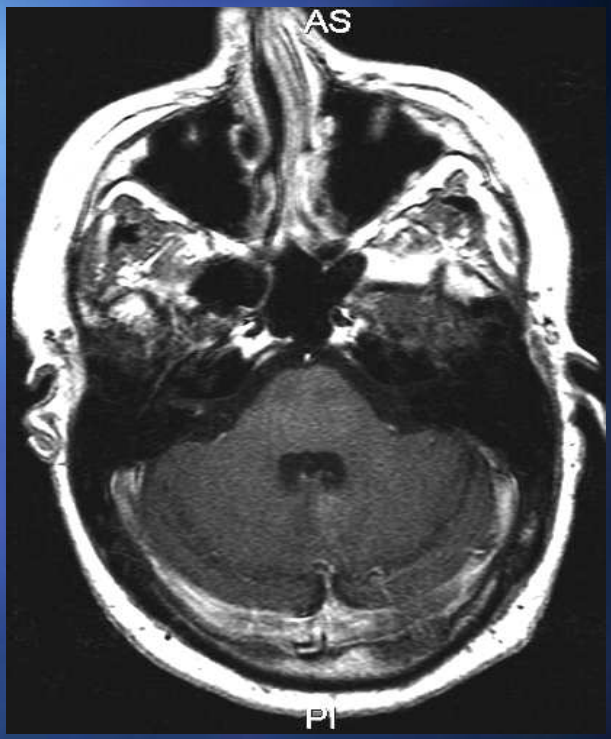
+ 7 mois

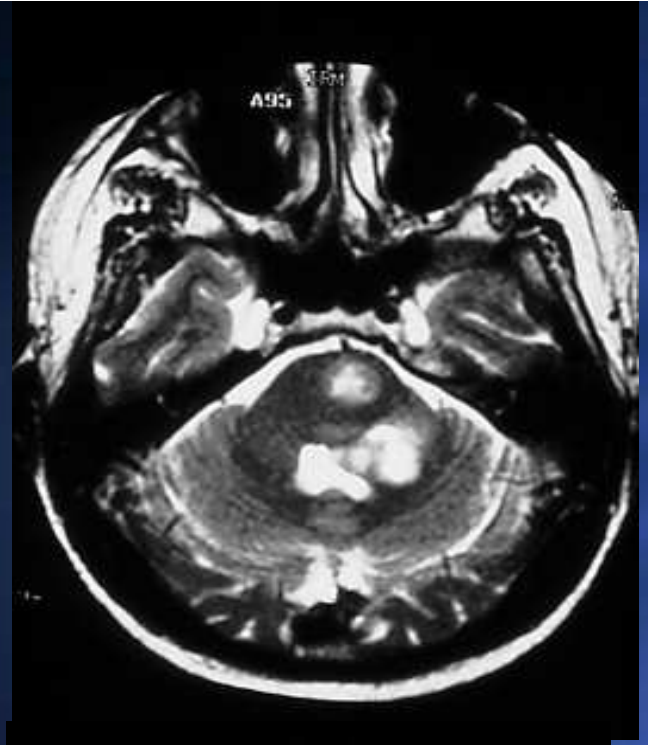

+ 4 mois
+ 3,5 ans
IRM
AS
PI

+ 1,5 an





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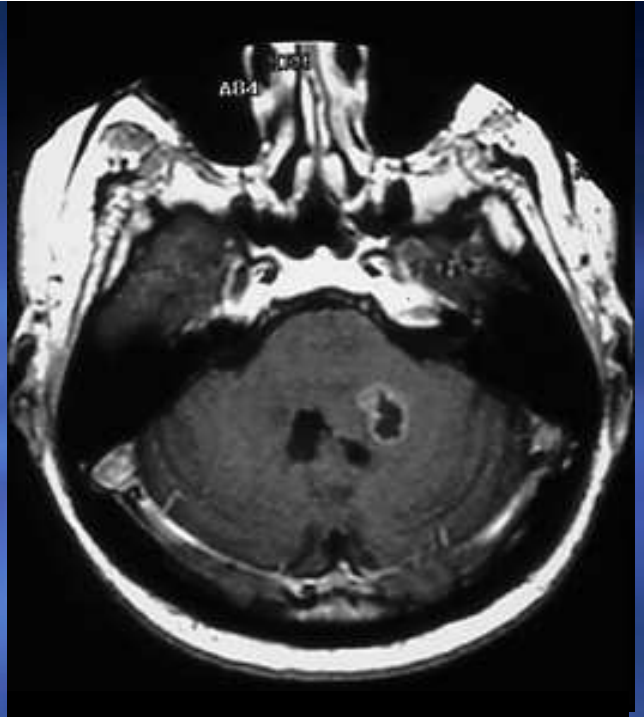
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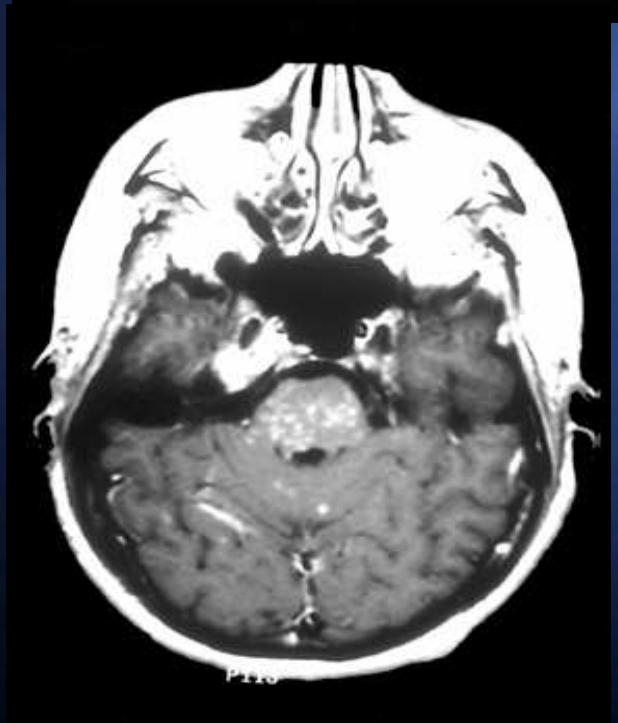
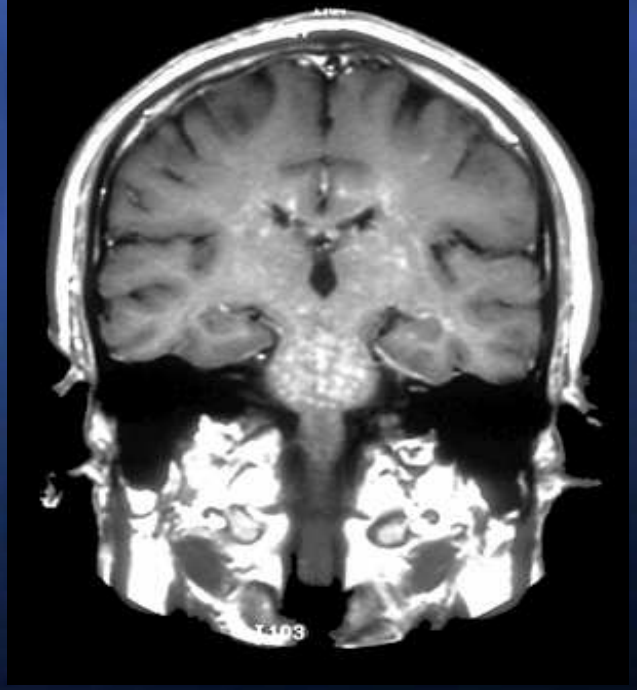
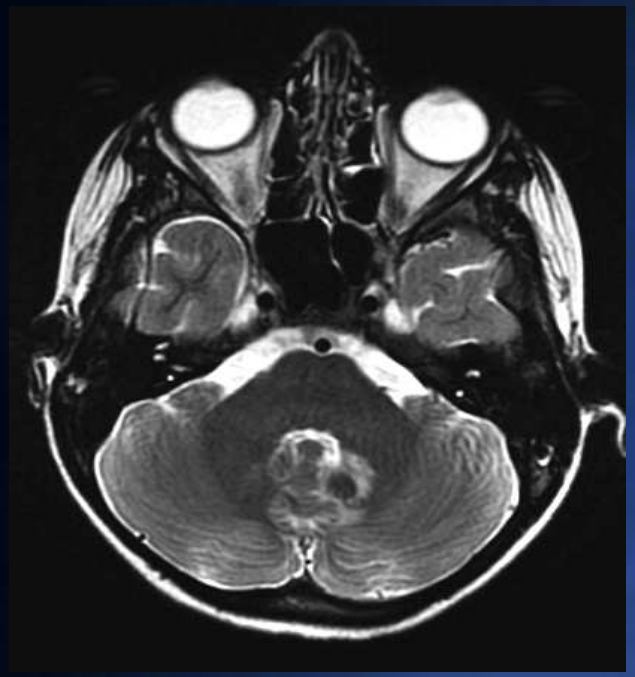
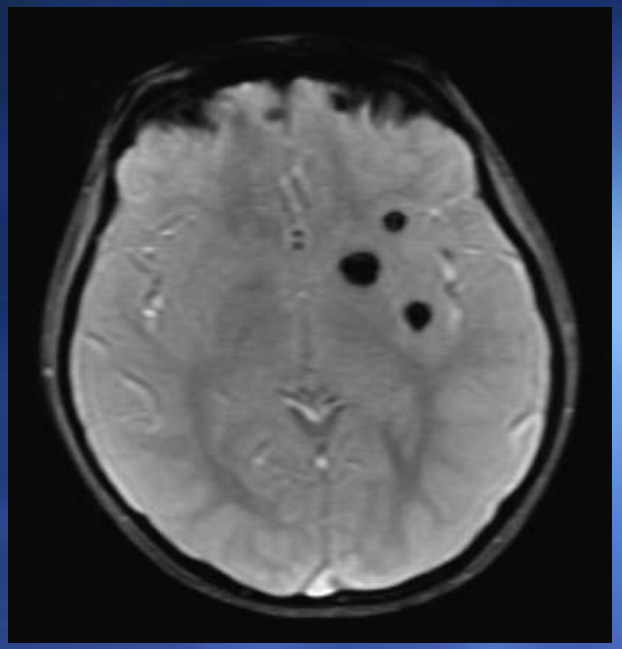
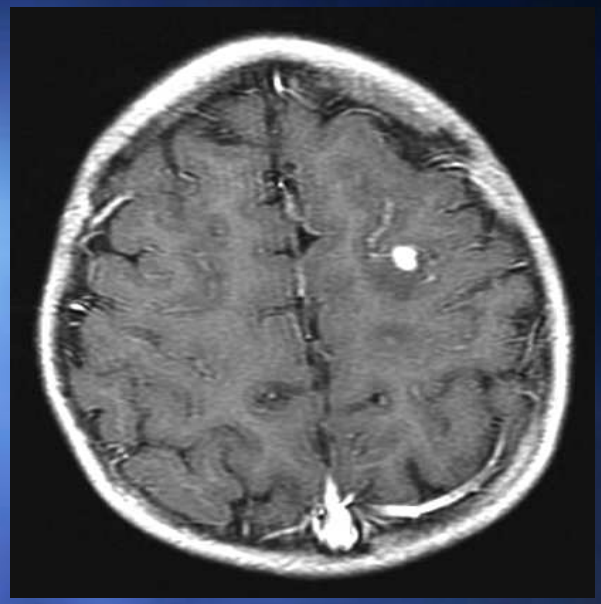
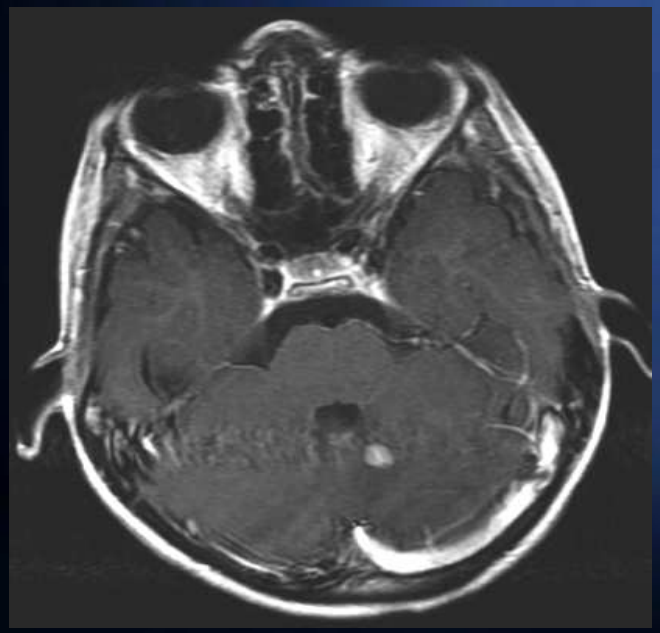
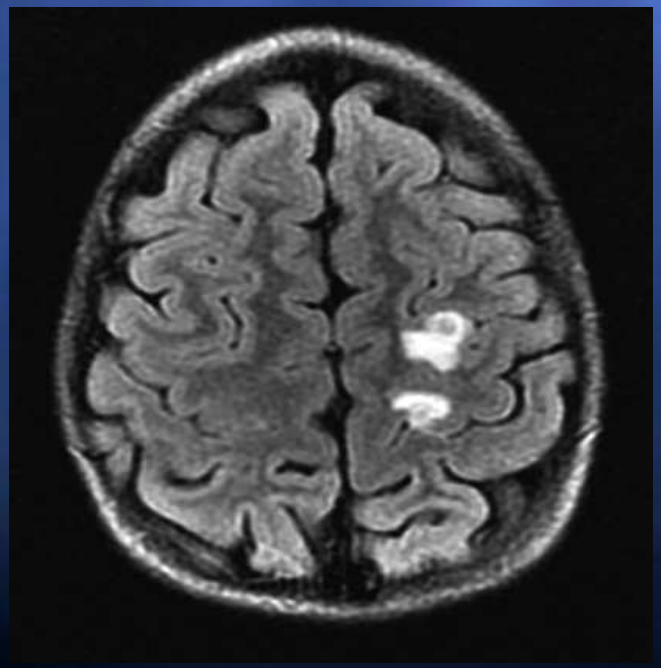
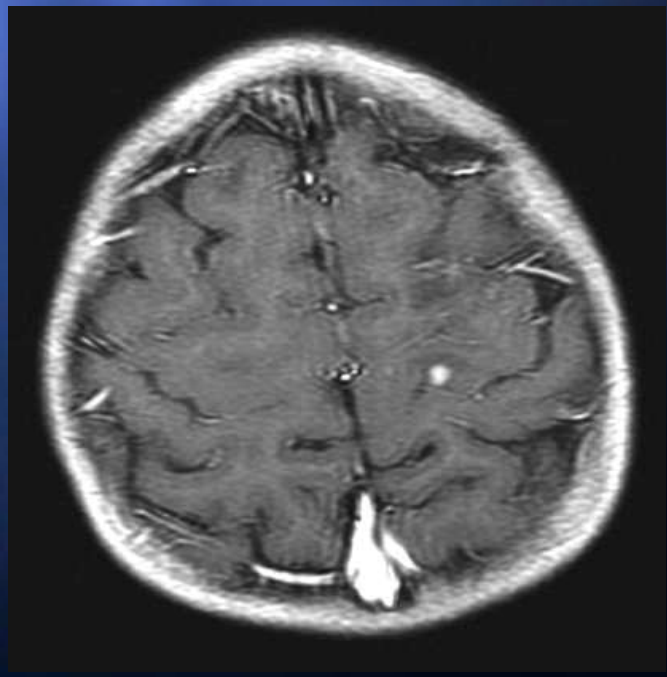
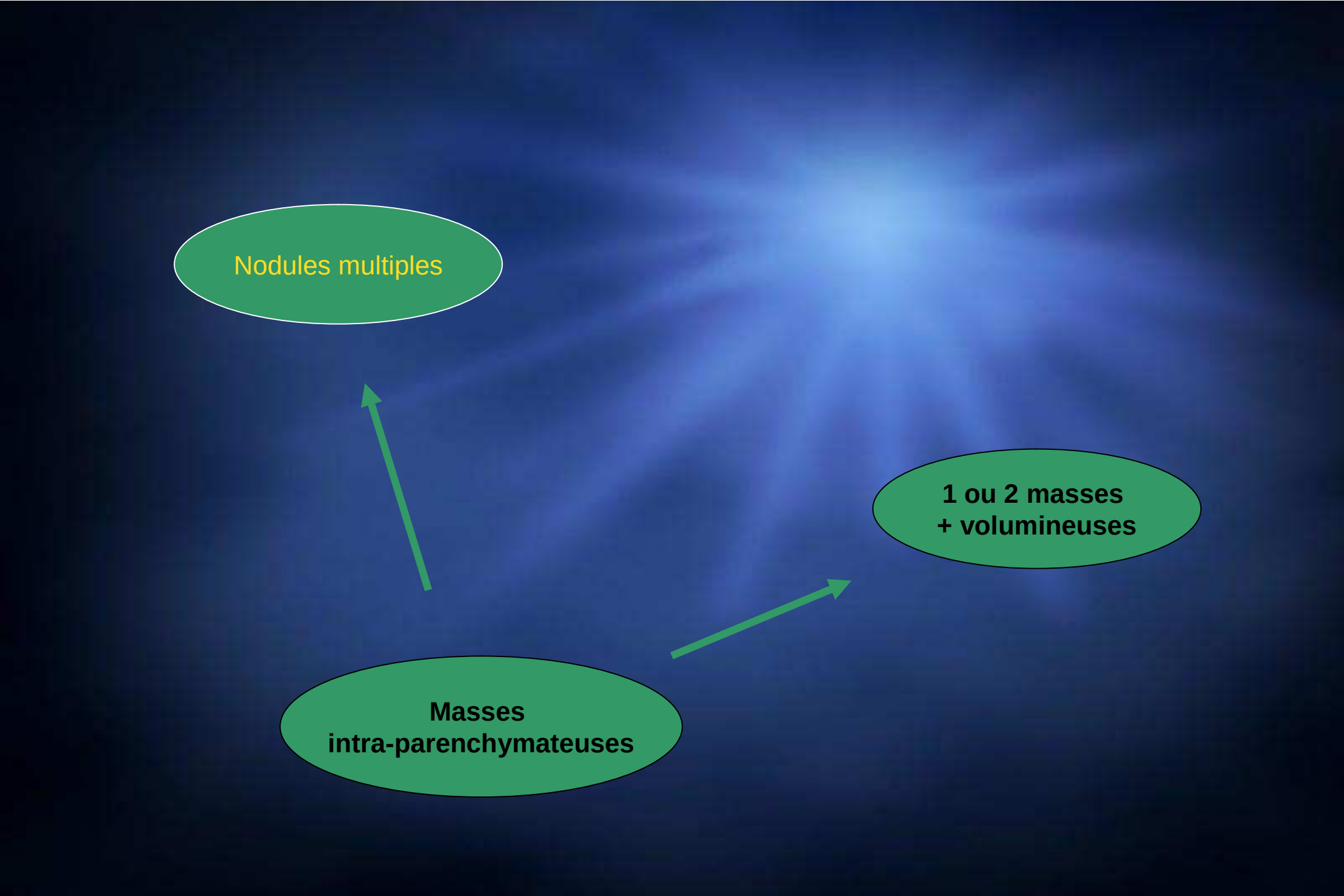
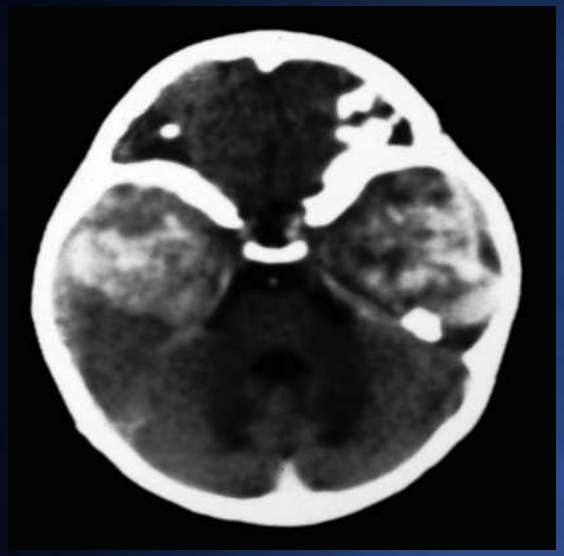
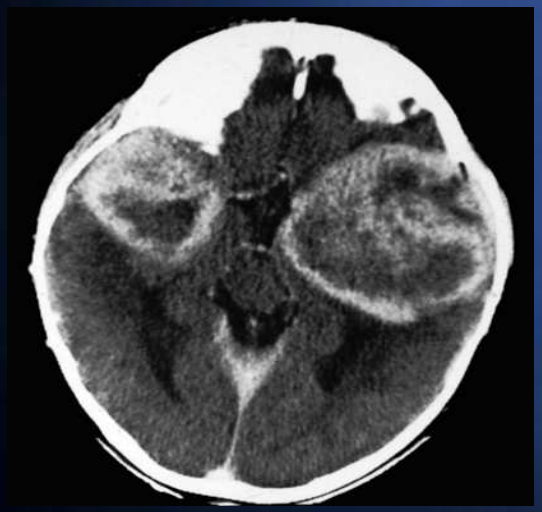




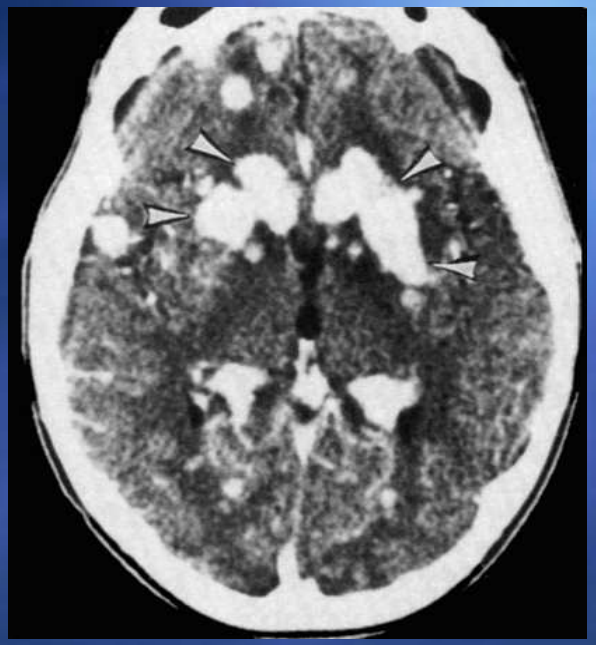







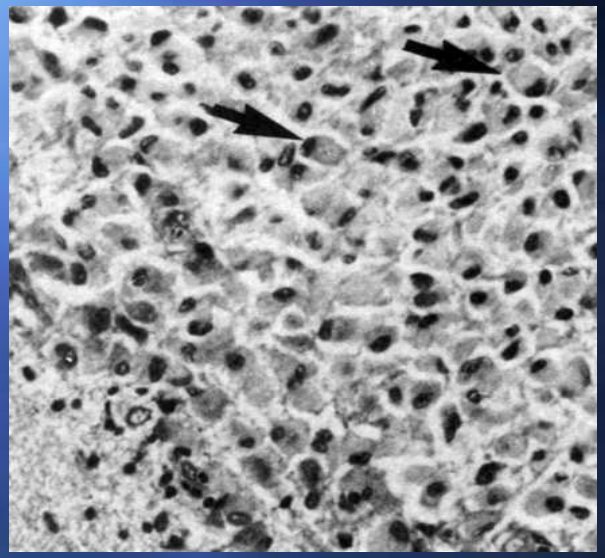

Nodules multiples
1 ou 2 masses
+ volumineuses
Masses
intra-parenchymateuses





Infiltration du neuroparenchyme gliotique
par de larges histiocytes





Ragland RL et al. AJNR 1991

Formes pseudo-dégénératives

Fosse Postérieure

Isolés / ou
Association lésionnelle

Atrophie

Hémisphères

Formes pseudo-dégénératives

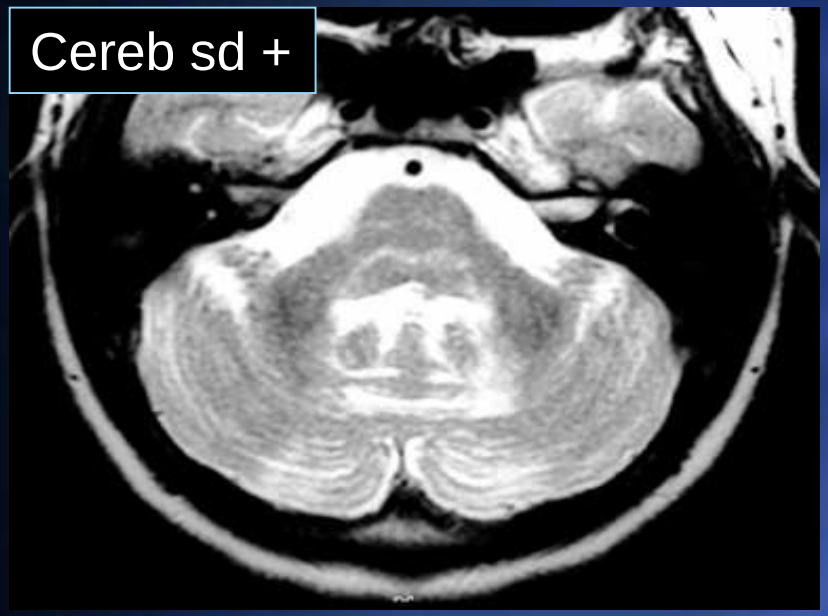
Fosse Postérieure

- *Noyaux dentelés ± SB adjacente > 50 % des cas*
- *Plancher V4 = 70%*
- *Tractus pyramidaux pontiques 20%*

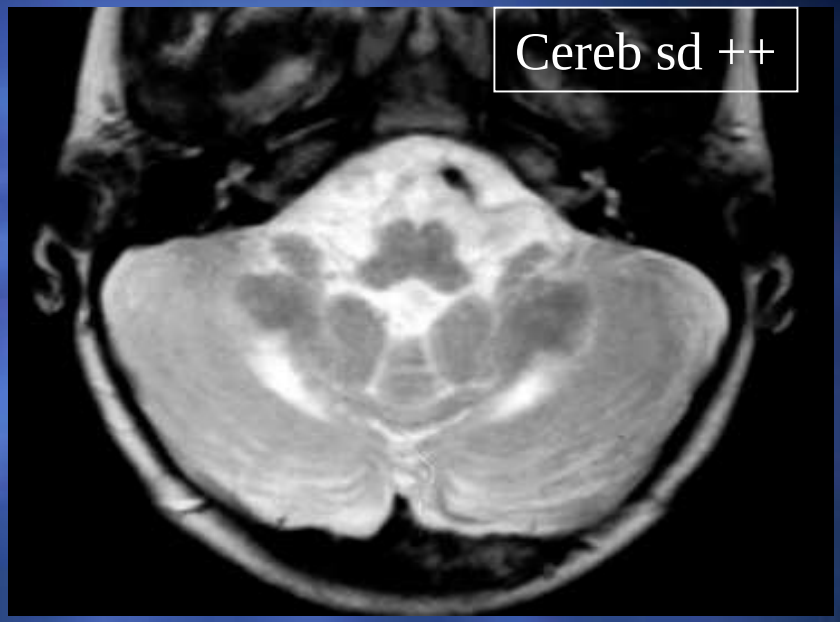
Fosse Postérieure

Noyaux dentelés ± SB adjacente > 50 % des cas

Cereb sd +



Cereb sd ++



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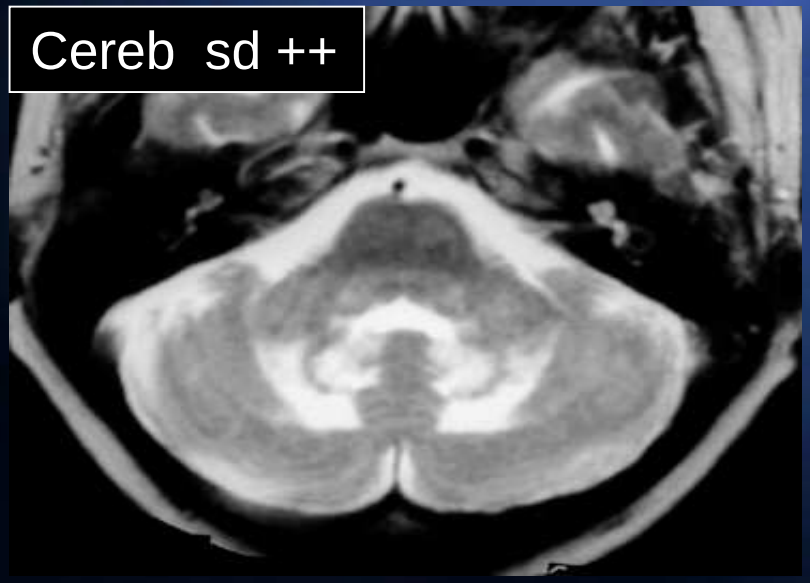
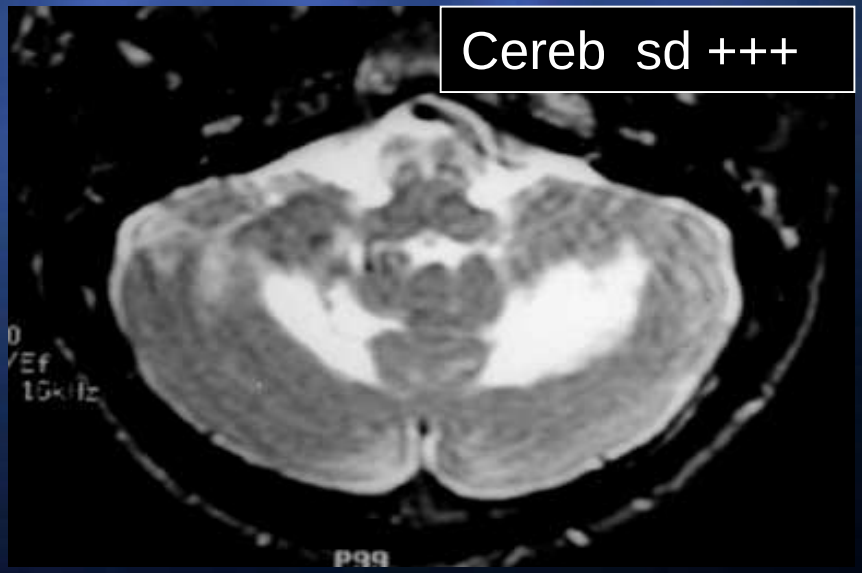
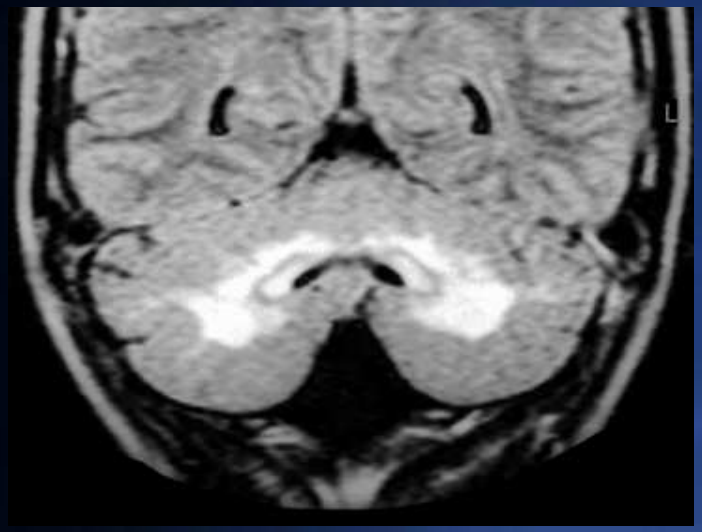
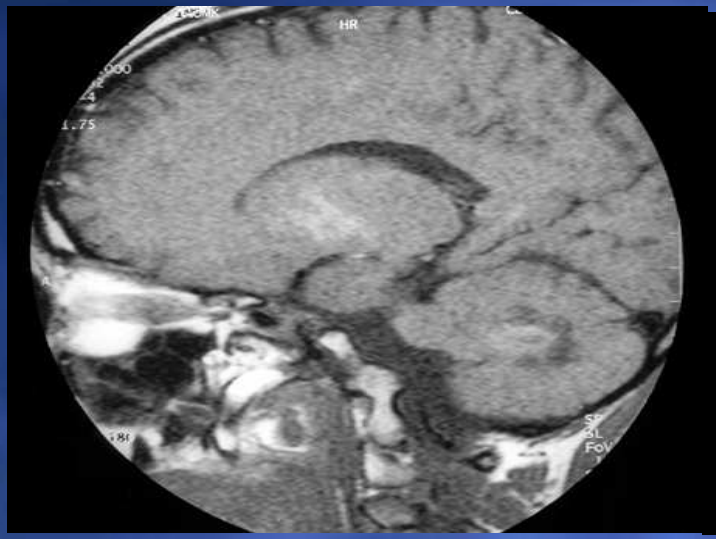


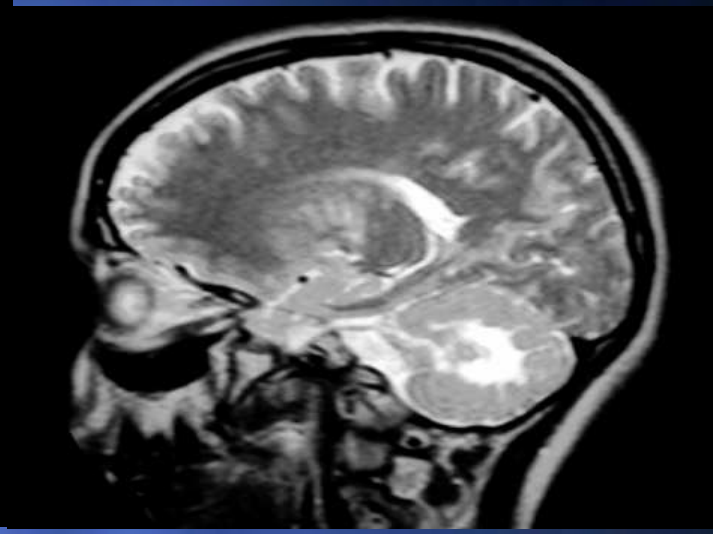
Cereb sd +++

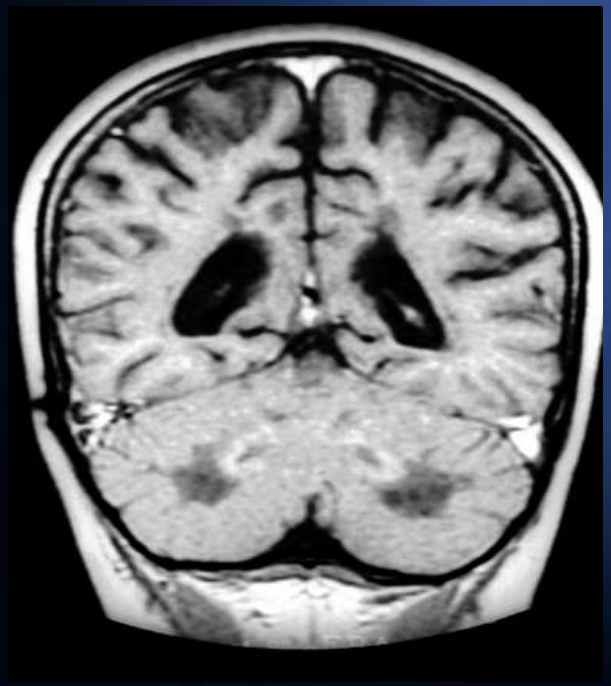


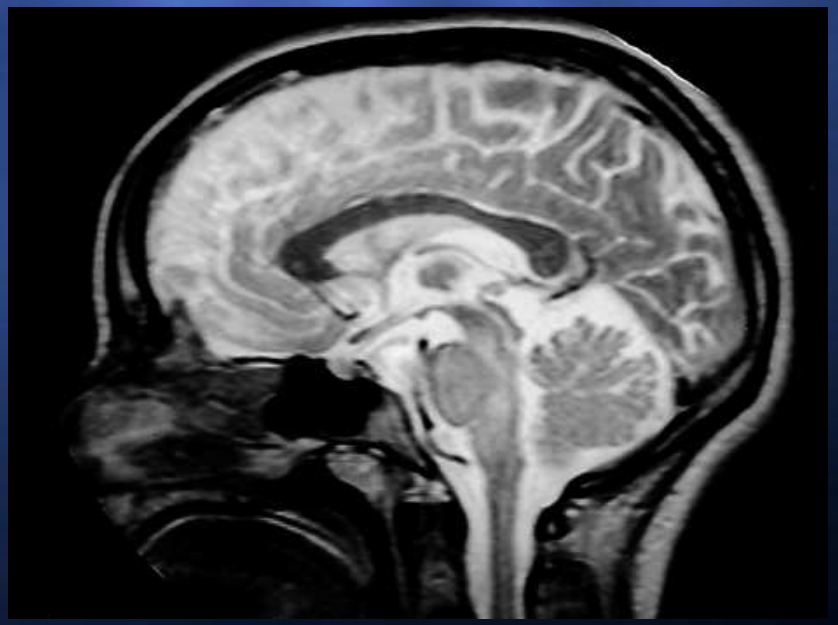
Noyaux dentelés ± SB adjacente > 50 % des cas





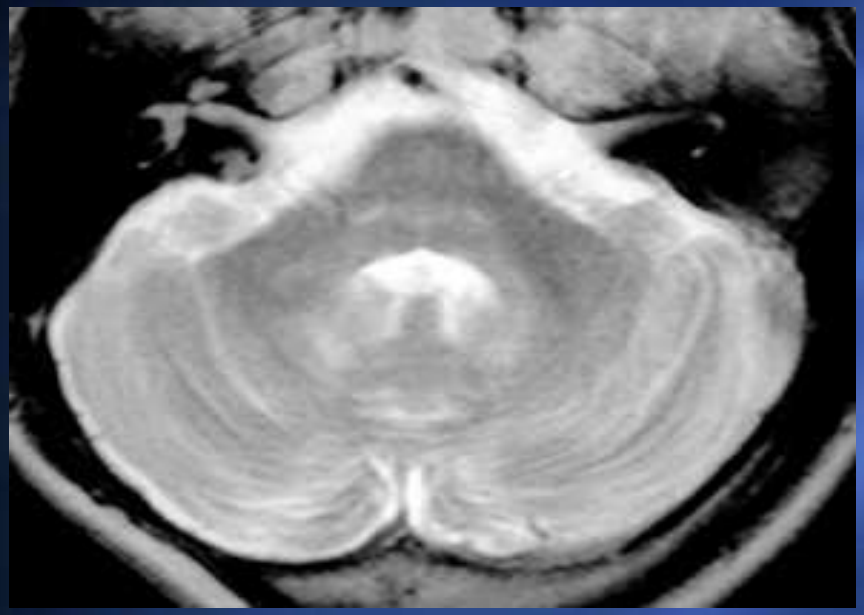


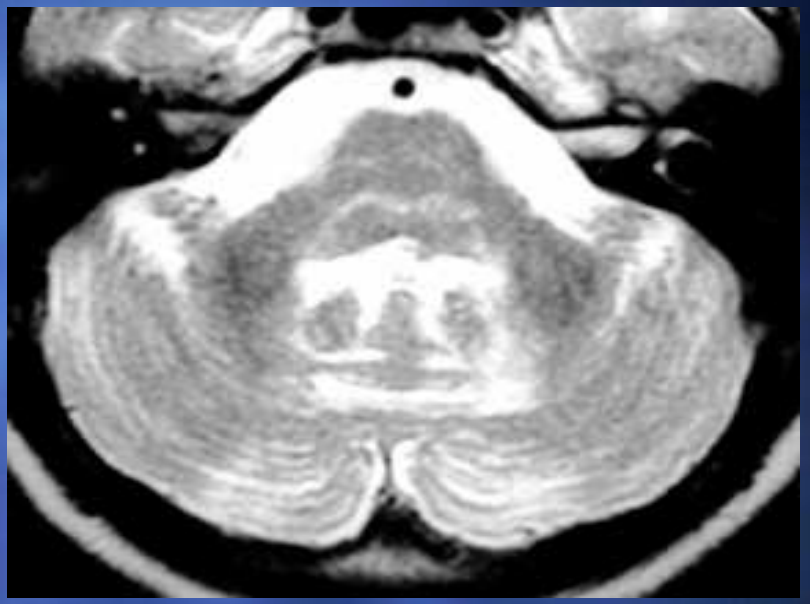


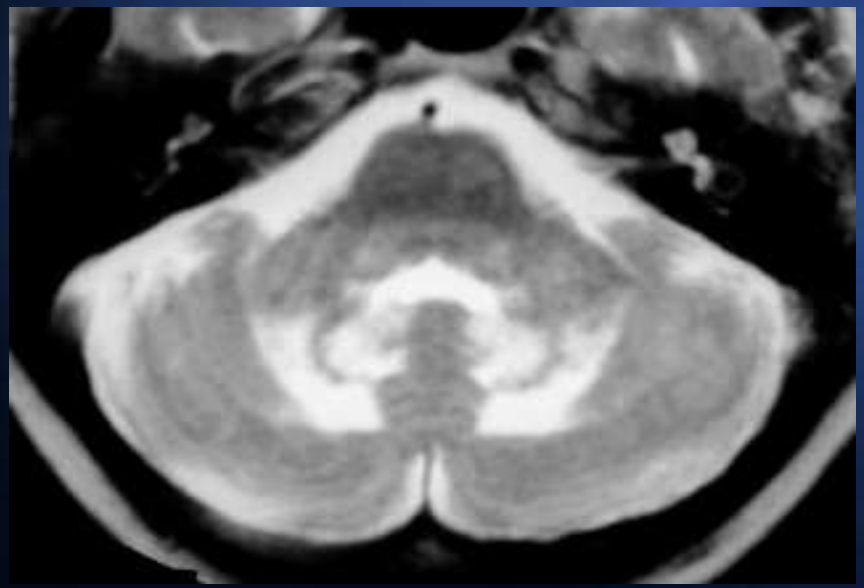


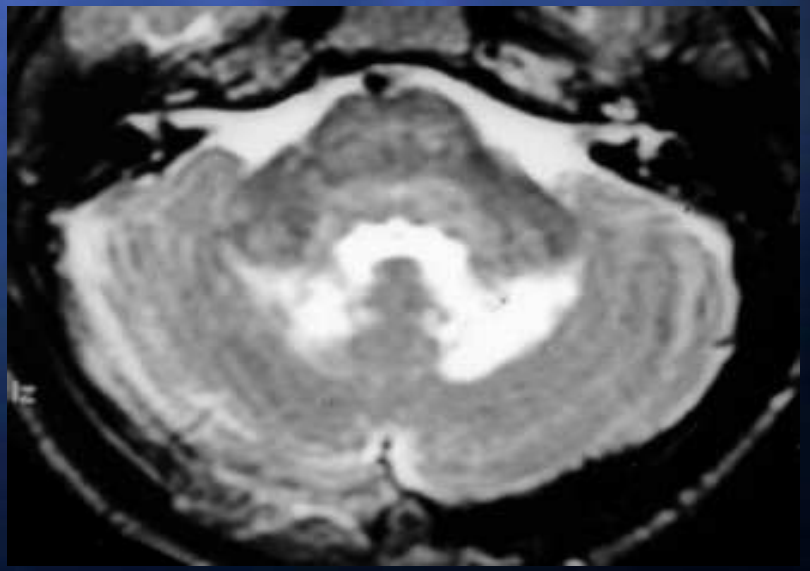
Fosse Postérieure

Hypersignal T2 Plancher V4 adjacent = 70%



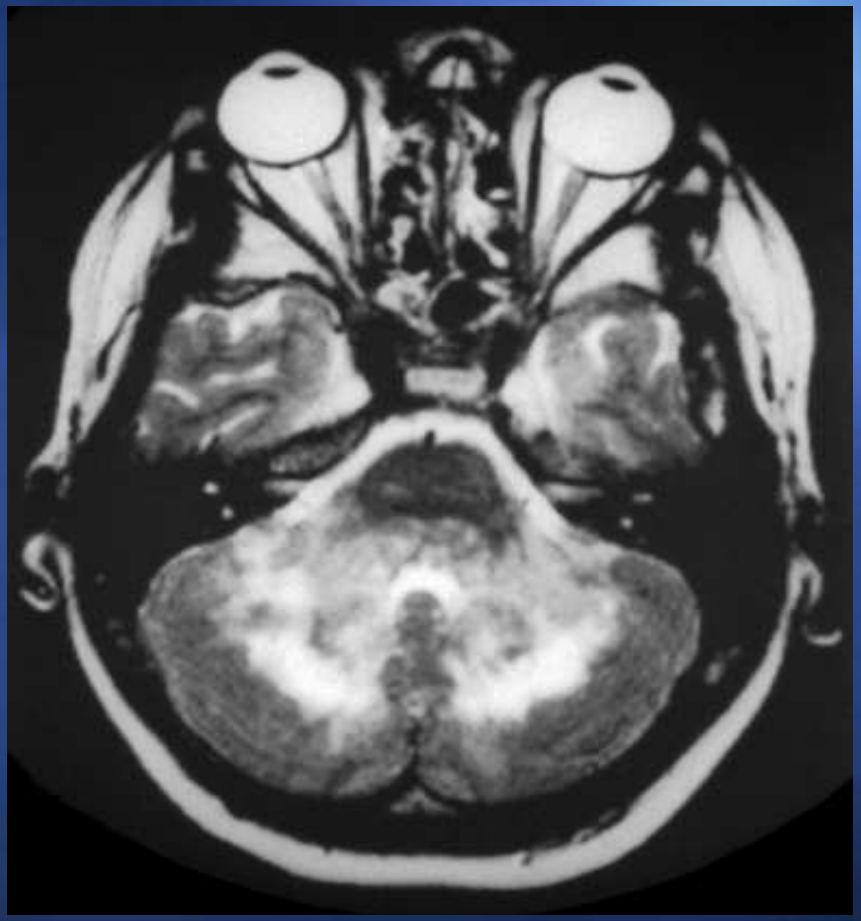






Fosse Postérieure

Hypersignal T2 Plancher V4 adjacent = 70%



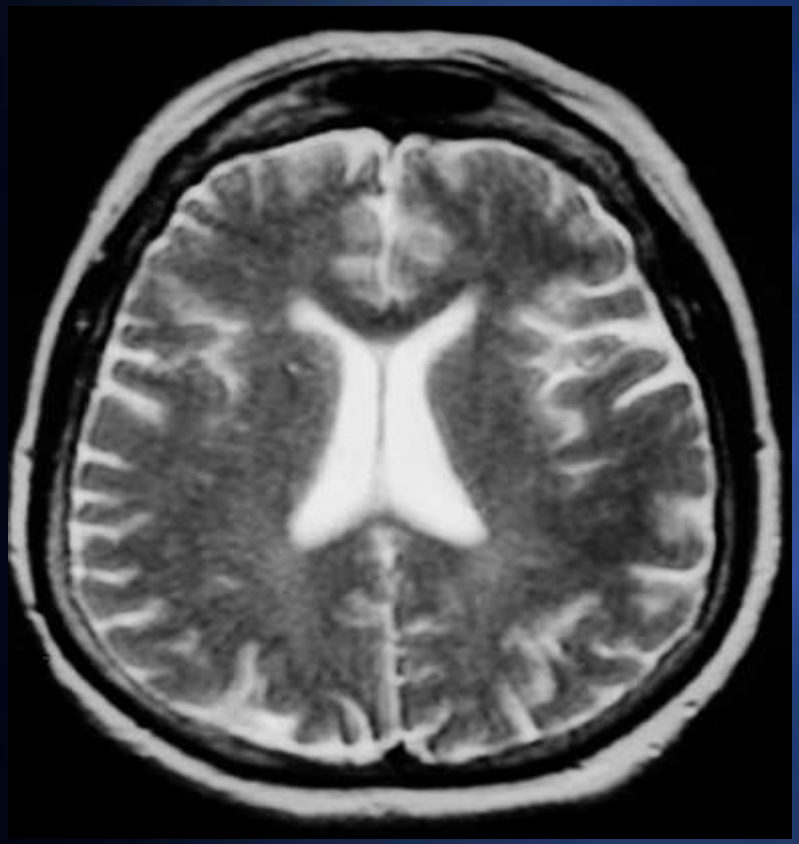
Formes pseudo-dégénératives

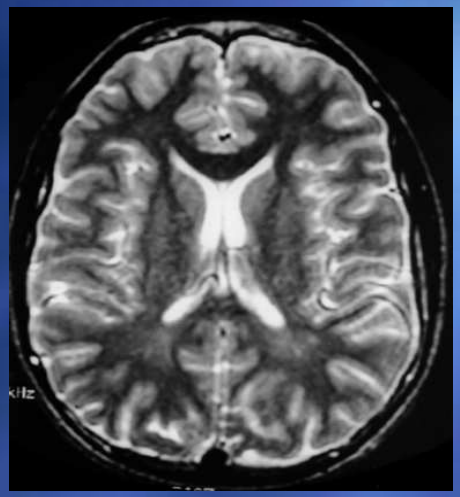
- Hyper T2 Substance blanche cérébrale > 50% des cas
- Atteinte des globes pâles 2/3 des cas

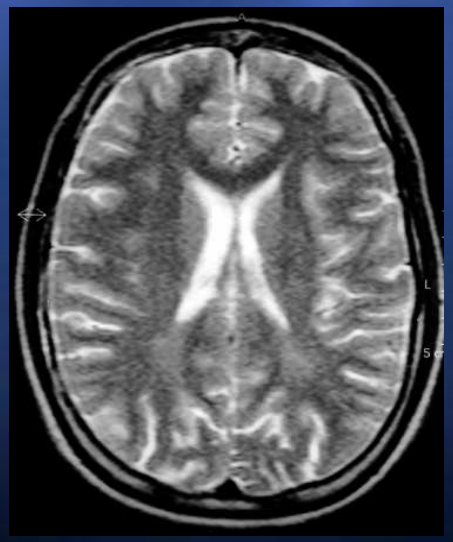
Hémisphères

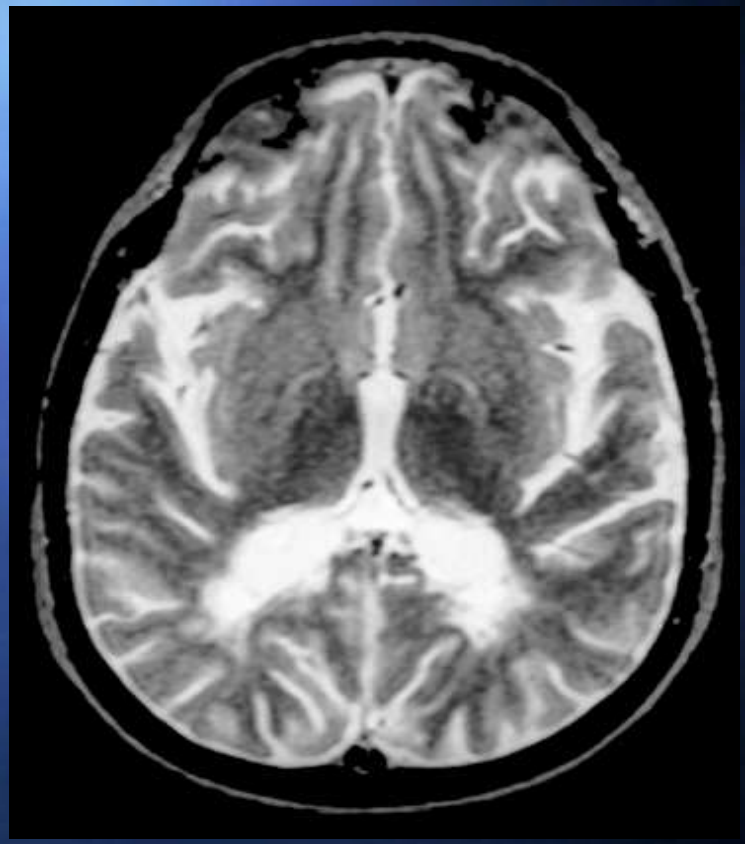
Hémisphères

Zones en hypersignal T2 Substance blanche cérébrale > 50% des cas



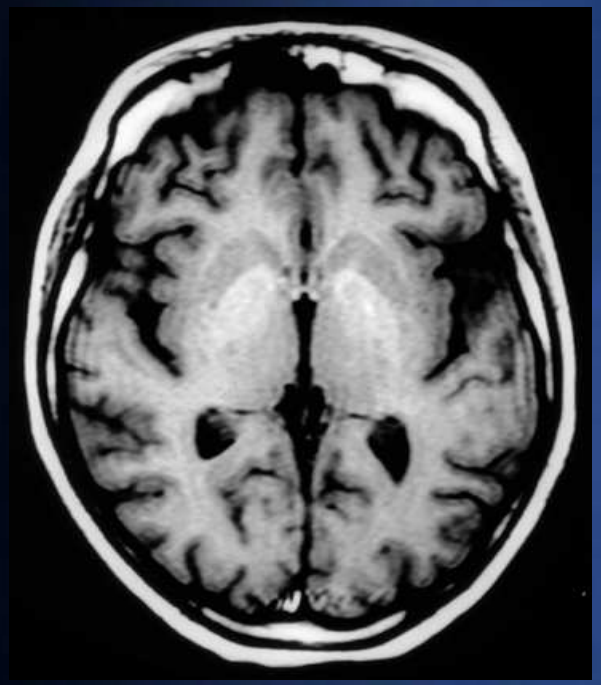


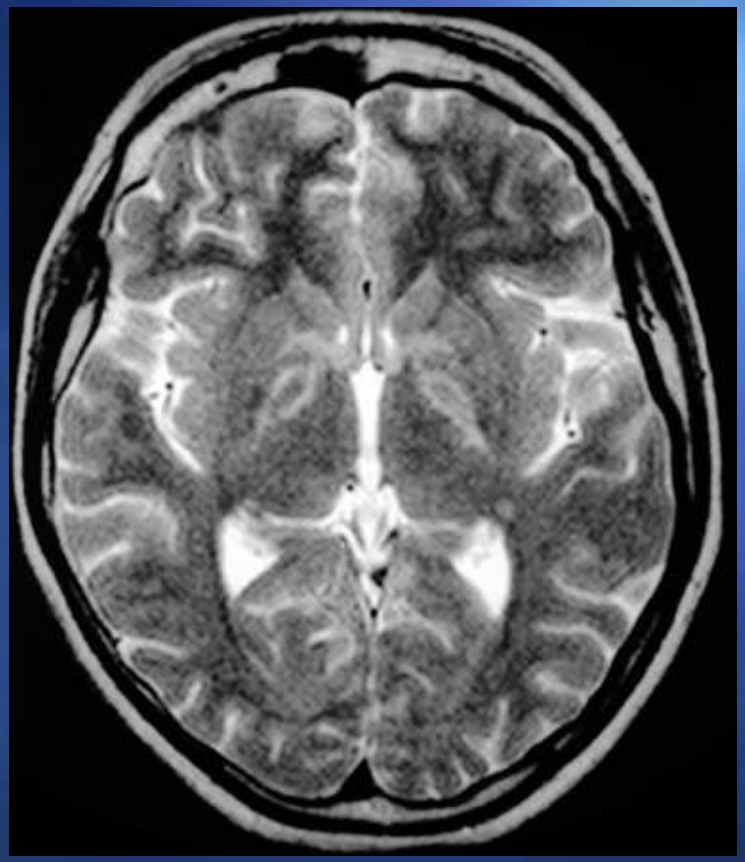




Hémisphères

Hypersignal T1 des globes pâles = 2/3 des cas





Insuffisance hépatique= 0
Nutrition parenterale = 0
Présence de macrophages à contenu lipidique/ radicaux libres

Formes pseudo-dégénératives

- *Atrophie cérébelleuse > 50% des cas*
- *Atrophie sus-tentorielle > ¼ cas*
- *Corps calleux 20%*

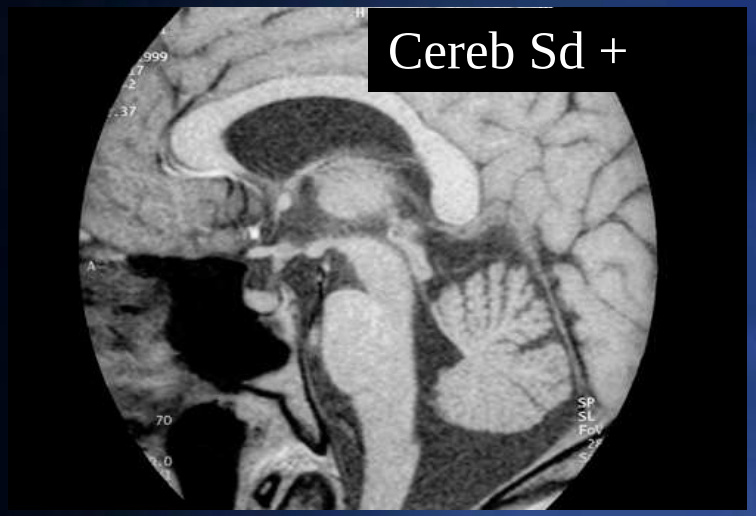
Atrophie

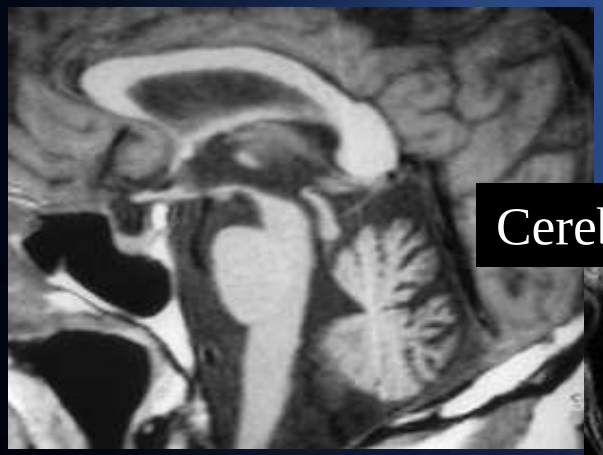
Atrophie

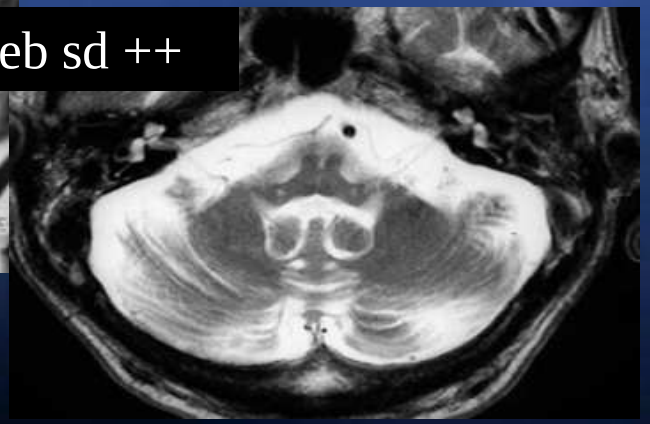
Atrophie Cérébelleuse : 57%

Atrophie sus-tentorielle: 28%

Corps calleux : 21%





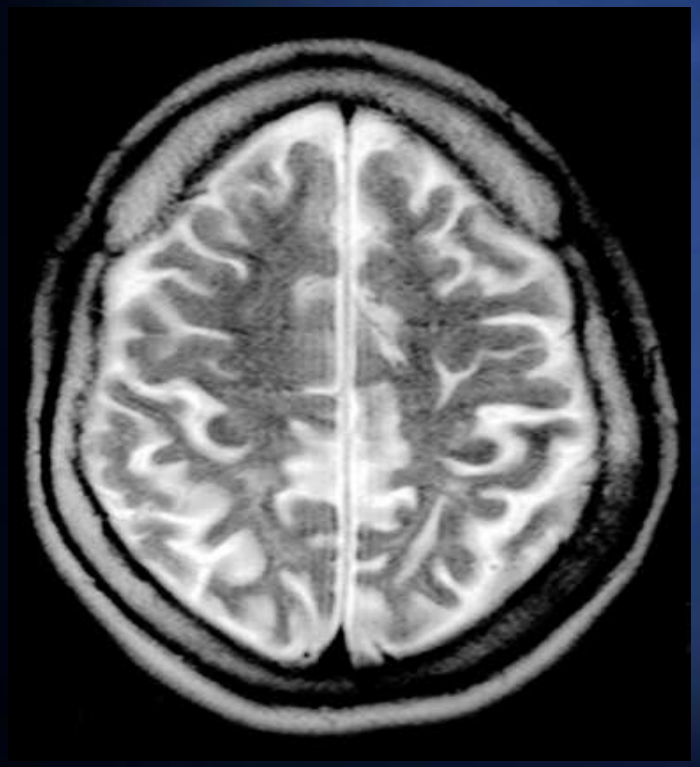


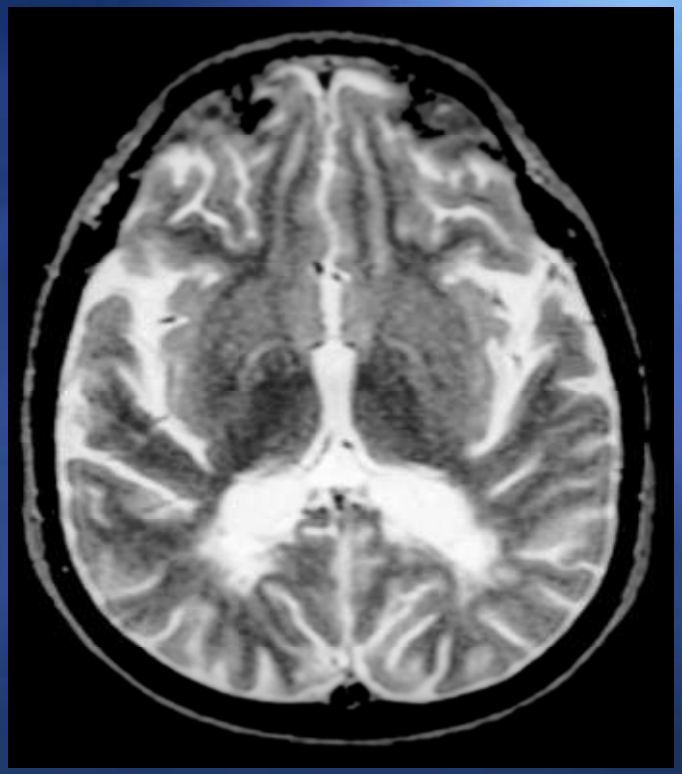
Atrophie

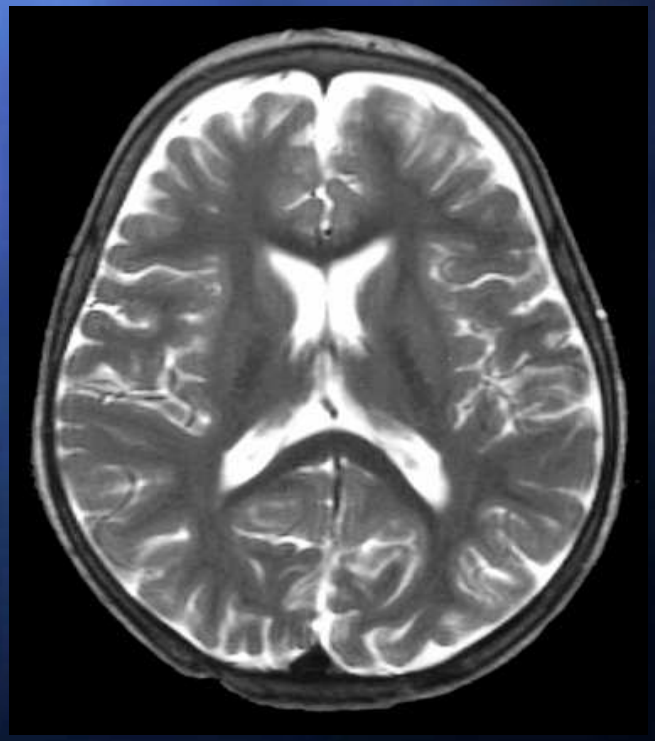
Atrophie Cérébelleuse : **57%**

Atrophie sus-tentorielle: 28%

Corps calleux : 21%







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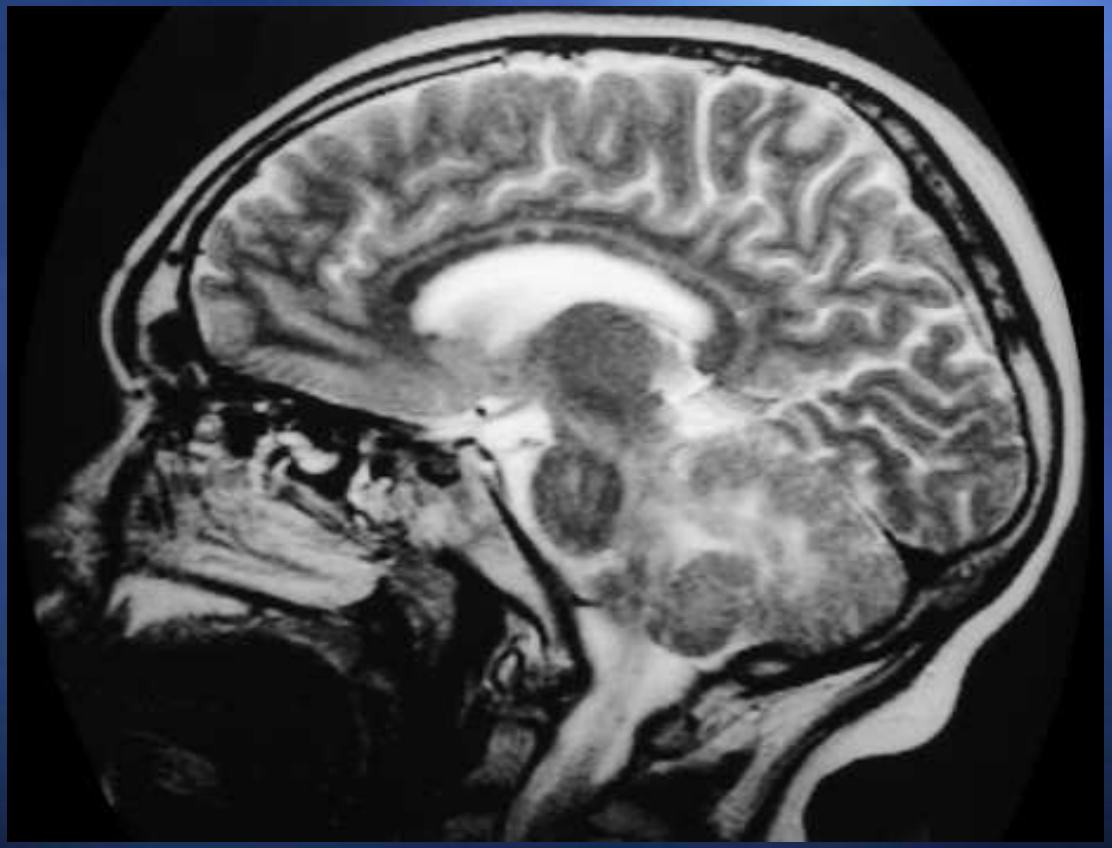

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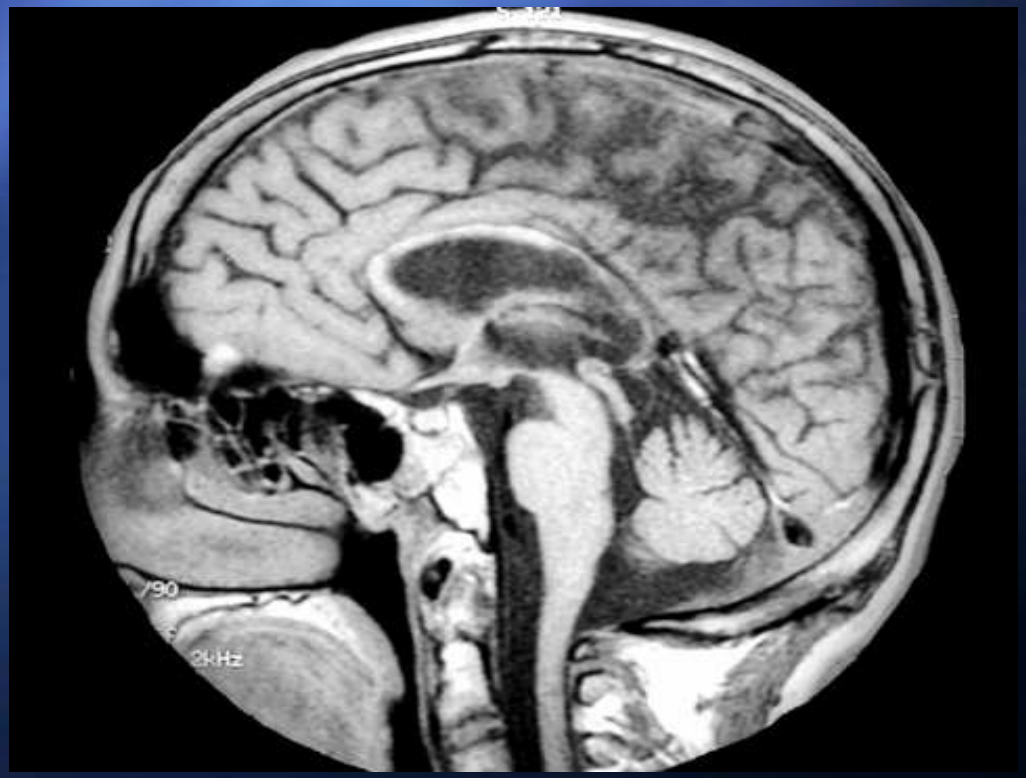
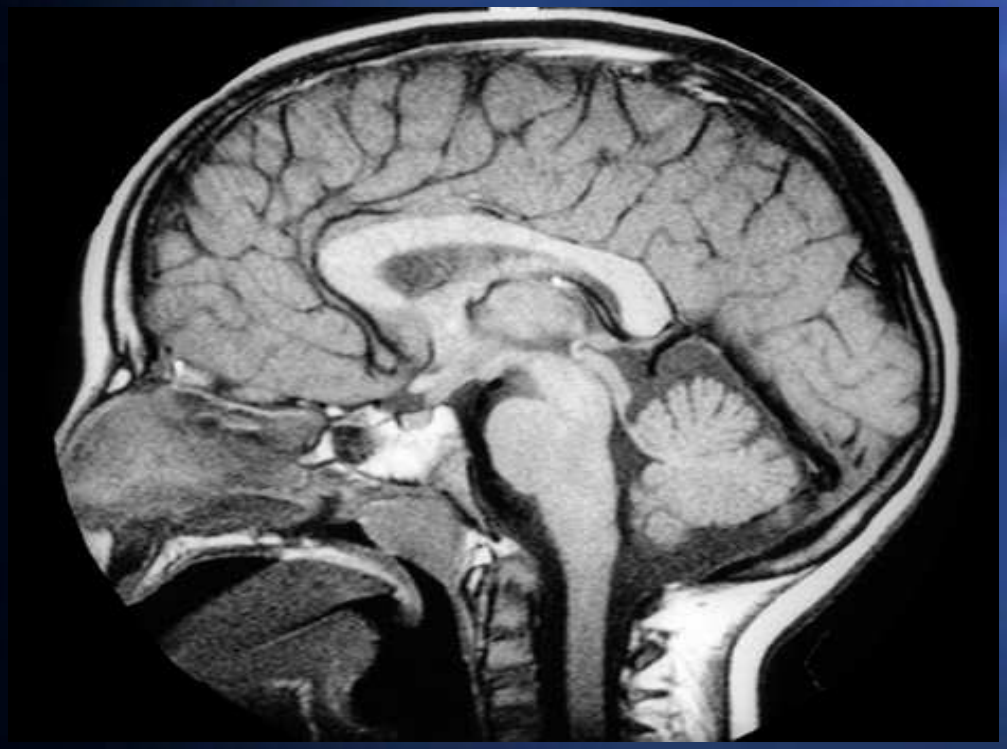
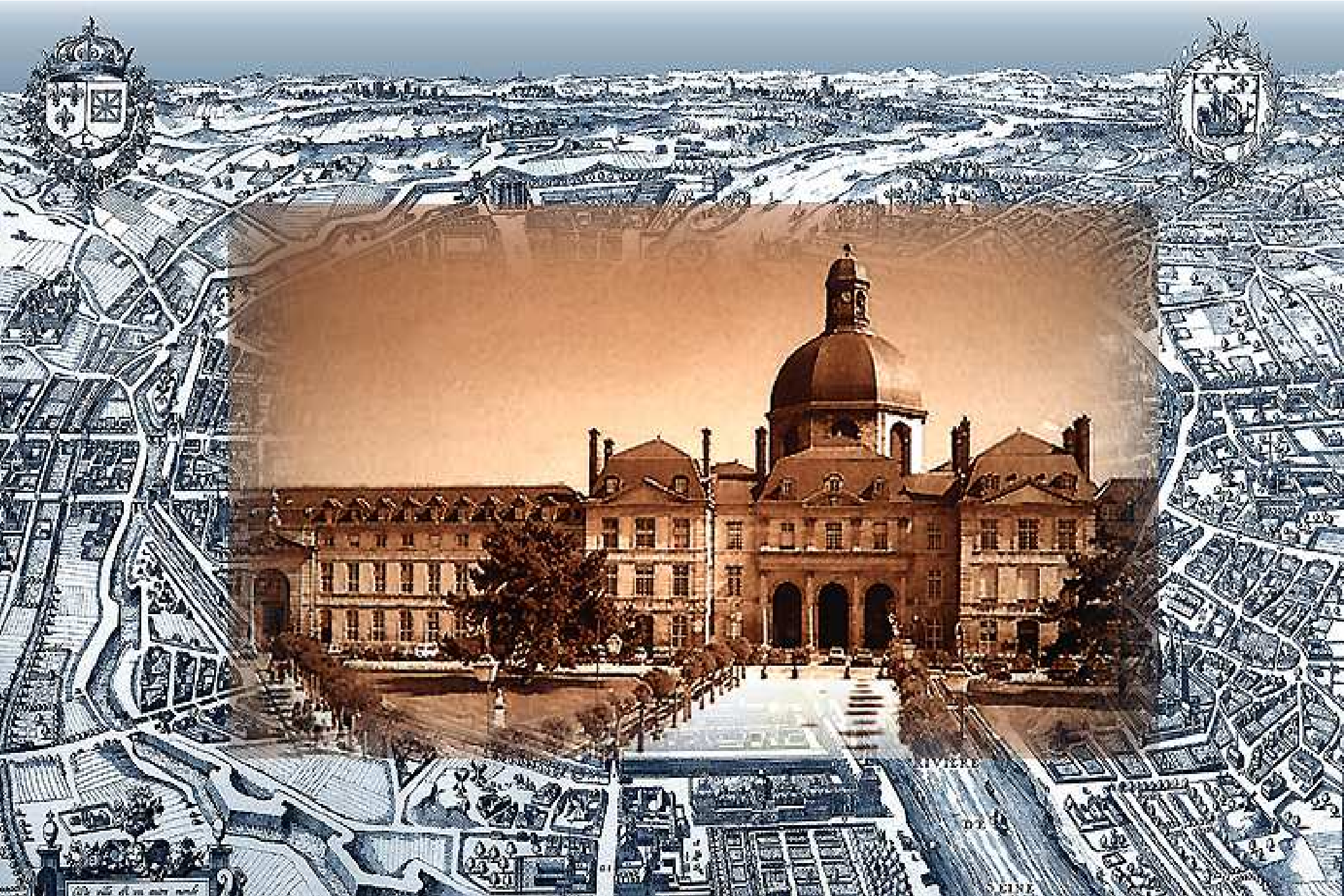


Evolution

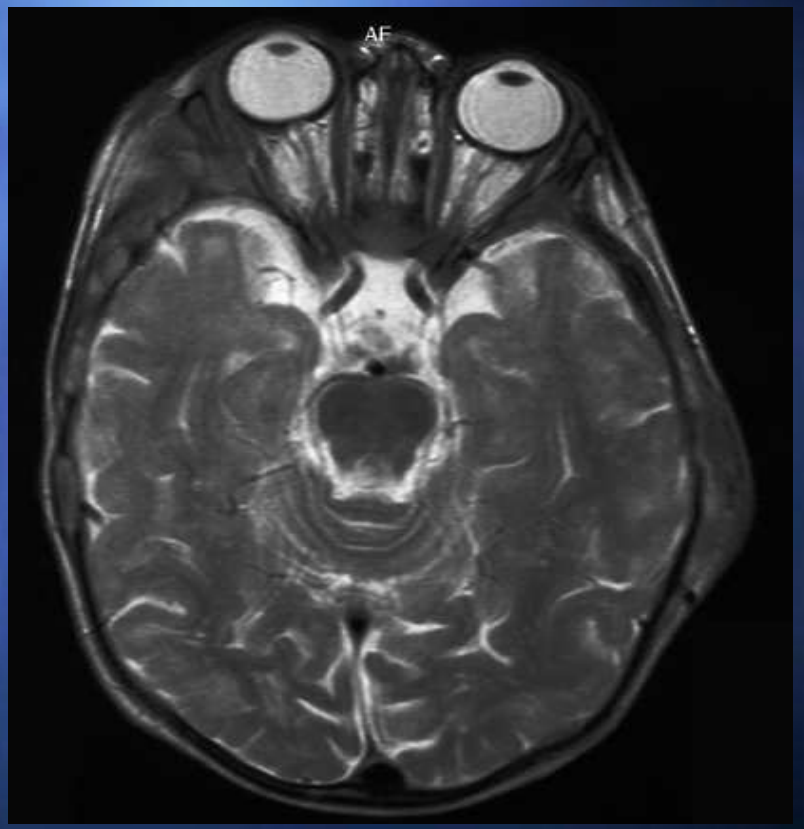
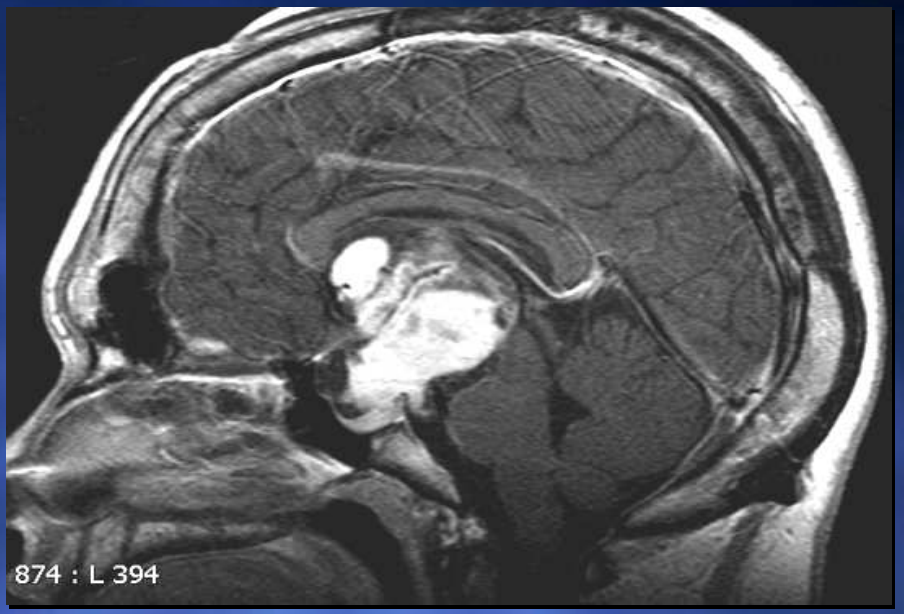


1996

1997


RIVIERE
DE
SEINE

Rajoutée – à garder?



Axe
Hypothalamo-hypophysaire
Dg différentiel