A biopsy reported a small round blue cell neoplasm [Figure 1]. Immunohistochemistry showed diffuse strong positivity for S100 [Figure 2] and Synaptophysin [Figure 3]. It was negative for Cytokeratin, Desmin, Leukocyte common antigen, CD99 and Vimentin, consistent with Esthesioneuroblastoma, Hyams Grade 2. Magnetic resonance imaging (MRI) [Figure 4] showed a lesion involving primarily left nasal cavity and bilateral posterior ethmoidal sinuses. The nasal septum, osteomeatal complex and ostia of frontal sinuses were also involved with occluded ostium of frontal sinuses leading to accumulation of secretions in left maxillary sinus and frontal sinus. MRI also showed extension to the left orbit, optic canal, cavernous sinus, and anterior cranial fossa. She was staged as Kadish Stage C.

After discussion in the multi-disciplinary tumor board, it was decided to offer her Neoadjuvant chemotherapy and adjuvant radiotherapy. She received chemotherapy with VAdrC–IE protocol (Vincristine 2 mg/m² Day 1, Adriamycin 75 mg/m² Day 1, Cyclophosphamide 1200 mg/m² Day 1, Ifosfamide 1800 mg/m² D1–D5, Etoposide 100 mg/m² Day 1-day 5 Q 3 weekly). She was reassessed with a CT Brain after six cycles, which revealed near complete response to chemotherapy [Figure 5]. Radiation was delivered using a 3D conformal plan to a total dose of 6000 cGy in 30 fractions. The target volume included the pre-chemotherapy tumor volume with margins. She tolerated treatment well except for Grade 1 skin reactions. She has been on regular follow-up with annual CT scans [Figure 6]. With 60 months of follow-up she continues to be disease free without any delayed complications of therapy.

**Discussion**

Esthesioneuroblastoma is a rare tumor originating in the upper nasal cavity. Craniofacial resection with adjuvant radiation is the...
most accepted approach in resectable cases. Limited surgery with non-craniofacial resection followed by chemotherapy and radiation have also been reported.\[3\] Chemotherapy along with radiation has been used in the management of Esthesioneuroblastoma in an effort to decrease the morbidity of surgery with mixed results. Esthesioneuroblastoma is moderately radiosensitive to post-operative doses of 60-66 Gy. Neoadjuvant chemotherapy followed by radiation and craniofacial resection has been advocated by the University of Virginia for Kadish Stage C.\[4\]

At Harvard a nonsurgical approach included neoadjuvant chemotherapy using Cisplatin and Etoposide followed by proton therapy showing excellent results.\[5\]

Mishima et al. achieved a complete response in eight out of 12 patients with an aggressive multi agent chemotherapy schedule.\[6\] Turano et al. reported a case successfully treated
using the same regimen, alternating Cisplatin Etoposide with Doxorubicin, Ifosphamide and Vincristine.\(^7\)

We however chose to use VAdrC IE, a schedule commonly employed for primitive neuroectodermal tumors. Our patient, an adolescent girl, the first ever reported case with this chemotherapy schedule showed an excellent response, which was consolidated with radiotherapy. This case report highlights the use of definitive chemoradiation in Esthesioneuroblastoma as a possible curative option. Further avenues of research are needed to demonstrate the efficacy of chemo radiation in this rare neuronal cancer.

**References**