

13 April 2022

Patient: Amin Adilov MRN: 1822248 DOB: 30/10/2012

Amin is now 9 years old and was diagnosed of stage 4 neuroblastoma MYCN non amplified in May 2019 in Moscow. Primary tumor is reported in the right adrenal gland. Extent of disease work-up showed B/BM infiltrates. Amin received chemotherapy according to the local protocol with 8 chemotherapy cycles and surgery on 12/9/2019. Subsequently he received ASCT (January 2020) and radiotherapy (Feb-Mar 2020). Despite all the treatment he never achieved complete remission showing primary refractory disease affecting bones and soft tissues of the primary tumor area. From March 2020 until February 2021 he received continued cycles of TMZ and cyclophosphamide with stable disease.

Amin arrived to our institution in April 2021. Upon arrival he was asymptomatic. The extent of disease work-up showed: BM (20/04/2021) negative. Whole body MRI (22/04/2021) one soft tissue nodule ($30 \times 18 \times 20$ mm) in the gastro-hepatic and a nodule ($19 \times 18 \times 13$ mm) in the left kidney hilium. MIBG (21/04/2021) showed abnormal uptakes in multiple bones (Curie score =10) and 2 retroperitoneal soft tissue masses where MRI showed.

With such results we started him on ICE chemo but developed hemorrhagic cystitis because of BK virus infection. The second cycle was with Irinotecan and Temozolamide (I/T). Evaluation after the 2 cycles showed: BM studies (08/06/2021) negative. CT of the abdomen (16/06/2021) similar soft tissue nodules (24 x 13 x 24 mm in the gastro-hepatic ligament and (22 x 20 x 16 mm) in the left para-aortic area. MIBG (10/06/2021) with partial response of the bone sites (Curie score =4) and persistent 2 retroperitoneal soft tissue masses. On 1/7/2021 surgical resection of the retroperitoneal masses was undertaken. Pathology showed ganglioneuroblastoma. A new disease work-up after surgery showed: MIBG (08/07/2021) Curie score=10 and persistence of the soft tissue uptake in the gastro-hepatic ligament.

With such results showing the soft tissue and bone refractory disease, he was enrolled into the chemo-immunotherapy trial NICE (naxitamab + GM-CSF with I/T and ICE). Amin began the first cycle NICE July 19 and the second on August 18, 2021. Disease evaluation after the 2 cycles showed: BM (30/08/2021) negative; MIBG (01/09/2021) negative. Amin had reached first CR after 2 cycles HITS, and 2.5 years post diagnosis. He then received 5 more cycles of naxitamab and GM-CSF only (13 September, 11 October, 8 November, 14 December 2021 and 3 January 2022). Disease evaluation after 2 cycles of only immunotherapy confirmed continued CR: BM (18/10/2021) negative; MIBG (22/10/2021) negative. The disease evaluation at the end of treatment confirmed continued CR: BM (12/01/2022) negative; MIBG (13/01/2022) negative. Amin was doing clinically very well, with no evidence of secondary side effects and running a normal life for his age. We then recommended disease check-up every 3 months with MIBG and BM studies. Also, craniospinal MRI should be performed once a year.



Given the clear benefit from immunotherapy in this patient, we advised the family to continue on a preventive plan of naxitamab and GM-CSF every 2 months until the bivalent vaccine becomes available at our center.

Sincerely,

DR. JAUME MORA
Scientific Director PCCB

