State University of New York
Health Science Center
Syracuse

Department of Pediatrics

Hematology/Oncology Division
including:
- Comprehensive Hemophilia Center
- Comprehensive Sickle Cell Center
- K.N.O.T. (Kids Now Off Therapy) Clinic
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December 25, 1996

RE: **Sophia B. Gettino**
DOB: 01/29/96
Private Pediatrician: Dr. W[redacted]
First Seen: 12/22/96 in 4C

DIAGNOSIS: 1. PRIMITIVE (SMALL BLUE CELL) NEUROECTODERMAL TUMOR CONSISTENT WITH PINEALOBLASTOMA or EPENDYMOBLASTOMA
2. OBSTRUCTIVE HYDROCEPHALUS

Sophia was seen by our service for evaluation of pineal tumor.

History of Present Illness: She had a 2-week history of inability to readily move her head or look up. She had decreased eating and impaired coordination. She became progressively irritable, lethargic with decreased activity. She quit crawling and sitting up, and her balance was poor with frequent fell. She had no emeses or seizures. On physical examination, she had large head circumference and tense anterior fontanelle. Pre-operatively, she had left sixth and fourth cranial nerve pareses and parinaud's syndrome (impaired upgaze and light dissociation of pupillary reflexes). Fundoscopic examination showed mild swelling of the optic disk without frank papilledema. Her blood pressure was 84/p and head circumference 49 cm.

Past-medical History: She was born by natural delivery following full term pregnancy. Her immunization was up-to-date.

Family History: Unremarkable. She lives with both parents.

MRI of the brain showed obstructive triventricular hydrocephalus secondary to a large heterogenous cystic pineal region mass. The mass measured 2 to 3 cm and enhanced with gadolinium.

On 12/18/96, she had left frontal endoscopic third ventriculostomy with biopsy of pineal tumor and placement of left frontal and 3rd ventriculostomies. The tumor was vascular. Pathologic examination of the "touch prep" sample from a biopsy of the posterior third ventricular/pineal region tumor showed hypercellular tumor composed of small blue cells with high nuclear-to-cytoplasmic ratio, slight nuclear pleomorphism and scant cytoplasm. There was a prominent pattern of rosette formation. Mitoses were present without necrosis. Spinal fluid cytology was negative for tumor cells.

On 12/20/96, she had left occipital transtentorial craniotomy and gross total resection of pineal region tumor. The operative findings were significant for greater than 2-cm diameter mass, with a firm fibrous and vascular capsule. Near total resection was achieved.

Because of her age, short onset of symptoms and pathologic findings, her prognosis unfortunately is very poor. The option of chemotherapy was presented to the family. One possibility was the Memorial Sloan-Kettering Cancer Center's protocol (Dr. Jonathan Finlay):

"DOSE-INTENSIVE CHEMOTHERAPY FOR CHILDREN LESS THAN SIX YEARS OF AGE NEWLY DIAGNOSED WITH MALIGNANT BRAIN TUMORS: A PILOT STUDY OF INTENSIVE INDUCTION CHEMOTHERAPY FOLLOWED BY CONSOLIDATION WITH MYELO-ABLATIVE CHEMOTHERAPY (THIOTEPA, ETOPOSIDE AND CARBOPLATINE) AND AUTOLOGOUS MARROW RESCUE

This protocol was given to the parents for their consideration. A central line will be placed on 12/26 for supportive care.

If I can be of any further help please do not hesitate to call me at 315-464-5294.

A[redacted]d, M.D., Ph.D.
Pediatric Hematology/Oncology

cc: parents