DYSEMBRYOPLASTIC NEUROEPITHELIAL TUMORS: CT, MR FINDINGS AND IMAGING FOLLOW-UP: A STUDY OF 53 CASES

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SUMMARY

Purpose: To evaluate CT and MRI features and long term imaging follow-up of a large series of dysembryoplastic neuroepithelial tumors (DNTs).

Patients and methods: We retrospectively analyzed CT (100 %) and MR imaging (83 %) findings of 53 patients with complex (n = 14), simple (n = 6) or non specific histological forms (n = 33) of DNTs. All patients underwent epilepsy surgery for the treatment of drug resistant partial seizures. Preoperative radiological follow-up from two to 10 years (81 %) and a post-operative follow-up from one to 13 years (92 %) were available.

Results: DNTs are intracortical tumors with no mass effect and no peritumoral edema. An associated deformity of the overlying skull was observed in 44 % of the 34 patients with a cortical lesion of the convexity. We found a contrast enhancement of the lesion in 21 % of cases, a calcic hyperdensity in 36 % of cases and a cystic part in 7.5 % of cases. DNTs were overlying skull was observed in 44 % of the 34 patients with a cortical lesion of the convexity. We found a contrast enhancement of the lesion in 21 % of cases, a calcic hyperdensity in 36 % of cases and a cystic part in 7.5 % of cases. DNTs were hypodense (82 %) on CT examinations and had a decreased signal on the T1 Weighted Images (95 %) and a hypersignal in T2 Weighted Images (100 %) on MR imaging. Eighty-one percent of patients had a mean preoperative radiological follow-up of four years and the tumor was stable in size in all cases; 92 % of patients had a mean post-operative radiological follow-up of 4.5 years and no recurrence was seen.

Conclusion: Three radiological features of DNTs are helpful for the diagnosis: cortical location, absence of mass effect and no surrounding edema. Clinical, radiological and histopathological findings have to be considered together in order to assess the diagnosis and to differentiate DNTs, which are stable lesions from gliomas.

Key words: dysembryoplastic neuroepithelial tumors, brain tumors, epilepsy, MRimaging, CT.

RéSUMÉ

Tumeurs dysembryoplasiques neuro-épithéliales: aspects scanographiques et en IRM (à propos de 53 observations.

Objectif de l'étude: Évaluer l'aspect en imagerie scanographique et IRM et le suivi radiologique au long cours d'une grande série de tumeurs dysembryoneuroépithéliales (DNT).

Patients et méthodes: Nous avons analysé de façon rétrospective les données scanographiques (100 %) et IRM (83 %) chez 53 patients présentant une DNT (14 patients présentaient la forme histologique « complexe », 6 patients la forme « simple » et 33 patients la forme « non spécifique »). Un suivi radiologique de 2 à 10 ans a été disponible dans 81 % des cas et un suivi radiologique post-opératoire de un à 13 ans était disponible chez 92 % des patients.

Résultats: Les DNT sont des lésions corticales sans effet de masse ni « adème » périlésionnel. Une empreinte sur la voûte osseuse est associée dans 44 % des 34 patients présentant une localisation tumoraire au niveau de la convexité. Une prise de contraste est présente dans 21 % des cas, des calcifications dans 36 % des cas et une composante kystique dans 7,5 % des cas. Les lésions sont hypodenses au scanner dans 82 % des cas et en hyposignal T1 (95 % des cas) et hypersignal T2 (100 % des cas) sur les examens IRM. 81 % des patients avaient un suivi radiologique moyen de 4 ans en préopératoire, et la lésion était stable dans tous les cas; 92 % des patients avaient un suivi moyen postopératoire de 4,5 ans et aucune récidive n'a été mise en évidence.

Conclusion: Trois signes radiologiques des DNT sont utiles pour le diagnostic: la localisation corticale, l’absence d’effet de masse et l’absence d’adème périlésionnel. Étant donné le polymorphisme de ces lésions, les données cliniques, radiologiques et anatomopathologiques sont à prendre en considération de façon globale, afin de poser le diagnostic et de différencier ces lésions non évolutives des gliomes ordinaires.

Mots-clés: tumeurs dysembryoneuroépithéliales, tumeurs du cerveau, épilepsie, IRM, TDM.

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Dysplasias of the cortex; 3. Glial nodules. Reproduced with permission from Dysplasie corticale; 3. Nodules gliaux. Figure reproduite de : « complexe » des DNT : 1. Élément glioneuronal spécifique ; 2. Dyplasie corticale ; 3. Nodules gliaux. Figure reproduite de : Neurosurgery 1988 ; 23 (5) ; 545-546, fig. 8.

The first morphologic variant of DNTs described by C. Daumas Dupert [1] in 1988, has been included in the revised World Health Organization histological classification of brain tumors in the category of « neuronal and mixed neuronoglial tumors » [2, 3, 6].

This first histological form of DNTs, referred to as the « complex » form, exhibits specific histological features that clearly distinguish them from ordinary gliomas ; a multinodular architecture, a specific glioneuronal element and foci of cortical dysplasia (figure 1). The specific glioneuronal element shows a columnar structure made up of bundles of axons lined by small tumoral oligodendrocytes (figure 2). Nodules may resemble astrocytomas, oligodendrogliomas or oligoastrocytomas (figure 3).

The other two histological forms of DNTs are the « simple » form [4, 6], which is composed only of the glioneuronal element, and the « non specific » form [6, 7], which does not show the specific glioneuronal element or a multinodular architecture. The « non specific » form may resemble any of the conventional forms of glioma.

Daumas Dupert [1, 2] postulated that DNTs have a dysembryoplastic origin, because of the presence of foci of cortical dysplasia, the participation of multiple and distinct cell lineages, the young age of patients at onset of symptoms and the presence on imaging studies of skull deformity adjacent to the tumor. Supratentorial cortical DNTs may derive from the external granular layer of the cortex, which is composed of primitive neuroepithelial cells that spread rapidly and cover the entire convexity of the cortex.

The aim of our study was to assess, in a large series of DNTs (n = 53), the radiological appearance and imaging follow-up on CT and MRI studies of these tumors according to their histological form (« complex » form n = 14, « simple » form n = 6, « non specific » form n = 33).

PATIENTS AND METHODS

The 53 patients in this study included all patients who underwent corticectomies for drug resistant partial seizures in our center between 1980 and 1995, and had had a histological diagnosis of DNTs. No more patients were recruited after 1995, in order to have a sufficient post-operative follow-up for all the patients. We excluded from our study patients without an accurate radiological examination (i.e. incomplete radiological examinations, or CT scans that used old generation technology and therefore could not demonstrate the tumor features). In order to exclude any confusion of nonspecific histological form of DNTs with gangliogliomas, we excluded from our study any tumor with lymphocytic infiltrates, which are typically absent in DNTs, but nearly always present in gangliogliomas (see discussion). About 60 % of the patients in our study (n = 32) were included in previous studies by Daumas Dupert et al. [1, 4, 7] in which the three histological forms of DNTs were first described.

All 53 patients, 13 female and 40 male, suffered from partial seizures, and their age at onset of symptoms ranged from one to 22 years (mean : 11.6 years) in 92.5 % of cases. In four cases the age at onset ranged from 34 to 40 years. All patients had partial seizures, 24 patients also had secondarily generalized seizures and none presented primary generalized seizures. Seizure duration at the time of surgery ranged from one to 37 years (mean 11.4 years). Epilepsy was disabling and drug resistant in all cases.

The interictal neurological examination was normal in 47 patients (88.6 %), one patient had a mild, stable sensitive deficit, another had a mild facial paresis, and a quadranopsia was disclosed in four patients. In all cases the neurological deficit was ignored by the patients and their families.

All patients had undergone corticectomies according to the Talairach technique, and surgical samples consisted of five to 20 fragments. 47 patients (88.6 %) had a total resection of the lesion and 6 had a partial surgery. Histologically, the series comprised 14 « complex » form of DNTs, 6 « simple » form, and 33 « non specific » forms. Histologically, the « non specific » histological forms resembled astrocytomas in seven cases (21.2 %), oligodendrogliomas in 15 cases (45.4 %) and mixed oligo-astrocytomas in 11 cases (33.3 %).

None of the patients had received radiation or chemotherapy.
All CT and MR scans were retrospectively reviewed by two of us (RS, JFM). Preoperative radiological examinations were performed at various institutions from 1980 to 1995. Preoperative CT examinations were available in all patients, 44 (83%) of whom also had preoperative MRI examinations. In 45 cases preoperative CT studies were obtained before and after intravenous administration of contrast medium. Eight patients had only an unenhanced CT examination, but three of them had an enhanced MRI examination. MRI scans included T1 Weighted Images, proton density WI and T2 Weighted Images in at least one plane. T1 Weighted Images after gadolinium intravenous administration were obtained in 18 patients (table I).

Forty-three patients (81%) had clinical and radiological preoperative follow-up data of two to 10 years (mean 4 years). Two or more CT or MR studies were obtained over this preoperative period. In the other 10 patients the preoperative radiological follow-up was of only one year.

**Radiological examinations**

All patients had immediate post-operative imaging: CT (n = 17) or MRI (n = 36) examination. In addition, long-term post-operative radiological follow-up of one to 13 years (mean 4.5 years) was available in 49 patients (92%).

**Studied parameters**

The following radiological criteria were evaluated as follows:
- **Location**: according to the predominant lobar repartition of the tumor: frontal, parietal, occipital, mesial-temporal and external-temporal.
- **Topography**: cortex and/or white matter involvement.
- **Size**: this corresponds to the maximal length of the lesion on CT or MRI scans.
- **Shape**: the shape of the lesion was defined as nodular (more or less spherical), or megagyri-like (impression of thickened cortex). We also evaluated the limits of the lesion: sharp or blurred.
- **Mass effect**: was defined as a displacement of normally adjacent structures.

**TABLE I. — Neuroradiological examinations in the 53 patients of the study.**

<table>
<thead>
<tr>
<th></th>
<th>« Complex » form</th>
<th>« Simple » form</th>
<th>« Non specific » form</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>No. of patients</strong></td>
<td>14</td>
<td>6</td>
<td>33</td>
</tr>
<tr>
<td><strong>CT studies</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Without contrast administration</td>
<td>n = 14</td>
<td>n = 6</td>
<td>n = 33</td>
</tr>
<tr>
<td>With contrast administration</td>
<td>n = 12 (86%)</td>
<td>n = 6 (100%)</td>
<td>n = 27 (82%)</td>
</tr>
<tr>
<td><strong>MR Examination</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Without contrast administration</td>
<td>n = 9 (64%)</td>
<td>n = 6 (100%)</td>
<td>n = 29 (88%)</td>
</tr>
<tr>
<td>With contrast administration</td>
<td>n = 4 (45%)</td>
<td>n = 3 (50%)</td>
<td>n = 19 (66%)</td>
</tr>
<tr>
<td></td>
<td>n = 5 (55%)</td>
<td>n = 3 (50%)</td>
<td>n = 10 (34%)</td>
</tr>
</tbody>
</table>

All patients had immediate post-operative imaging: CT (n = 17) or MRI (n = 36) examination. In addition, long-term post-operative radiological follow-up of one to 13 years (mean 4.5 years) was available in 49 patients (92%).
— A ventricular print of the lesion or a bone deformity adjacent to the lesion: were evaluated separately and were not considered as a mass effect.

— Contrast enhancement: contrast enhancement was evaluated on immediate post-contrast administration scans. CT (n = 43) and MRI (n = 11) were evaluated separately. We also analyzed the shape of the contrast enhancement.

— Other components of the tumor: calcic components were assessed on non-enhanced CT scans. Cyst was defined as a well-delineated liquid spherical component. We differentiated between macro-cysts (size superior to 1 cm) and micro-cysts (size less to 1 cm).

— Peritumoral edema: was defined as « digit like » peritumoral T2 hypersignal of the white matter on MRI examinations, or as hypodense « digit like » surrounding abnormalities on CT scans.

Imaging follow-up

Pre surgical evolution of the tumor was assessed according to the following changes: the size of the tumor, newly appearing contrast enhancement, calcifications or cystic formation.

The post-operative out come was assessed on last imaging control as follows: recurrence or no recurrence of the lesion after complete resection, stability or increased size of the lesion after partial resection.

RESULTS

Radiological features

Forty-four patients had both CT and MRI examinations, and nine patients had only CT scan exami-
The tumor appeared isointense on T1 Weighted Images in nine patients (20 %), with a homogeneous, pseudocystic appearance in 41 patients (91 %), and composed only of calcic components. CT scans disclosed a calcic hyperdensity in 19 cases (36 %), 12 of which had small punctuated foci of calcifications. The margins between the tumor and normal parenchyma were sharp on T2 Weighted Images in 42 of the 44 patients with MRI examinations. None of the lesions, including large tumors, showed a mass effect. A print of the tumor on the lateral ventricle was found in five cases (9.4 %), but in no case the ventricle was displaced.

A deformity of the overlying skull was apparent in 15 (44 %) of the 34 patients with a cortical lesion of the convexity (figure 4). In addition, an enlargement of the temporal fossa was observed in three (10 %) of the 34 patients with a temporal tumor.

Focal tumoral enhancement was observed in 10 (21 %) of the 48 patients with enhanced CT or MRI examinations. Contrast enhancement exhibited a nodular pattern in five cases (figure 5), was ring shaped in two cases (figure 6) and showed a heterogeneous shape in three cases. In one patient with both CT and MRI examinations, contrast enhancement was observed on MRI but not on CT scan.

On plain CT examinations the tumor had a hypodense appearance in 43 of the 53 patients (81 %). The hypodensity was homogeneous in 20 cases (38 %) and heterogeneous in 23 cases (43 %). In four cases (7.5 %) the lesion appeared hyperdense and composed only of calcic components. CT scans disclosed a calcic hyperdensity in 19 cases (36 %), 12 of which had nodular calcifications (figure 4), and seven of which had small punctuated foci of calcifications. These calcifications were observed either alone or in association with a hypo- and/or isodense component.

Four of the 53 lesions (7.5 %) showed a microcystic component, without any enhancement of the cyst wall.

On MR imaging the tumor had decreased signal intensity on T1 Weighted Images (figure 7) and an increased intensity signal on T2 Weighted Images in 41 patients (91 %), with a homogeneous, pseudocystic appearance in nine patients (20 %) (figure 8). The tumor appeared isointense on T1 Weighted Images and hyperintense on T2 Weighted Images in three cases.

In no case, even in lesions with a contrast enhancement, did we find the pattern of « digit like » peritumoral T2 hypersignal of the white matter suggestive of surrounding edema.

All tumors involved the cortical ribbon, which was usually markedly thickened. In 12 cases signal abnormalities also extended into the sub cortical white matter (figure 9).

It is noteworthy that, in patients who had both CT and MRI examinations (n = 44), MRI demonstrated more clearly the cortical location and radiological features of the tumor than the CT scans. In six patients, CT examinations were normal whereas MRI revealed signal abnormalities. Small lesions were better demonstrated by MRI examinations. The margins between the tumor and normal tissue were better defined on T2 Weighted Images than on CT examinations.

Imaging features according to the histologically forms DNTs (table II)

Histologically « simple » forms of DNTs (n = 6) showed a pseudo-cystic, well delineated, homogeneous appearance, with no calcifications or any enhancement after contrast examination (100 % of enhanced CT and/or MR examinations) (figure 5 and figure 2b corresponding histological sample).

Histologically « complex » and « non specific » forms had a variable histopathological and radiological appearance. Histologically nodules may resemble oligodendroglomas or astrocytomas or mixed gliomas. In these histological forms, nodular enhancement (figure 5 and figure 3b corresponding histological sample) of a « non specific form » of DNT) or ring-like contrast enhancement (figure 6 and figure 3c corresponding histological sample of a « complex form » of DNT) was observed in 10 patients (21 %). Also, calcifications (figure 4 and figure 3d corresponding histological sample of a « complex form » of DNT) were encountered in 19 patients (40 %). Radiological findings in these patients are summarized in table II.

Preoperative radiological follow-up

Two- to 10-year preoperative imaging follow-up (CT and/or MRI examinations) was available in 43 patients. In all cases tumors remained stable in size. However, in two patients with a histologically « complex form » of DNTs, while the size of the lesion remained stable, sequential imaging demonstrated changes in the shape of contrast enhancement. In these two cases the nodular enhancement became ring-shaped. In a third case of a histologically
« complex » form we observed a newly appeared ring-like enhancement on MR T1 weighted imaging sequences.

Post-operative radiological follow-up

Of the 53 patients in the study 39 had complete surgical removal of the tumor, whereas 14 patients had only partial surgery. Post-operative radiological follow-up (range: 1 to 13 years), available in 49 patients, revealed no radiological evidence of recurrence of lesions in patients who had undergone complete resection. Furthermore, in the 6 patients who had undergone partial surgery, we found no growth of the residual lesions.

DISCUSSION

DNTs are stable lesions in young patients, with no or few radiological changes on pre-operative imaging follow-up and no recurrence after surgery in our series, but histologically they may closely resemble low- or even high-grade astrocytomas, oligodendroglia or mixed oligo-astrocytomas. Their clinical presentation (partial epilepsy beginning before 20 years of age, with no or with mild and stable neurological deficit) together with the radiological features are key criteria for the differential diagnosis from ordinary gliomas. Several neuroradiological studies [8-12] have already been published on DNTs, but included only a limited number of patients (from 3 to 16), and all were based on the classical histological forms of DNTs, i.e. the « complex » and the « simple » forms.

This study, which is based on a series of 53 cases of DNTs diagnosed in patients who underwent epilepsy surgery between 1980 and 1995, is the first radiological study to include the recently described « non specific » histological form of DNTs. Moreover, in this study covering a period of time during which epilepsy surgery was not common practice, 81% of patients had an imaging follow-up (mean: four years) before they were referred for epilepsy surgery. This, and a long post-operative follow-up (mean 4.5 years) in most of the patients (92%), allowed us to confirm that DNTs are carcinologically stable lesions.

In accordance with their histological polymorphism DNTs may show variable appearance on CT or MRI scans, they may exhibit contrast enhancement (21%), and may contain calcifications (36%). Nevertheless, DNTs may have pseudo-cystic appearance in histological « simple forms ». However, there are three constant radiological features, which coupled with the clinical onset, distinguish DNTs from « ordinary gliomas »: cortical location, absence of mass effect and absence of surrounding edema. In accordance with other studies based on epilepsy surgery, the majority of tumors in this series were located in the temporal lobe (64%), and most of these were located medially. However, it should be noted that DNTs may be encountered in any other part of the supratentorial cortex.

The cortical location of the lesion was more clearly visualized on MR imaging, especially on T2 Weighted Images, than on CT scans or T1 Weighted Images. However, large tumors may obviously exceed the thickness of the normal adjacent cortex on MR scans. Even in these large lesions, neurons are identified either in the deeper part of the lesion or in

| TABLE II. — Radiological findings in the 3 histological forms of DNTs. |
| --------------------------- | --------------------------- | --------------------------- |
|                             | « complex » form (n = 14)   | « simple » form (n = 6)     | « non specific » form (n = 33) |
| CT imaging                 |                            |                            |                              |
| Normal                     | n = 0                      | n = 0                      | n = 6                        |
| Abnormalities:             |                            |                            |                              |
| Hypodense lesion           | n = 14 (100 %)             | n = 6 (100 %)              | n = 23 (70 %)                |
| Hyperdense lesion          | n = 0                      | n = 0                      | n = 4 (12 %)                 |
| MR imaging                 |                            |                            |                              |
| T1-WI:                     |                            |                            |                              |
| Hypointense                | n = 9 (100 %)              | n = 6 (100 %)              | n = 26 (90 %)                |
| Isointense                 | n = 0                      | n = 0                      | n = 3 (10 %)                 |
| T2-WI: Hyperintense        | n = 9 (100 %)              | n = 6 (100 %)              | n = 29 (100 %)               |
| Calcic components          | n = 3                      | n = 0                      | n = 16                       |
| Contrast enhancement       | n = 5                      | n = 0                      | n = 5                        |
| Cystic component           | n = 0                      | n = 0                      | n = 4                        |
FIG. 4. — Imaging appearance of a “complex” form of DNTs:
  a) Non-enhanced CT scan reveals a parietal lesion with calcific hyperdensity.
  b) Calvarial remodeling is evident on bone window CT scan.

FIG. 4. — Aspect radiologique d’une forme “complexe” des DNT:
  a) Coupe scanographique sans injection mettant en évidence une lésion corticale pariétale calcifiée.
  b) L’empreinte sur la voûte osseuse est mieux visualisée sur la fenêtre osseuse.

FIG. 5. — Imaging appearance of a “non specific” form of DNTs.
  a) Coronal proton-weighted MR image shows a hyperintense lesion of the right internal temporal cortex.
  b) Sagittal T1-weighted MR image after gadolinium administration demonstrates a nodular enhancement of the lesion. CT scan was normal in this patient.

FIG. 5. — Aspect radiologique d’une forme “non spécifique” des DNT:
  a) Coupe coronale en séquence densité de protons mettant en évidence une lésion nodulaire hyper intense du cortex temporal interne droit.
  b) Prise de contraste nodulaire de la lésion sur la séquence sagittale T1 avec injection de gadolinium. Le scanner de ce patient était considéré comme normal.

FIG. 6. — Imaging appearance of a “complex form” of DNTs: coronal T1-weighted MR image reveals a cortico-subcortical fronto-parietal lesion with a ring-like enhancement. Note the lack of surrounding edema.

FIG. 6. — Aspect radiologique d’une forme “complexe” des DNT : coupe coronale T1 avec injection de produit de contraste mettant en évidence une lésion fronto-pariétale cortico-sous-corticale prenant le contraste de façon annulaire, et sans “œdème” périléSIONnel.

FIG. 7. — Imaging appearance of a “complex” form of DNTs: coronal T1-weighted MR image reveals a right medial temporal hypointense, heterogeneous lesion, with no mass effect.

FIG. 7. — Aspect radiologique d’une forme “complexe” des DNT : coupe coronale T1 mettant en évidence une lésion hypointense, hétérogène, sans effet de masse, corticale au niveau du lobe temporal droit interne.
the adjacent white matter [7]. Since, DNTs occurs during embryogenesis, their development may induce neuronal migration disorders. As a result, part of the neurons may, like megagyri, remain ectopic (figure 10) [7].

The radiological features that we have described correspond to a routine radiological semiology. However, routine MR imaging techniques may be dramatically improved by the use of surface coils [13]. In future, surface coil imaging of the cerebral cortex might allow the detection of neuronal ectopia associated with DNTs, and thus might provide an additional radiological criterion for the differential diagnosis between DNTs and « ordinary gliomas ».
Another radiological feature frequently encountered in DNTs is a deformity of the overlying skull (71% of patients with a lesion of the convexity). Assuming that the development of DNTs takes place during embryogenesis, it may be postulated that they occur before the ossification of the vault, and that as a result the bone is modeled around the tumor.

The three histological forms of DNTs show different radiological features. Histologically, the « simple » form of DNTs consists exclusively of a « specific glioneuronal element » that contains an abundant pale eosinophilic interstitial fluid and has a characteristic semi-liquid appearance. On CT and MRI examinations this morphological variant of DNTs is seen as a homogeneous cystic-like cortical lesion that does not show contrast enhancement. None of the « simple » form of DNTs in our study showed calcic hyperdensity. In accordance with their histological heterogeneity, the « complex » and « non specific » forms have polymorphic radiological features. They may or may not show contrast enhancement or calcifications. Contrast enhancement revealed either a nodular, patchy or ring-like appearance. Histologically this latter pattern corresponds to glomeruloid capillaries [1, 3, 7]. Contrast enhancement in DNTs, in contrast to ordinary gliomas, is not associated with peri-tumoral edema or mass effect. The radiologist should bear in mind that DNTs may have variable appearance on imaging and that the categorization into « simple », « complex » or « non specific » histological forms has no clinical or therapeutic implications but merely reflects varying difficulties in terms of diagnosis.

« True » cysts are uncommon in DNTs, and are usually small. These radiological features distinguish DNTs from gangliogliomas. The differential diagnosis between gangliogliomas and DNTs is the main diagnosis problem [4, 14]. Like DNTs, these tumors are more common in children and young adults with chronic partial seizures. Small gangliogliomas may be predominantly cortical. Histologically, the differential diagnosis between the « non specific » form of DNTs from gangliogliomas may also be problematic because the ganglion cells, which are often present only focally, may not be observed on non representative samples. However lymphocytic cuffing is typically observed in gangliogliomas but is absent in DNTs [7]. A few radiological studies [14-16] showed that gangliogliomas have contrast enhancement in the solid part of the tumor and are associated with a large cyst whereas cysts are uncommon in DNTs. In our study a small cyst (less than 1 cm) was found in only four patients. Therefore, the presence of a large cyst on imaging and the presence of lymphocytic cuffing on histological examination are important criteria to distinguish gangliogliomas from DNTs. Delineating these two tumor types makes sense from a prognostic point of view because, although gangliogliomas are benign tumors, slow growth or malignant transformation [14] is possible, whereas DNTs are carcinologically stable lesions.

In addition to gangliogliomas, oligodendrogliomas are a differential diagnosis of DNTs. Oligodendrogliomas usually occur after 18 years of age (mean age : 35 years). Radiological studies [17-20] showed that oligodendrogliomas are usually cortico-sub cortical cerebral tumors with mass effect or surrounding edema, and may have calcic components and contrast enhancement. The diagnosis of ordinary glioma could be straightforward in patients with mass effect or surrounding edema. Nevertheless, in patients with low-grade oligodendrogliomas the differential diagnosis with DNTs is more difficult. In most instances patients with low-grade oligodendrogliomas present with partial or generalized seizures after 20 years of age, and on imaging the tumor involves the cortex but also extends in the white matter [21, 22]. However, in a minority of cases partial seizures may occur before the age of 20 years, and on examination the tumor may present a predominantly cortical location and appear well delineated on MR imaging. Therefore, in such instances, especially in patients aged between 18 and 22 years, the differential diagnosis between DNTs and oligodendrogliaoma is more difficul to establish.

In all cases preoperative radiological follow-up demonstrate that DNTs are stable lesions. Although in three cases changes in the pattern of contrast enhancement or newly formed contrast enhancement were observed on control imaging, the size of the lesion remained unchanged. No tumoral recurrence if resection was complete, or growth of the residual tumor if surgery was partial, could be observed on post-operative control imaging.

A few radiological series have reported CT and MR imaging of DNTs. DNTs have been described as well-demarcated cortical or cortico-subcortical lesions, with no peritumoral edema. The neuroradiological features of DNTs reported by Kuriowa [8, 12] (n = 10) and Koeller [10] (n = 6) and in the 16 cases included in the study of Ostertun [9] were consistent with those observed in our study. However, Ostertun [9] found a slight (n = 5) or obvious (n = 4) mass effect in nine cases, one of which was related to an intra-tumoral hemorrhage. We found no mass effect in any of our patients; however, a print on the ventricle (n = 3) or the calvarium (n = 15) were considered separately in our study.

Kuriowa [8, 12] and Koeller [10] did not mention a long-term follow-up. Contrary to previous reports, Ostertun et al. [9] found growth of the tumor in two out of the six patients in whom serial preoperative radiological findings were obtained. However, in one case with « tumoral growth » and peritumoral edema, changes were attributed to tumour hemorrhage, and in the other case changes in size were at-
tributed to the growth of a cystic component. In these two cases histopathological evaluation showed no signs of malignant transformation. DNTs may contain hamartomatous vessels, as observed in three cases in our study, which may lead to intra-tumoral hemorrhage. Cases of intra-tumoral hemorrhage leading to a marked mass effect have recently been reported [23]. We also observed the occurrence of a hemorrhagic infarct in two « complex » forms of DNTs that were responsible for contrast enhancement and mass effect (personal observation). In DNTs, growth of the cystic part of the lesion, changes in the shape of contrast enhancement, the occurrence of calcification or contrast enhancement, or intra-tumoral hemorrhage may be observed but do not imply malignant transformation. To date only one case of malignant transformation has been reported [24].

The diagnosis of DNTs may be difficult to establish histologically not only in the « non specific » forms but, for obvious reasons of surgical sampling, also in the simple or complex forms [4, 5, 7]. The diagnosis of DNTs must be suggested by the radiologist when all the following criteria are met: 1) partial seizures beginning before age of 20 years, 2) no neurological deficit other than a stable and probably congenital neurological deficit, 3) cortical topography of the lesion better demonstrated on MRI, 4) no mass effect on CT and MRI examinations and 5) no peritumoral edema. As observed in this study a later onset of symptoms occurs in a minority of patients (n = 4). While the age of the patients cannot be considered as a criterion of exclusion, in such instances the diagnosis of DNTs should be considered with more caution. This study, as most of the other series in the literature, is based on epilepsy surgery, and thus includes patients with long-term chronic seizures. However, nowadays CT and MRI tend to be systematically performed soon after the first seizures and it is thus likely that the different forms of DNTs, including the recently described « non specific » histological form, are much more frequent in children than previously thought. The role of the radiologist is critical in terms of establishing the diagnosis of DNTs and also for the follow-up of the lesion and the post-surgical follow-up. This implies that the same radiological examination protocol must be used, throughout so to allow tumoral changes or recurrence to be detected. These diagnosis difficulties increase also the risk of over diagnosing DNTs. In absence of specific morphological features, a diagnosis of DNT should not be made unless all of the five criteria described above are met. By bearing in mind that, in children or young adults, tumors that resemble « ordinary gliomas » on CT and MRI examinations may be DNTs, radiologists would certainly contribute to avoiding inappropiate treatment, such as radiotherapy or chemotherapy, in patients with normal life expectancy.


REFERENCES


