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A CASE REPORT OF ALVEOLAR RHABDOMYOSARCOMA IN RIGHT PARASELLAR REGION IN AN ADULT

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ABSTRACT

Primary parasellarrhabdomyosarcoma extending to parameningeal region are very uncommon and malignant tumors, that occur predominantly in the posterior fossa of pediatric patients. A 29 years old female presented with right frontal headache and 3rd cranial nerve palsy. MRI showed lesion in right parasagittal region, extending to right pterygo-maxillary fissure and maxillary sinus. She was treated by right frontal craniotomy and right parasellar mass was removed. Immunohistochemistry suggested alveolar rhabdomyosarcoma. Patient has been treated by Intensity Modulated Radio-therapy of 60 Gy in 30 fractions. Patient has completed the treatment and now patient is on chemotherapy and on regular follow up.

Key words: Rhabdomyosarcoma, Intensity Modulated Radiotherapy, Paraseller region, parameningeal region.

INTRODUCTION

Rhabdomyosarcoma is the commonest soft tissue sarcoma in childhood. It is a mesenchymaltumour, having two main histological subtypes, [1] Alveolar, [2] Embryonal. It is more common in children aged upto 5 years. In the pediatric population they are preferentially located in the infratentorial compartment. Only sporadic cases are described in adults. Treatment for these lesions is not yet standardized, but maximal safe resection followed by radiation therapy is widely suggested, followed by chemotherapy [3]. We report the case of a 29 year old female with MRI brain showing lesion in right parasagittal region, extending to right pterygomaxillary fissure and maxillary sinus. She was treated by right fronto-temporal craniotomy and right parasellar mass was removed. Immunohistochemistry suggested alveolar rhabdomyosarcoma. Patient has completed Intensity Modulated Radio Therapy.

CASE HISTORY

A 29 years old female presented with difficulty in vision and ptosis suggestive of 3rd cranial nerve palsy and frontal headache for 6 days. Her MRI Brain was suggestive of lesion at right parasellar region encasing cavernous part of right Internal Carotid Artery involving right orbital apex. Lesion was extending to right masticator space involving right medial and lateral pterygoid muscle causing erosion of posterior wall of right maxillary sinus and medially extends into it. Medially, it also extends into right nasal cavity, right middle turbinate, ethmoid sinus and sphenoid sinus. Laterally, it causes compression over right temporal lobe. The extra-axial component of lesion measures 35*32 mm and the masticator component measures 41*31 mm.

TREATMENT

The patient underwent right frontotemporal craniotomy and subtotal excision on 26/2/2015. Biopsy and Immunohistochemistry suggested Alveolar Rhabdomyosarcoma (Vimentin, Desmin and Actin were positive).

Her POST-OP MRI BRAIN showed lobulated extra axial mass lesion is seen in sellar-parasellarregion,right middle cranial fossa, right sphenoid sinus, right posterior ethmoid air cells ,right orbital apex,

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lateral wall and floor of right orbit. Lesion is extending into inferior orbital fissure into right infratemporal fossa. There is involvement of wall of right orbit, skull base on right side, right pterygoid plate, posterolateral and medial wall of right maxillary sinus. Lesion involves pterygoid fossa, medial and lateral pterygoid muscle and temporalis muscle. It is obliterating right cavernous sinus and meckel's cave and abutting right petrous portion of ICA s/o residual lesion of approx. 51*52*59 mm. Subdural hematoma is seen in right frontal and temporal region maximum width of 8 mm in temporal region.

Metastatic work-up like, bone marrow biopsy, CSF cytology and CT scan abdomen and pelvis were normal. Patient was advised Intensity modulated radiotherapy as a modality of treatment and planning was done using Oncentra treatment planning system. The dose to the Planning Target Volume was 60 Gy in 30 fractions by 2 Gy per fraction. Then the patient was referred to medical oncologist for adjuvant chemotherapy. Patient is given injection VCR 2 mg (1.4 mg/m^2) intravenously on Day 1, injection Actinomycin D 1 mg(.075mg/kg) in 500 cc normal saline on days 1 to 5 upto maximum of 1 mg/kg and injection Cyclophosphamide 1800 g in 500 cc normal saline.

Fig 1. CT- scan of brain image showing disease

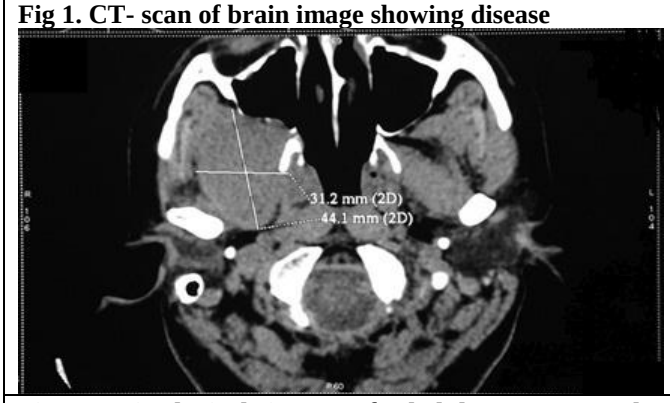


Fig 2. MRI- brain image showing disease

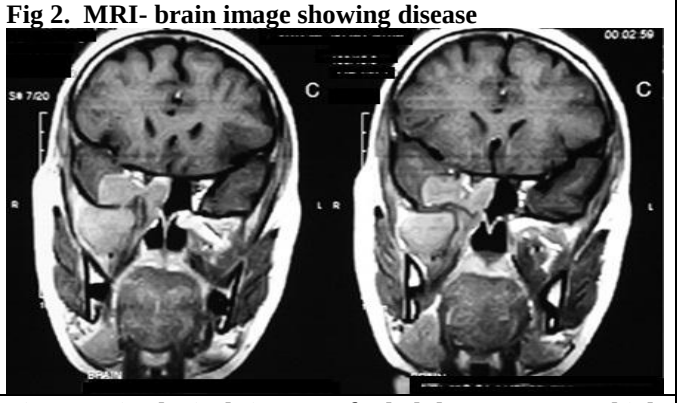


Fig 3. Histological image of rhabdomyosarcoma-low power

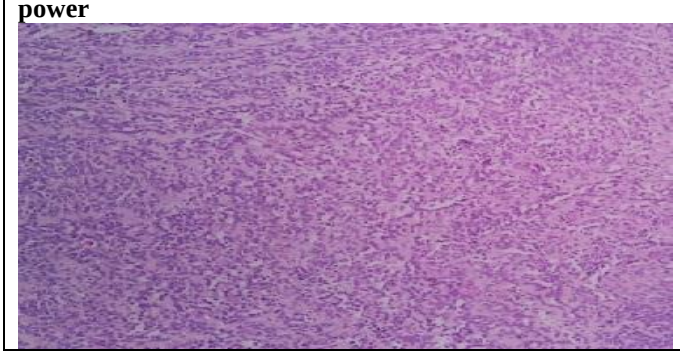
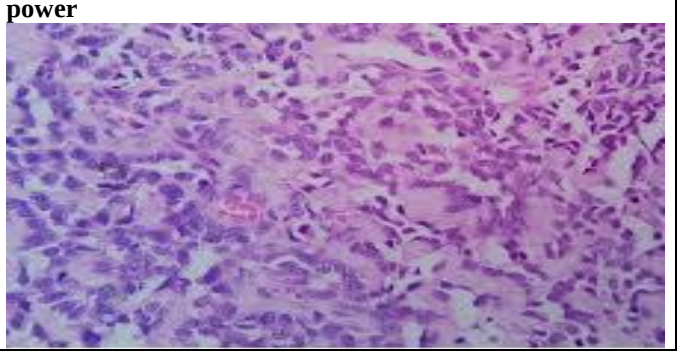


Fig 4. Histological image of rhabdomyosarcoma-high power



DISCUSSION AND CONCLUSION

Rhabdomyosarcoma is a highly malignant soft tissue sarcoma that arises from unsegmented, undifferentiated mesoderm or myotome-derived skeletal muscles. It may occur at any site of body like orbit, head and neck, genitourinary, extremities, trunk and parameningeal area. Rhabdomyosarcoma are more common in children and uncommon in adults. Adult RMS has very poor outcome and must be treated aggressively using multimodality treatments. There are histological subtypes of rhabdomyosarcoma like alveolar, embryonal, botryoids, anaplastic, spindle cell and undifferentiated [4].

Primary cerebral rhabdomyosarcoma extending to parameningeal area is mainly a disease of childhood. It is uncommon in adults and its treatment is mainly surgery and followed by radiotherapy which is very difficult due to the critical organs which are present nearby. According to literature available till date there are very few cases of adult cerebral rhabdomyosarcoma [5]. Rhabdomyosarcomas constitute a unique group of soft tissue neoplasms that share a propensity to undergo myogenesis, a well-defined biologic process that primarily occurs during embryonal and fetal development. Rhabdomyosarcomaof brain has to be differentiated from an intracranial extension from skull or para-meningeal sites (orbit, nasopharynx, paranasal sinuses, middle ear, external auditory canal), but also from a metastatic seeding from a systemic rhabdomyosarcoma and from mixed primary cerebral tumors of the central nervous system in which rhabdomyoblastic areas are found in combination with sarcomatous, neuroectodermal, mesenchymal or

teratomatousfeatures. This tumor is thought to derive from myogenic precursor cells and belongs to the group of small round blue-cell tumors. The rhabdomyosarcoma must be differentiated from other brain tumours. For confirmation of the diagnosis Immunohistochemistry is must. Different Immunohistochemistry markers are actin, vimentin, desmin and myogenin.

Rhabdomyosarcoma is treated with multimodality approach which includes surgery which is followed by chemotherapy followed by radiotherapy at 9 weeks of treatment except in patients requiring emergency radiation treatment at day 0 or immediately after surgery for spinal compression and those who had high risk parameningealfeatures(direct intracranial extension, base of skull erosion, or cranial nerve palsy). In patients with primary tumours originating from cranial-parameningeal sites(nasopharynx-nasal cavity, paranasal sinuses pterygopalatine-infratemporal fossa)examination for involvement of meninges or meningeal impingement is must by either MRI or CT scan or by CSF cytology. The prognosis depends on the primary site, the histological subtype and tumour size. Good prognosis for survival are; tumour to be smaller than 5 cm, the patients under 20 years of age, lack of regional or distant metastasis and negative surgical margins [2,3]. Here is a summary of few cases of RMS brain in adults [5].

Differential diagnosis should include high-grade glioma, lymphoma, metastases and PNET. Histological confirmation is essential for the definitive diagnosis. According to Intergroup Rhabdomyosarcoma Study-4: results for patients with nonmetastatic disease patients of stage 3 of IRSG presurgical staging system are to be treated with sugery first if possible otherwise radiotherapy from day 0. In all stages of rhabdomyosarcoma the most common modality to improve prognosis is local control. The sellar and parasellar region is an anatomically complex area that represents a critical junction for important contiguous structures like, normal brain, eyeballs lens optic nerve, inner ear, brain stem etc. making the radiation therapy difficult. So, here we planned intensity modulated radiotherapy for this patient.

We gave 2 Gy per fraction total 60 Gy to planning target volume by photons using 9 beams. The maximum dose which the target received was 66.93Gy . The doses to lens, eyeball and optic nerve were well within limit.

The patient well tolerated the treatment with some side effects of oral ulceration and headache which were controlled by symptomatic medications. There was improved vision which was reduced because of the disease during treatment. After radiotherapy the patient is to be given chemotherapy as follows (given with response rate forrhabdomyosarcoma)vincristine 59%, dactinomycine 24%, cisplatin15% to 21%,dacarbazine 11%,mitomycin 36%,etoposide 15% to 21%,ifosfamide 86%,irinotecan 23% and topotecan 46%. Various regimens used are VAC that is vincristine, Adriamycin and cyclophosphamide and or ICE (ifosfamide, carboplatin and etoposide). The prognosis depends on the primary site, the histological subtype and tumour size [6]. Good prognosis for survival are; tumour to be smaller than 5 cm, the patients under 20 years of age, lack of regional or distant metastasis and negative surgical margins [2,3]. Tumours of the orbit have a better prognosis. Parameningeal RMS has more recurrence rates and has much more early metastasis because of the possibility of intracerebral spread [1]. Distant metastasis occurs in adults at the time of diagnosis in 60% of cases [1]. The 5-year survival rates are 56% to 65% [1].

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CONFLICT OF INTEREST:
The authors declare that they have no conflict of interest.

REFERENCES

1. Crist WM, Anderson JR, Meza JL, Fryer C, Raney RB, *et al*. Intergroup rhabdomyosarcoma study-IV: results for patients with nonmetastatic disease. *J ClinOncol*, 19, 2001, 3091-3102.
2. Simon JH, Paulino AC, Smith RB, Buatti JM. Prognostic factors in head and neck rhabdomyosarcoma. *Head Neck*, 24, 2000, 468-473.
3. Vania P, Paolo CC, Massimo T,*et al*. Primary cerebral alveolar rhabdomyosarcoma in adult
4. Ulutin C, Bakkal BH, Kuzhan O. A Cohort Study of Adult Rhabdomyosarcoma: A single Institution Experience. *World Journal of Medical Sciences*, 3, 2008, 54-59.
5. Montone KT, Barr FG, Zhang PJ, Feldman MD, LiVolsi VA. Embryonal and alveolar rhabdomyosarcoma of parameningeal sites in adults: a report of 13 cases. *Int J SurgPathol*, 17, 2009, 22-30.
6. Wu TH, Huang JS, Wang HM, Wang CH, Yeh KY. Long-term Survivors of Adult Rhabdomyosarcoma of Maxillary Sinus Following Multimodal Therapy:Case Reports and Literature Reviews. *Chang Gung Med J* , 33, 2010, 466-471.