

UKRAIN TREATMENT OF ASTROCYTOMAS IN GIRL WITH TUBEROUS SCLEROSIS: A CASE REPORT

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Summary: *A 13-year-old girl with tuberous sclerosis and subependymal giant cell astrocytomas was treated with Ukrain. Although neurosurgical operations were also performed, complete tumor excision was not possible. Ukrain led to a great improvement in clinical status and also to partial regression of astrocytoma growth, which can be observed to date.*

Introduction

Ukrain, a semisynthetic drug, is a chelidonine thiophosphoric acid derivate with immunomodulatory and cancerostatic properties in cancer patients (1). Its efficacy has been shown in humans in a number of tumors with different localization: mammary gland, stomach, large intestine, lung, ovaries, urinary bladder, kidney, melanoma, cholangiocarcinoma, sarcomas, etc. (2). In view of these data and the description of good efficiency of Ukrain in two cases of astrocytoma (3, 4), we proposed the use of Ukrain in the case of tuberous sclerosis with subependymal giant cell astrocytomas.

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Tuberous sclerosis, or Bourneville-Pringle disease, is a complex genetic disorder characterized by the formation of multiple hamartomas of different organ systems (5). Since the first description of the classic triad of adenoma sebaceum, epilepsy and mental retardation by Vogt (6), various manifestations have been added to the clinical picture of the disease: classic shagreen patch; unguis fibroma; retinal hamartomas; renal angiomyolipomas; cardiac rhabdomyoma; and bilateral polycystic kidneys (7). Less than 2% of patients have subependymal giant cell astrocytoma (8).

Patient and method

A 13-year-old girl complained of severe headache when she was first observed in our clinic in

October 1996. The patient was also depressed and uncommunicative. The clinical features of tuberous sclerosis, seizures and adenoma sebaceum, were present. Computed tomography (CT) scan of the brain showed a 2.2-cm tumor in the region of the septum pellucidum, extending into Monro's foramen and consequently obstructing the cerebrospinal fluid (CSF) pathway, especially at the left side. Following two previous interventions to evacuate the epidural hamartoma and to drain the left ventricle due to hydrocephalus, respectively, the patient underwent an operation for partial tumor extraction in November 1996. On histological examination, a giant cell astrocytoma was found.

After the operation severe headache remained, vomiting two to three times per week appeared, and CT scan showed cerebral hypertension. In December 1996, magnetic resonance imaging (MRI) showed the presence of five nodes with diameters ranging from 7 mm to 3.4 cm on both sides of the brain; however, the tumor seen previously by CT scan in October 1996 was not shown. On February 14, 1997, ventriculoperitoneal shunt surgery was carried out.

The patient and her parents were informed about the possible side effects of Ukrain (Nowicky Pharma, Vienna, Austria) and agreed to undergo the therapy with it. The first Ukrain injection was administered on January 1, 1997, and the course lasted until February 13, 1997. Ukrain was administered intravenously at a dose of 5 mg (5 ml) twice a week, and the total dose administered over the course of treatment was 35 mg (35 ml). In general Ukrain-monotherapy was carried out until June 1997 with the same dosage regimen.

Results

Even after the first relatively short course of Ukrain therapy, on February 15, 1997, examination by X-ray did not reveal any progressive tumor growth nor

appearance of any new nodes. The patient's neurological condition improved and postoperative CT scan showed no hydrocephalus. During the treatment period both psychic and somatic status improved, the girl was lively and friendly, and managed to study at school. All follow-up CT examinations showed the regression of the non-operated nodes; only two nodes instead of five remained, with diameters of 2.5 cm and 4 mm, respectively. Furthermore, the CSF pathway obstruction was not shown. No side effects or allergic reactions were associated with Ukrain administration. The patient remains in remission to date.

Discussion

In view of the young age of the patient and the development of the clinical picture of hypertension in CSF pathways, the therapeutic approaches in this case were limited. While a number of symptomatic neurosurgical operations were performed, progressive growth of intracranial tumors made the prognosis unclear.

Ukrain administration led to a great improvement in clinical status and also to partial regression of astrocytoma growth, which can be observed to date. Accordingly, we propose the use of Ukrain as a cytostatic and immunomodulatory agent in the case of tuberous sclerosis with astrocytoma development.

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