ALVEOLAR Rhabdomyosarcoma of Sinonasal Tract

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Submitted: 20 December 2016 / Accepted 25 December 2016

A biopsy of 27 years old male was received with a history of bloody nasal discharge for last 1 year, and sinusonal growth. Tissue was composed of multiple pieces collectively measuring 3 x 3 cm in aggregate. Histologically, tumour was composed of round blue cells arranged in nested pattern. The tumor cells showed scanty cytoplasm, moderate nuclear pleomorphism, hyperchromatic nuclei and indistinct nucleoli. Brisk mitotic activity was noted. (Figure) No spindled cells, myxoid change and necrosis was seen.

Immunohistochemically, the tumor cells showed strong cytoplasmic desmin positivity. Tumor cells showed negative expression of CK, P40, NSE, Calretinin, Synaptophysin. In situ hybridization for EBV-encoded RNA was also negative (Figure). Final diagnosis was rendered as alveolar rhabdomyosarcoma.

The differential diagnosis of round blue cell tumors in Head and neck region include rhabdomyosarcoma, olfactory neuroblastoma, sinonasal carcinoma, ewing’s sarcoma, small cell carcinoma and lymphomas. In this case, tumor cells showed nested pattern and strong desmin positivity favoring alveolar rhabdomyosarcoma.

Rhabdomyosarcoma is a malignant tumor of mesenchymal tissue derivation. It represents most common soft tissue sarcoma of pediatric population[1]. Among alveolar and emryonal rhabdomyosarcoma, alveolar most commonly occur in adolescents than young children[2].

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Approximately 40 percent of rhabdomyosarcoma involve head and neck region. Lesion in sinonasal tract commonly presents as symptoms of sinusitis, cranial nerve palsies, proptosis and mass[3]. Immunohistochemically, rhabdomyosarcoma show positivity for desmin, myogenin, myoD1, and muscle specific actin.

Histologically, alveolar rhabdomyosarcoma cells show cohesive nests of round cells, outlined by fibrous septa with picket row configuration. Alveolar rhabdomyosarcoma has reciprocal translocations which are absent in embryonal rhabdomyosarcomas. These translocations, t(2;13)(q35;q14) and t(1;13)(p36;q14), generate the fusion genes PAX3–FOXO1 and PAX7–FOXO1, respectively[2]. The treatment modalities for alveolar rhabdomyosarcoma include surgery, chemotherapy and radiotherapy. Prognosis depends upon stage and histological type. As compared to embryonal rhabdomyosarcoma, alveolar type behave in more aggressive fashion[4].

References:
Figure: (A) Tumour arranged in nests, composed of round blue cells with moderate cytoplasm, hyperchromatic nuclei and indistinct nucleoli. (B) High power view of tumour cells with nuclear pleomorphism and few rhabdoid cells. (C) Desmin strongly positive in tumour cells showing cytoplasmic staining pattern. (D) Tumour cells negative for CK, P40, Calretinin, Synaptophysin, NSE and EBER.