ORIGINAL RESEARCH PAPER

Oncology

KEY WORDS: Squamous-cell lung carcinoma, pancreas, metastasis, case-report

INTRA CEREBRAL DYSGERMINOME REVEALED BY SECONDARY AMENORRHEA: ABOUT A CASE WITH THE LITERATURE

A.Debbagh*	Medical Oncology Service, Instruction Military Hospital Mohamed V, Rabat *Corresponding Author
K. Alaoui Slimani	Medical Oncology Service, Instruction Military Hospital Mohamed V, Rabat
Y.Sbitti	Medical Oncology Service, Instruction Military Hospital Mohamed V, Rabat
M.Ichou	Medical Oncology Service, Instruction Military Hospital Mohamed V, Rabat

Introduction:
The germinal tumors of SNC are extremely rare tumors, their incidence in the literature does not exceed 1 % of all the brain tumours, essentially represented by dysgerminomes (1). Their modes of revelation are multiple and varied. They are curable tumors especially as they arise at the young subject. The treatment relies on the radiotherapy, however the contribution of the chemotherapy allows an optimization of the global therapeutic strategy (2). In this work we report the case of a young patient with dysgerminone from the supra-sellar region revealed by secondary amenorrhea successfully treated with the combination of first-line chemotherapy followed by radiotherapy.

Clinical case:
This is a 22-year-old patient with no significant pathological history, who consulted for neglected secondary amenorrhea for 3 years, associated with a poly-poly-polydypsin syndrome in a context of general state preservation. The clinical examination at admission was without any particularities.

The initial hormonal assessment revealed anterior pituitary insufficiency (Prolactinemia and disrupted cortisolemia) supported by endocrinology.

The brain MRI performed (Figure 1) was in favor of a supra-sellar tumor filling the supra-sellar cistern, spreading the cerebral peduncles and pushing back the 3rd ventricle.

A stereotaxic biopsy scanned-guided anatomically pathological morphologically (Figure 2) and an immunohistochemical profile (Figure 3) of a dysgerminoma.

The CTAP scanner made it possible to affirm the isolated nature of the supra-sellar lesion, thus eliminating possible dysgerminoma metastases.

After multidisciplinary consultation, a neo-adjuvant chemotherapy was decided and the patient received 3 cycles of Carboplatin (AUC 6) associated with Etoposide at a rate of 100 mg / m^2 for 03 days (D1 = D21) with a good tolerance. The evaluation at the end of the chemotherapy showed a complete morphological response (Figure 4).

Subsequently, total ventricular irradiation was performed at the dose of 24 Gy (15 fractions of 1.6 Gy) with a complement of 10 Gy on the tumor bed.

Currently, after a 27-month follow-up, the patient remains asymptomatic under hormone replacement therapy and the control status remains unspecific.

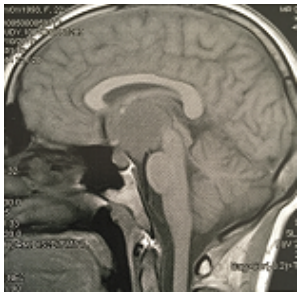

Figure 1 Brain MRI showing a dysgerminome

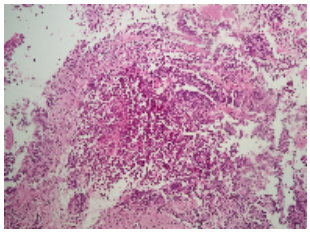

Figure 2 : Morphological aspect of an intra-cerebral dysgerminome (G×100)

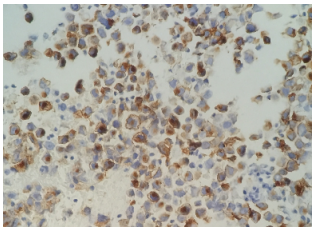

Figure 3 : Positive marking of the tumoral cells with Ac anti CD117 (G×400)

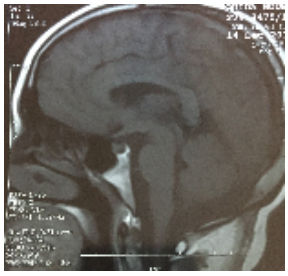

Figure 4 Mass disappearance after treatment

Discussion:
Extra gonadal germ tumors are rare, typically mediastinal, and more rarely retro-peritoneal. Dysgerminomas account for less than 1% of all brain tumors (1), and mainly affect the child and young

adult especially during the 2nd decade with male predominance (2). These tumors are preferentially localized at the level of the pineal region in 60-70% of cases and in the supra-sellar region in about 40% of cases (1.3) as was the case with our observation. The usual modes of revelation when the tumor is supra-sidelastic are diplopia, visual acuity and disturbance of the hypothalamic-pituitary axis, more rarely secondary amenorrhea as in our case (4). MRI is the examination of choice, dysgerminoma is in the form of a heterogeneous mass, isointense with iso- or hyposignal in T1-weighted sequence and in the form of a discrete hypersignal in T2 (5,6). The search for dissemination of the disease should be systematic either by looking for neoplastic cells in the cerebrospinal fluid or by performing a spinal MRI (3, 5, 7). In fact, histopathological examination is a crucial element for diagnosis and therapeutic management. Histologically, pure dysgerminomas are similar to primordial germ cells (3,4,5). Furthermore, the treatment is based on radiotherapy preceded by neo-adjuvant chemotherapy to reduce the target volumes and doses of irradiation: Sifat et al offer to administer 4 courses of chemotherapy (Etopside, Cisplatin) followed by irradiation of 40 Gy of the tumor bed and 25 Gy of the craniospinal axis if the disease is diffuse (5), Vederine et al offer 3 courses of chemotherapy (Carboplatin, Etoposide) supplemented if complete response, an irradiation of 26 Gy in the ventricular volume and a complement of 10 Gy in the tumor bed (1), as in our case. Although rare, metastases of dysgerminomas are possible and are most often secondary to seeding from the cerebrospinal fluid.

Conclusion:

The management of intracranial dysgerminomas represents an important therapeutic challenge to guarantee to young patients a cure without aftereffects. The excellent radiosensitivity of this tumor is accompanied by a potential toxicity, which requires defining a combination of optimal treatment in which the contribution of chemotherapy seems to have its place.

References :

1. Vedrine L, Bauduceau O, Fayolle M, Le Moulec S, Ceccaldi B. Combined chemotherapy and radiation therapy for intracranial germinomas. The Val-de-Grace hospital experience. Cancer Radiother 2005; 9: 335-40.
2. Alifdal M, El Abbadi N, Maftah M, Banani A, Benjaafar N, Bellakhdar F. Double localization of intracranial germinoma. Neurochirurgie 1995; 41:424-8
3. Louis DN, Sheithauer BW, Budka H, Von Demling A, Kepes JJ: meningiomas in Kleihues P, Cavanee WK (eds): World Health Organization. Pathology and Genetics of Tumours of the Nervous system. Lyon IARC Press, 2000.
4. Agranat P, Jedynak P, Epardeau B, Extra JM, Mignot L. Cerebral germ cell tumors in adults. A retrospective study of 19 cases.Bull Cancer 1995;82:650-9.
5. Sifat H, Haddadi K, el Ghazi E, Errihani H, Kamouni M, Mansouri H, Hassouni K, Bakkali H, Kanouni L, Gaye M, Kebdani T, Benjaafar N, el Gueddar BK. Central nervous system germinoma: retrospective study of six cases. Cancer Radiother 2002; 6:273-7.
6. El Abbadi N, Maaqili MR, Arkha Y, Amarti A, Bellakhdar F. Cerebellar primary germinoma. Case report. Neurochirurgie 2002; 48:351-4.
7. Gauvrit J.Y, Soto Ares G, Hamon-Kerautret M, Blond S, Pruvo JP. Imagerie des tumeurs de la région pinéale. Feuillets de Radiologie 1997 ; 37 : 287-299