Commentary

In the current issue of journal of neurosciences in rural practice, the image and case reported entitled "Intra cranial Rosai-Dorfman disease" (RDD)[1] illustrates the main difficulty to reach the diagnosis of that rare non langerhans cell histiocytosis.

As reported here, this disease firstly described by Destombes[2] in 1965, usually presented in children and adolescents with massive painless cervical lymphadenopathy, often with associated fever, mild anemia, polyclonal hypergammaglobulinemia, and an elevated sedimentation rate, may present in adults with extranodal form,[3] and, in a few cases, about 5% as an intracranial isolated entity.[4]

Interestingly, the difficulty to reach that diagnosis appears once the intracranial process is showed by computed tomography (CT) or magnetic resonance imaging (MRI). Indeed, different features in the neuro imagery may be encountered: Solitary or multiple, well demarcated, predominantly homogeneous dural-based extra-axial masses associated with surrounding vasogenic edema as described by Zhu and colleague.[5] Especially, intracranial RDD may mimic solitary or multiple meningioma.[6] It is important to note in addition to the author's commentary that epidemiologic clinical data may be helpful in order to distinguish RDD from other dural-based lesions.[6] Patients with intracranial RDD are usually younger than those with sporadic meningioma or neurofibromatosis type 2 with an average of 40 years. The last point about imagery findings of these lesions mimicking meningioma in many radiological respects is that they are characterized by an absence of calcification on CT and hypointensity on T2-weighted MRI,[7] which differs from the case reported here.

Management of diagnosis and treatment should consider the degree of emergency: Is that intracranial process threatening life or function like in the present case or not? In such situation, neurosurgery aiming total or near resection is indisputable. After a literature review, intracranial RDD is not described as bleeding tumor but one case of complication after gross total resection was reported[8] with a postoperative hemorrhage (and a low-density lesion in the right frontal lobe) similar to the present case. The main point is the preservation of venous drainage, especially closed to the midline and on the skull base which is not specific of RDD surgery. It is probably the reason why some authors as Tomio and colleague[8] suggest a near total resection and adjuvant radiotherapy in skull base or peri sinuses localizations.

This case report dwells on the histopathological diagnosis and this is the only means to assess the diagnosis of RDD. The pathognomonic signs are as noticed here the emperipolesis and foamy histiocytes. Histochemistry permit to reach that rare diagnosis by showing spindle cells and lymphocytic infiltration, staining CD 68+, not mentioned in the case, and PS 100. Among the others markers mentioned, the CD1a is particularly important to distinguish RDD from Langerhans cell histiocytosis.

Besides, consideration has to be paid to the polyclonal tissue different from plasmacytoma or lymphoma tumoral tissue.[6]

Another burning issue is raised by the author when the treatment is evoked: If surgery appears to be the only means to permit both to treat the vital or functional threaten and to obtain tumoral tissue to analyze, outcome is probably not as good as mentioned. Adeleye and colleague who gave a large review of literature cases (111 cases),[4] found 45 cases without recurrence (41%), 28 with stable disease (25%), 12 with relapse/growth (11%) and 4 death (4%) but only 19 cases (17%) among them with data after three years of follow-up. In fact, recurrences are not exceptional and not necessary during the three years following surgery.[4,6]

Overall the main interest of this image and case report is to present a rare intracranial differential diagnosis important to know of dural-based lesion like meningioma. Many important characteristics were already mentioned: Histopathological diagnosis, importance of surgery. Furthermore, and in addition to the other characteristics mentioned in the present summary, attention has to be paid to the real need of strong data and especially long follow-up and therapeutic strategy from a prospective study.

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