Pediatric neuroblastoma: Long-survivor

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Introduction. Neuroblastoma is an embryonal tumor of the sympathetic nervous system and his first localization is the adrenal gland. It is the most common solid tumor in childhood, 90% are diagnosed in the first 5 years of life and generally in advanced stages. Factors of poor prognosis are the abdominal location, the undifferentiated cells, more than one year old and amplification of N-myc gene (shown in 20% of cases). Tumors are sensitive to chemotherapy and radiotherapy. The estimated overall survival is 30-50% CASE: 7 year old girl with a VP shunt valve for hydrocephalus secondary to Chiari malformation type II and she presents incontinence. At 2 years old is diagnosed with stage IV neuroblastoma, with N-myc amplified, adrenal primary and metastatic involvement (lymph node and bone), achieving complete remission after chemotherapy. She was treated with adrenalectomy and autologous hematopoietic stem cell (bone marrow biopsy negative) followed by abdominal radiotherapy given 21 Gy. At 5 years to diagnosed she reports radiographic progression at mediastinal, retroperitoneal and bone. Receive 5 cycles of chemotherapy with complete response, autologous hematopoietic progenitors and mediastinal radiotherapy given 21 Gy by residual lesion. At present, after 5 years of diagnosis, the disease is stable and without relapse.
Conclusions. We report a case with poor initial prognosis, diagnosed at 2 years old with stage IV, located at abdominal, unresectable with amplified N-myc gene, but with a multidisciplinary treatment the patient is asymptomatic and disease-free.

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Pediatric tumors: Embryonal rhabdomyosarcoma GIII

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Six year old, female patient. Oncological history: Due to a gastrointestinal affectation and a severe general involvement, she visits the ER where she is found to have GI bleeding and a tumor in hypogastrium. She is urgently surgically intervened on April 15, 1981, and a mass 10 cm × 8 cm in size is removed from anterior and posterior bladder, which at that moment has tore and is bleeding profusely and has a cerebroid aspect; the pathological anatomy is consistent with a grade III embryonal rhabdomyosarcoma. Upon this finding, chemotherapy is decided upon according to protocol (Vincrisul, actinomycin D and cyclophosphamide during 4 cycles). Later on, she is giving a RTE-2D treatment with Cobalt 60 by means of 4 fields, two laterals and a anterior and posterior that encompass the entire volumen of the tumorectomy. Total dosage of 42 Gy. Fractionation 5 × 150 cGy. Renal protección was given at lateral and posterior fields, and hepathic protection with an anterior field. The treatment lasted from 10/06/81 to 22/07/81. She tolerated it well. Evolution: Patient remained in follow ups by pediatrics. Following her diagnosis of primary hypogonadism, secondary to pelvic radiotherapy, a substitute hormonal therapy with ethinilestradiol is started in 1992 but was suspended due to lack of growth. She then, received growth hormone but discordant corporal proportions were observed. Reason for which she was given microgynon, experimenting menarche that same year. At age 17, in 1993 during physical exploration, the patient is found to have a grade I thelarche, grade II pubarche and grade III axilarche. From there on she has had a normal secondary sexual development. Current status Toxicity outstanding: sterility. Currently this patient is symptom free (ILE 360 months).

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Perivascular epithelioid cell tumor of the uterus (PECOMA) in a 21-year-old patient treated of meduloblastoma in her childhood (at the age of 5)

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Introduction. Patient diagnosed at the age of 5 from a posterior fossa meduloblastoma. At the age of 21, new diagnosis of a PECOMA uterus tumor. Long survival with low side effects.
General review. Medulloblastoma is the most frequent solid tumor (CNS) in children. Survival rate at five years is <50%. Common treatment is based on surgery, radiation and chemotherapy. Wide range of side effects associated with treatment. Uterus PECOMA is an infrequent (<1 case in 100,000) tumor that is related with the Tuberous Sclerosis (TS) spectrum of diseases. It is considered as an unknown behavior malignant tumor. Recent data show relation between TS tumorigenic pathways and medulloblastoma.
Our case. Our patient suffer from Tuberous Sclerosis. She was diagnosed and treated at the age of 5 with complete surgical resection followed by chemoradiation (30 Gy to craniospinal axis and 50 Gy to posterior fossa as a boost with a 2D planning) and adyuvant chemotherapy. At the age of 21, in March 2012, in a gynecological review due to metrorrhagia, she was diagnosed from a PECOMA uterus tumor. Simple hysterectomy was done. No adjuvant treatment associated. No evidence of recurrence of both tumor at the present time. Carrying out a normal life.