TIENTS: Patient A: A 17 months old male presented with non-metastatic bilateral CPC. A de novo mosaic germline TP53 mutation was identified. After near-total resections, 16 months of standard chemotherapy were administered; 18 months later, localized tumor growth developed, again near-totally resected. Two cycles of re-induction chemotherapy were administered followed by three cycles of thiotepa/carboplatin with autologous hematopoietic cell rescue (AuHCR) and subsequently 21 months of sirolimus and thalidomide, continuing without residual or recurrent disease. Patient B: A 30 months old male presented with left lateral ventricular non-metastatic CPC. A de novo TP53 germline mutation was identified. Following subtotal resection, craniospinal irradiation with boost was administered followed by eight cycles of standard chemotherapy; 18 months later, localized recurrence developed; gross total resection was followed by 15 months of standard dose chemotherapies; four months thereafter, a second local recurrence developed, again gross totally resected. He then received one cycle of high-dose cyclophosphamide followed by three cycles of thiotepa/carboplatin with AuHCR. Subsequently he received sirolimus and thalidomide for 12 months, complicated by progressive pancytopenia. A small localized CPC recurrence was noted, gross totally resected, concomitant with myelodysplastic syndrome; he underwent an allogeneic matched unrelated donor marrow transplantation. CONCLUSIONS: Marrow-ablative chemotherapy with post-transplant targeted biological therapy may afford durable survival for select children with recurrent CPC.

RARE-32. PEDIATRIC METASTATIC SKULL BASE CHORDOMA WITH TP53 MUTATION – A CASE REPORT AND REVIEW OF THE LITERATURE

Akira Shimada[1], Kazuhiko Kurozumi[1], Kiichiro Kanamitsu[1], Hisashi Ishida Ishida[1], Kaori Fujiwara[1], Kana Washio Washio[1], Tadashi Kumamoto[2], Chitose Ogawa[2], and Masahiro Shin[3]; [1]Okayama University Hospital, Okayama, Japan, [2]National Cancer Center, Tokyo, Japan, [3]Tokyo University, Tokyo, Japan

Chordoma is an uncommon bone tumor arising from notochordal remnant, which accounts for 1–4% of all bone malignancies. It commonly occurs along the cranial-spinal axis, and skull base is one of most frequent sites, representing 35–49% of all chordoma cases. Surgical resection is widely accepted as the first choice of treatment. There are only limited number of reports about pediatric chordoma cases, and its biological behavior including genetic backgrounds were largely unknown. Here, we present a 5 year-old girl with a large aggressive skull base chordoma of 6 cm in maximum diameter, which eventually had multiple systemic metastasis. We initially tried chemotherapy based on the protocol for the osteosarcoma, but in vain. Because the tumor was highly vascularized on angiography, after embolization of the feeding arteries and bilateral internal maxillary arteries, endoscopic endonasal surgery was performed. The tumor was sufficiently removed, achieving effective mass reduction, and the residual tumors involving the lower cranial nerves and craniocervial junction were additionally treated with Gamma Knife radiosurgery. However, one month later, it showed systemic metastasis to bilateral cervical lymph nodes and lung. We tried chemotherapy with nivolmab and imatinib for this patient, whereas they showed the partial effect. The genetic analysis revealed somatic TP53 c.569C>T, (p.P190L) mutation in chordoma specimen. In the past literature, we found only one study of the adult chordoma cases, in which majority of the patients had somatic TP53 mutation (p.P72R). Further investigation with large number of the cases is essential to clarify the molecular biology of pediatric chordomas.

RARE-33. GIANT CELL TUMOR OF THE SKULL BASE WITH A HISTORY OF A SUCCESSFUL RESPONSE TO DENOSUMAB AND LATER DEVELOPING A SECOND TUMOR

Manushak Avagyan[1,2], Gevorg Tamamyan[1], Samvel Danielyan[1], Martin Harutyunyan[2], Nune Karapetyan[3], Nerses Karamyan[4], Hovhannes Vardevanyan[5], Armen Mkhitaryan[6], and Samvel Bardakhchyan[1]; [1]Hematology Center After Prof. Yeolyan, Yerevan, Armenia, [2]Yerevan State Medical University, Yerevan, Armenia, [3]Yerevan, Yerevan, Armenia, [4]"IRA Medical Group" Radiotherapy Center, Yerevan, Armenia, [5]Armenian-American wellness center, Yerevan, Armenia, [6]"Histogen" Armenian-German Scientific Practical Center of Pathology, Yerevan, Armenia

BACKGROUND: Giant cell tumor of bone(GCTB) is a rare neoplasm with unpredictable behavior, possible malignant transformations, and/or lung metastases. Surgery is usually the treatment of choice. In unresectable or metastatic cases, treatment with denosumab is a new treatment option. CASE PRESENTATION: A 14-years-old female presented with cachexia, dysphagia, diplopia, discoordination, strabismus, and multiple cranial nerve palsies in 10.2015. MRI revealed intra-extracranial mass arising from C2 vertebrae, compressing the medulla oblongata and the left cerebellar hemisphere, invading to the sphenoid bone and nasopharynges. Biopsy showed a GCTB. Surgical resection was done, which was incomplete because of tumor location (cranial nerve and vertebral artery involvement). Then local radiation therapy was performed 50.4Gy. During RT patient's condition declined and MRI showed disease progression. Treatment with denosumab 120mg q4w was initiated in 03.2016, which yielded successful results. Disease was under control for three years until 03.2019. Then she returned with clinical symptoms of diplopia and severe headache. MRI showed local tumor progression. Repeated biopsy revealed undifferentiated pleomorphic sarcoma, which could be either a malignant transformation of GCTB or a new tumor. The patient later underwent two cycles of chemotherapy with Ifosfamide/Doxorubicin. MRI after 2nd cycle showed marked tumor progression. The patient didn't receive any further treatment because of cachexia and died due to disease progression in 12.2019. CONCLUSION: To our knowledge, this is the youngest patient ever reported with a skull base tumor with such a clinical development, successful and long-time remission with denosumab and with such a chemotherapy-resistant malignant transformation or second cancer.

RARE-34. UK CHILDREN'S CANCER AND LEUKAEMIA GROUP (CCLG): GUIDELINES FOR THE MANAGEMENT OF MENINGIOMA IN CHILDREN, TEENAGERS AND YOUNG ADULTS

Elwira Szychot[1,2], John Goodden[3], Whitfield Gillian[4], and Sarah Curry[5,6]; [1]Royal Marsden Hospital, London, United Kingdom, [2]Institute of Cancer Research, London, United Kingdom, [3]Leeds General Infirmary, Leeds, United Kingdom, [4]Christie Hospital, Manchester, United Kingdom, [5]Southampton Children's Hospital, Southampton, United Kingdom, [6]Our Lady's Children's Hospital, Crumlin, Ireland

Primary tumours of the meninges are rare accounting for only 0.4–4.6% of all paediatric tumours of the central nervous system. Due to the rarity of these tumours in children, and the consequent absence of collaborative prospective trials, there is no clear consensus on how the unique characteristics of paediatric meningiomas impact clinical status, management approach, and survival. Much of the evidence and treatment recommendations for paediatric meningiomas are extrapolated from adult data. Translating and adapting adult treatment recommendations into paediatric practice can be challenging and might inadvertently lead to inappropriate management. In 2009 Traunecker *et al.* published guidelines for the management of intracranial meningioma in children and young people on behalf of UK Children's Cancer and Leukaemia Group (CCLG). Ten years later we have developed the updated guidelines following a comprehensive appraisal of the literature. Complete surgical resection is the treatment of choice for symptomatic meningiomas, while radiotherapy remains the only available adjuvant therapy and may be necessary for those tumours that cannot be completely removed. However, significant advances have been made in the identification of the genetic and molecular alterations of meningioma, which has not only a potential value in development of therapeutic agents but in surveillance of childhood meningioma survivors. This guideline builds upon the CCLG 2009 guideline. We summarise recommendations for the diagnosis, treatment, surveillance and long-term follow up of children and adolescents with meningioma.

RARE-35. PINEOBLASTOMA IN CHILDREN SIX YEARS OF AGE OR LESS: FINAL REPORT OF THE HEAD START I, II AND III EXPERIENCE

Mohamed S. AbdelBaki[1], Mohammad H. Abu-Arja[2], Tom B. Davidson[3], Jason R. Fangusaro[4], Joseph R. Stanek[1], Ira J. Dunkel[5], Girish Dhall[6], Sharon L. Gardner[7], and Jonathan L. Finlay[1]; [1]Nationwide Children's Hospital, Columbus, Ohio, USA, [2]New York-Presbyterian Brooklyn Methodist Hospital, Brooklyn, Y, USA, [3]Children's Hospital Los Angeles, Los Angeles, CA, USA, [4]Children's Healthcare of Atlanta, Atlanta, GA, USA, [5]Memorial Sloan Kettering Cancer Center, New York, NY, USA, [6]The University of Alabama at Birmingham, Birmingham, AL, USA, [7]The Stephen D, Hassenfeld Children's Center for Cancer & Blood Disorders, New York, NY, USA

BACKGROUND: We report the outcomes of patients with pineoblastoma enrolled on the Head Start I-III trials. METHODS: Twenty-three children were enrolled between 1991–2009. Treatment included maximal surgical resection followed by five cycles of intensive-chemotherapy and consolidation with marrow-ablative chemotherapy and autologous hematopoietic cell rescue (HDCx/AuHCR). Irradiation following consolidation was reserved for children over six years of age or those with residual tumor at the end of induction. RESULTS: The median age was 3.12 years (range:0.44–5.72). Three patients withdrew from the protocols and two patients experienced chemotherapy-related mortality. Eight patients experienced progressive disease (PD) during induction chemotherapy. Ten patients received HDCx/AuHCR; eight experienced PD post-consolidation. Seven patients received craniospinal irradiation (CSI) with a median dose of 20.7 Gy (range:18–36 Gy) with boost(s) (median dose 27 Gy, range:18–36 Gy); three received CSI as adjuvant therapy (2 post-HDCx/AuHCR) and four upon progression/recurrence. The 5-year progression-free survival (PFS) and overall survival