

cell 5 (9%) and anaplastic 2 (4%). 19 cases were only reported as medulloblastoma WHO IV. Using immunohistochemical marker beta catenin and histology 43 tumours were subdivided into WNT (26), SHH (13), non-WNTnonSHH (4) groups.

Conclusion: There was an obvious male predominance in this group. 78% of cases were diagnosed within 3 months of onset of symptoms. 57% of cases had high risk disease.

ST-1_V1.5

INFLAMMATORY MYOFIBROBLASTIC TUMORS: A TALE OF THREE STORIES

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Background/Objectives: Inflammatory myofibroblastic tumors (IMT) are unusual intermediate malignant potential tumors that occur in soft tissues and visceral organs of children and young adults. The tumors consist of myofibroblastic spindle cells and inflammatory cells. These tumors can be locally aggressive, recur or rarely metastasize. There are no clear consensus guidelines on treatment for patients with such tumors. We describe three cases from our tertiary academic paediatric institute in Singapore.

Design/Methods: A retrospective analysis of paper, electronic records, and histology slides of patients diagnosed with IMT in our hospital between 1997-2012 was performed.

Results: Three patients with IMT were identified. All patients had intra-abdominal tumors (omentum, spleen, supra-renal) with one patient also having a concurrent IMT located in the left hemi thorax. Histopathological findings reported positivity for ALK -1 stain in 2 patients. Primary complete surgical resection of the tumors was attempted without success. And hence adjuvant chemotherapy was administered in all cases. A variety of chemotherapeutic agents in combinations were used as per physician preference. Two patients also received monotherapy with non-steroidal anti-inflammatory drug (NSAID) when the tumors did not respond to chemotherapy. Both the patients on NSAID therapy showed transient partial responses. One patient with aggressive IMT died due to neutropenic sepsis. The mTOR inhibitor sirolimus was started on one patient who is now alive with stable disease three years after commencement. A local recurrence in one patient was treated with further surgery and she now remains in complete remission. These two patients remain on active follow up.

Conclusion: Sirolimus appears to offer disease control in children with inoperable or resistant IMT. Further studies are required to understand and manage these fascinating mesenchymal tumors better.

ST-1_V1.6

OUTCOMES OF SURGERY FOR RENAL TUMOURS WITH INTRAVASCULAR EXTENSION

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Aim: The aim of this study was to review the management of children with Wilms' tumour who had intravascular thrombus.

Material and methods: The data regarding presentation, treatment received, surgical management, complications and outcomes were evaluated. All patients received neoadjuvant chemotherapy except in two.

Results: The study cohort included 31 patients with intravascular thrombus treated from 2006 to 2015. The thrombus extent at presentation was: Infrahepatic inferior vena cava (IVC) -19, retrohepatic IVC -6, suprahepatic IVC -1 and Atrium -5. There was complete clearance of IVC in 11/31 patients with neoadjuvant chemotherapy. Regression of the thrombus occurred in 17/31 patients. Due to regression of the tumor thrombus from the atrium, cardiopulmonary bypass could be avoided in 2 patients. In all patients the thrombus had to be dissected off from the tunica intima due to

dense fibrosis around it. The only major complication was massive bleeding in one patient with atrial thrombus. There was no perioperative or 30-day postoperative mortality. The 3-year OS and EFS was 89.3% and 77.8% respectively.

Conclusions: Intravascular tumor thrombus extension has favourable outcomes after contemporary multidisciplinary treatment. Chemotherapy aids in surgery, with tumor regression and thus may obviate the need of cardiopulmonary bypass in atrial thrombus.

ST-1_V1.7

PEDIATRIC MEDULLOBLASTOMA: EXPERIENCE AT TATA MEDICAL CENTER, KOLKATA

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Background: Medulloblastoma is the commonest pediatric malignant brain tumor. Cure-rates of ~75% have been achieved in the West. Indian centers have reported inferior survival. The aim of the study was to analyze the clinical profile and outcome in children treated at a new referral cancer hospital in Eastern India.

Methods: A retrospective analysis of case records of children (<18-years) diagnosed with medulloblastoma, between June 2011-July 2016, was performed. Modified Chang staging, using MRI of brain and spine, and analysis of cerebrospinal fluid was used. Children aged >3-years, with post-operative residual tumor <1.5cm², M0 stage, and non-large cell/anaplastic histology, were classified as 'standard-risk.' 'Standard-risk' and 'high-risk' children received cranio-spinal irradiation with 23.4Gy in 13 fractions, and 36Gy in 20 fractions, over 3.5-4 weeks, respectively. Posterior fossa and/or tumor bed was boosted to a total dose of 55.8Gy in 31 fractions. Following radiation, 'standard-risk' children received 8 cycles of cisplatin, lomustine and vincristine. 'High-risk' patients received 8 cycles of cisplatin, cyclophosphamide and vincristine, or, ifosfamide, cisplatin and etoposide (physician's discretion). Analysis was performed using IBM-SPSSv20. Kaplan-Meier method was used for survival analysis.

Results: Twenty-six children were enrolled. Median age was 6-years (range:0.9-13.5); 4 (15.3%) were ≤3-years at diagnosis. 54% were males. Median symptom-interval was 3.7-months (range: 1-12). Symptoms included vomiting (20; 77%), headache (17; 65%), unsteadiness (14; 54%) and cranial nerve palsy (2; 7%). Location of tumor (n=22) included midline, cerebellum (9; 41%), lateral cerebella (5; 23%) and roof of 4th ventricle (8; 36%). Modified Chang staging: T (n=17): T1 (1; 6%), T2 (2; 12%), T3 (5; 29%), T4 (9; 53%), and, M (n=22): M0 (9; 41%), M1 (3; 13.5%), M3 (7; 32%), M4 (3; 13.5%).

Seven (32%) children were standard-risk and 15 (68%) were high-risk; data was missing for the rest. Eight (31%) refused treatment. Extent of surgical resection (n=16) included gross total resection: 7 (44%), near total resection: 3 (18.5%), sub-total resection: 6 (37.5%). A ventriculo-peritoneal shunt had been inserted in 11/20 (55%). Median time from surgery to initiation of radiotherapy was 41-days (range:34-51).

At a median follow-up of 28.5-months (range:0.1-55), 12 (46.5%) were alive, 9 (35%) of whom had completed, and 3 (11.5%) were on treatment; 3 (11.5%) had relapse/progression, 2 (7.5%) had abandoned treatment, while 1 (3.5%) had received palliative care. Among the 4 children ≤3-years, 1 had disease progression and 3 refused treatment. 4-year EFS (for those who completed therapy was 66.1 ± 1.2%; standard-risk: 100%, high-risk: 45 ± 1.8%. Treatment-related complications included posterior fossa syndrome (7; 39%), sepsis (4; 22%), shunt obstruction (1; 5%) and hearing loss (4; 22%).

Conclusion: Limitations of the study included high prevalence of treatment-refusal/abandonment (10; 38%) and non-availability of molecular subtyping for more accurate risk stratification. Nevertheless, this study shows that when properly diagnosed and treated, the outcomes of children with standard risk medulloblastoma in India are comparable with the west. Further improvements in outcome require providing social support to reduce treatment-refusal/abandonment, standardization of care for