CASE REPORT

RHABDOMYOSARCOMA OF MAXILLARY SINUS IN AN ADULT FEMALE

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HOW TO CITE THIS ARTICLE:
Annapurna Rao B, Ajay Kumar B, Krishna Prasad P, Rama Koteswara Rao N. "Rhabdomyosarcoma of Maxillary Sinus in an adult Female". Journal of Evidence based Medicine and Healthcare; Volume 2, Issue 22, June 01, 2015; Page: 3365-3369.

ABSTRACT: Rhabdomyosarcoma of Maxillary sinus is a rare tumor usually occurring in children. A 30 year old adult female patient attending the ENT department of our Hospital presented with rapidly progressing tumor arising from Left maxillary sinus. It was extending into the left orbit, Nasal cavity and oropharnyx as seen in the CT scan. Histopathology and Immuno-histochemistry positive to CD-99, Desmin and Myo-D1 confirmed the diagnosis of Rhabdomyosarcoma. Five cycles of chemotherapy regimen with Vincristine, Adriamycin and Cyclophosphamide lead to complete resolution of tumor and regression of Proptosis of left eye.
KEYWORDS: Rhabdomyosarcoma, Maxillary sinus, Adult female.

INTRODUCTION: Squamous cell carcinomas are the most commonly occurring malignant tumors of maxillary sinus. Rhabdomyosarcoma is common in children and younger age groups. The present case of Rhabdomyosarcoma of Para nasal sinus is presented in view of its rarity as it presented in a young female patient.

CASE REPORT: A 30 year old female presented with Proptosis of left eye (Fig. 1), Diplopia in left eye, left nasal obstruction, discharge from the nasal cavity and severe pain in the orbit since of 3 months. These complaints started one month following the removal of an Antrochoanal polyp at a rural Hospital. Histopathology was not done at that time. There was no family history of malignancy and no previous history of exposure to radiation in the patient.

A thorough ENT examination was done. On Anterior rhinoscopy sero sanguineous fluid was seen in the left nasal cavity along with a fleshy polypoidal mass in the left middle Meatus. On posterior rhinoscopy a mass was shown entering the oropharnyx from nasopharnyx. A diagnostic nasal endoscopy confirmed the above findings. Biopsy was taken from mass in the left nasal cavity and sent for Histopathological examination.

Earlier Diagnostic CT scan of PNS showed near to total opacification of left maxillary sinus with widening and obstruction of left meatal complex. There was extension of polypoidal soft tissue mass via left Osteo-meatal complex into left nasal cavity obliterating it. The mass was protruding into the oropharnyx through the left posterior choana. Laterally there was thinning and erosion of lateral bony wall of the left maxillary sinus with extension of soft tissue into left peri-sinus fat plane and obliterating it. Superiorly there was thinning and erosion of floor of left orbit with extension of soft tissue into the left orbit with loss of fat plane adjacent to recti muscles. Proptosis of eye ball was noted.

MRI scan (Fig. 2) of Para nasal sinuses was undertaken to help in staging the tumor before starting the treatment. It showed a mass measuring 2.7x3.8x4.3cm (APxTVxCC) with

heterogeneous signal intensity. The mass was seen in the left maxillary sinus with erosion of postero lateral wall. The mass was extending in to infra temporal fossa. Medially the lesion was extending into Pterigo maxillary fossa, Pterygomaxillary fissure, inferior orbital fissure and orbital apex with intra orbital extension measuring 2.6x2.0x2.3 cm. The left maxillary ostium was widened with extension of mass into maxillary ostium, middle meatus, nasal cavity and Nasopharnyx through posterior choana.

Histopathological examination of the biopsied material (Fig. 3) showed fragments of tumor tissue with prominent vascularity associated with wide areas of hemorrhagic necrosis. Most of the tumor cells are round to spindle cell type having high nuclear cytoplasmic ratio in an edematous loose stroma. Focal myxoid areas are seen in the stroma. Histological Grade 2 (3+1+1) of FNLCC grading was assigned to the tumor.

Immuno-histochemistry showed MIC-2, CD-99(Fig. 4), Desmin (Fig. 5) and MYO-D1 (Fig. 6) positivity, whereas Cytokeratin, CD-45, Chromogranin and Synaptophysin were Negative. KI-67 showed positivity in 40% of cells. Thus a diagnosis of Rhabdomyosarcoma was made based on Histopathology and Immuno-histochemistry findings. Hematological parameters, Chest X-ray and ultrasound abdomen showed normal findings. There was no evidence of distant metastasis.

Stage IIB (T2B, N0, M0, G2) of AJCC[1] staging system was assigned to the tumor with grade T2B; consists of tumor diameter >5cm and located in deep location.

Since the tumor was unresectable, Chemotherapy followed by Radiotherapy was advised to the patient. Chemotherapy with Vincristine (1.9mg), Adriamycin (100mg) and Cyclophosphamide (1000mg) was started. After six months of follow up, she received five out six cycles of Chemotherapy. Tumor was completely resolved and Proptosis of left eyeball also resolved (Fig. 7) with mild blurring of left eye vision. Diagnostic Nasal endoscopy showed almost normal left maxillary sinus, oropharnyx and nasal cavities.

DISCUSSION: Malignant tumors of Para nasal sinuses are rare. Of all the Para nasal tumors, 77% of malignant tumors arise in the maxillary sinus.[2] Most common malignant lesion is Squamous cell carcinoma, whereas Rhabdomyosarcoma (RMS) of Para nasal sinuses is rare and occurs mostly in children.

Rhabdomyosarcoma (RMS) is the most common sarcoma in childhood and approximately 40% of Rhabdomyosarcoma occur in Head and neck[3] with about 20% occur in Nasal cavity, Nasopharynx and Nasal sinuses.[4] RMS of the Para nasal sinus accounts for 10% to 15% of adult head and neck RMS.[5] Ahmed and Toskos[6] reported 14 cases of Para nasal sinus Rhabdomyosarcoma, 9 out 14 cases occurred in ethmoidal sinus and 5 cases occurred in Maxillary sinus. Erkul et al[7] reported RMS in 32 year old male in Ethmoid sinus and Tsung-Han Wu et al[8] reported 23 cases of adult maxillary sinus Rhabdomyosarcoma.

Clinical history of rapid progression of tumors in the Head and neck region in children and young adults need to be evaluated thoroughly to rule out various sarcomas. Early diagnosis by histopathology and Immuno-histochemistry helps in rapid initiation of treatment. Histological grade, tumor size and depth are essential for staging.[1] Histological grade of sarcoma is one of the most important parameters of staging system.[1] Multimodality treatment involving surgery, chemotherapy and radiotherapy leads to better outcome with increased survival rates.[8]

CONCLUSION: Para nasal sinuses are uncommon sites for adult Rhabdomyosarcoma. High index of clinical suspicion and early diagnosis by Histopathology and Immuno-histochemistry leads to early start of treatment. Since there are no known etiological factors in our case and also in previously published cases with increasing case reports of adult Para nasal sinus Rhabdomyosarcoma, further studies are needed to evaluate the etiological factors.

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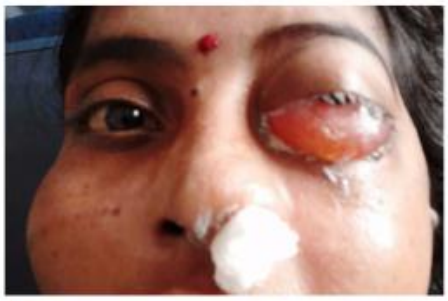

Fig. 1: Proptosis of Left eye

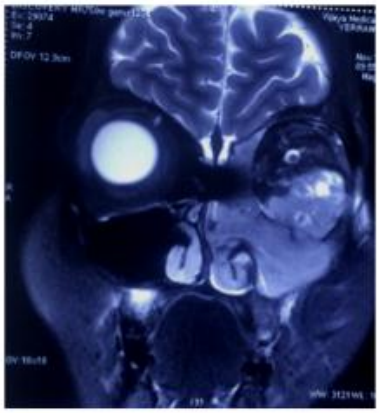

Fig. 2: MRI show heterogenous signal intensity mass seen in left maxillary sinus extending in to left orbit and left nasal cavity.

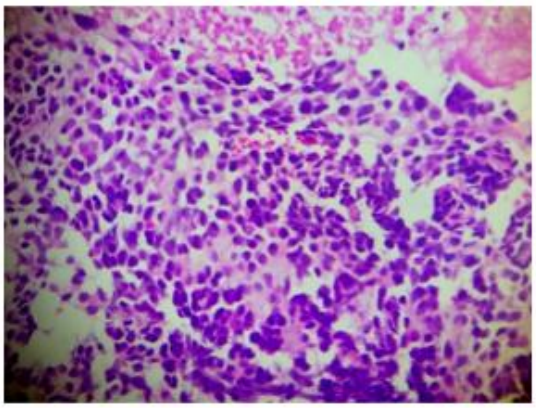

Fig. 3: Microphotograph showing round blue cells arranged in sheets along with prominent vascuarity and necrosis. (H & E; 400x)

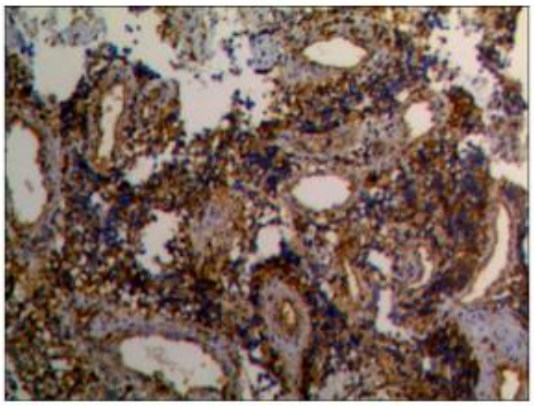

Fig. 4: Microphotograph showing CD-99 positivity in the tumour cells. (IHC; 100x)

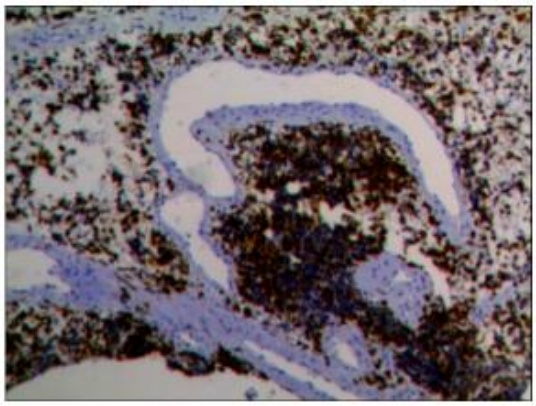

Fig. 5: Microphotograph showing Desmin positivity in the tumour cells. (IHC; 100x)

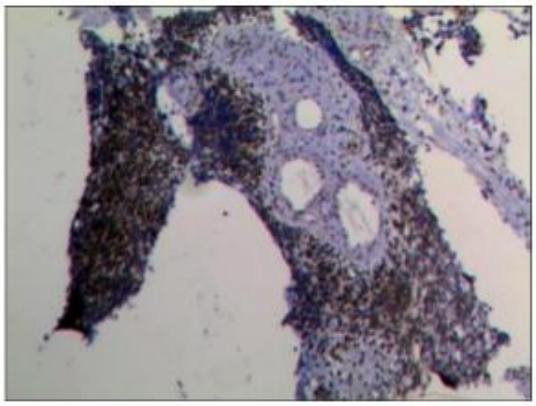

Fig. 6: Microphotograph showing Myo-D1 positivity in the tumour cells. (IHC; 100x)

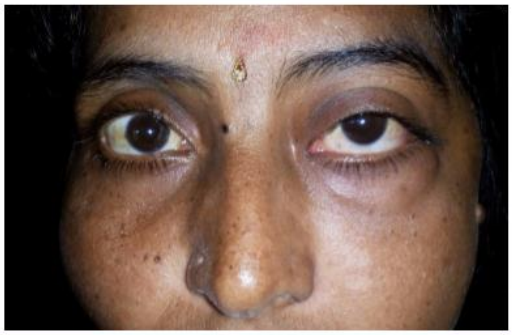

Fig-7: Complete resolution of tumour with normal looking left eye after 5 doses of chemotherapy.

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Date of Submission: 23/05/2015.
Date of Peer Review: 24/05/2015.
Date of Acceptance: 26/05/2015.
Date of Publishing: 01/06/2015.