Pakistan Journal of Otolaryngology 2014; 30 : 24-25

Case Report

Nasopharyngeal Angiofibroma with Intracranial Extension; Post Radiotherapy Down Staging and Surgical Excision

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ABSTRACT: Nasopharyngeal angiofibroma is a locally aggressive, histologically benign, vascular neoplasm. This neoplasm accounts for 0.05% of head and neck tumors and affects almost exclusively male adolescents. Surgery is considered as the primary treatment of nasopharyngeal angiofibroma. Other treatment modalities such as radiotherapy and chemotherapy are still recommended for intracranial extension involving the cavernous sinus or the internal carotid artery. We report a rare case of nasopharyngeal angiofibroma of stage 4b with intracranial extension. The treatment consisted of radiotherapy and post radiation down staging of tumor. New stage 4a is then treated with surgical excision post angioembolization via lateral rhinotomy approach.We discuss the role of radiotherapy in down staging the tumor and making it possible to excise the tumor effectively in advance stage 4b of nasopharyngeal angiofibromas.

Key Words: Nasopharyngeal angiofibroma, Intracranial extension, Lateral rhinotomy, Down staging tuuor.

INTRODUCTION: Nasopharyngeal angiofibroma is a slow growing, locally aggressive, histologically benign vascular tumor affecting the adolescent males. Incidence showed it as 0.05% of all head and neck tumor. Various treatment modalities like surgery, radiotherapy, embolization, chemotherapy and hormonal therapy are used and each has its own limitations. Radiotherapy is the treatment of choice for patients of advance stage. This case is reported due to its rarity, as it was an advance stage lesion and radiotherapy was given to patient. On followed up the patient showed a good response in terms of hemostasis, reduction in size and down staging thus making it possible to remove surgically. This case has given us astonishing results and made an advance stage of untreatable case to treatable with using radiotherapy as prime modality and then surgical excision.

CASE REPORT: A 18-year young boy presented in January 2010 with history of right sided nasal blockage, bleeding and purulent discharge from right nostril for last 1 year. Examination showed patient was anaemic in his general physical examination and on local examination there is widening of the bridge of the nose. Anterior rhinoscopy showed purulent discharge in the right nasal cavity and reddish lobulated mass occupying whole right side. Posterior rhinoscopy showed mass occupying right half of nasopharynx. Decreased nasal patency of right side. Sense of olfaction was absent on right side. Other ENT exam was unremarkable. A computerized tomography (CT) scan with contrast and MRI done that showed large lobulated vascular mass around right pterygopalatine fossa extending into nasal cavity, nasopharynx on right side, infratemporal fossa through pterygomaxillary fissure, intracranially extending across the base of skull, eroded sphenoid sinus and involving right cavernous sinus.The mass in its greatest dimensions was 75 X 56 mm.The final diagnosis was stage IVb nasopharyngeal angiofibroma with to intracranial extension (Fig-1). In view of the intracranial extension, the patient was not considered for surgical resection and hence radiotherapy was started. External beam radiotherapy was delivered through a wedge pair of anterior and oblique co-planar cobalt beams to a dose of 4600 cGy/ 23 Fr /5Fr/wk/ 31 days after doing a CT simulation. The therapy schedule was uneventful. Patient was reviewed six month later because he was lost to follow up by patient as the patient is from a distant place. A follow up CT and MRI scan was done. Repeated scan showed mass in the region of nasopharynx predominantly on the right side extending in to infratemporal fossa through pterygomaxillary fissure, eroded sphenoid sinus but no involvement or extension seen in cavernous sinus. Findings are consistent with previously known diagnosis of angiofibroma. It measured 45 X 42 mm.Now the stage of the tumor was found to be as stage 4a (Fig-2). Then as a protolcol of treatment of stage 4a, angioembolization was done and excision of lesion was done with utilizing lateral rhinotomy approach (Fig-3). Patient is now on regular follow up and is stable.

DISCUSSION: Nasopharyngeal angiofibroma is type of a slow growing, locally aggressive, histologically benign vascular tumor affecting the adolescent boys. This exclusive occurrence among boys is probably linked to the androgen. The site of origin is thought to be either from the superior aspect of the sphenopalatine foramen or from the pterygo-palatine fossa, in the recess behind the sphenopalatine ganglion at the exit of the pterygoid canal[1]. A clinical triad of unilateral nasal obstruction, recurrent spontaneous epistaxis and nasal drainage is diagnostic of nasopharyngeal angiofibroma. Most often biopsy is not done as it is highly vascular but a biopsy done as in our case confirms the diagnosis. Three classification systems, used to stage nasopharyngeal angiofibroma are Fisch's, Chandler's and Radowski's adaptation of Sessions' classification are essentially based on the extent of the disease[2]. Various treatment modalities like cryotherapy, sclerosing therapy, hormonal treatment, radiotherapy[3], chemotherapy, embolization

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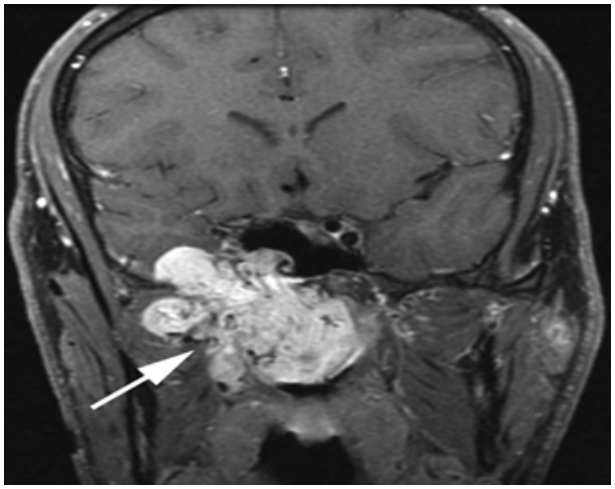

Figure-1: Scan of patient showing stage 4-B.

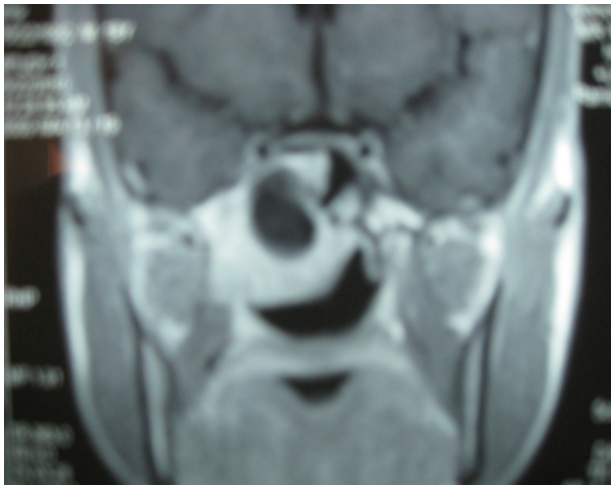

Figure-2: Scans Repeated After Radiothrapy Showing down Staging.

and surgery[4,5] are used to treat nasopharyngeal angiofibroma. Since it is rarely seen, there are no standard guidelines available in the literature especially for advanced cases. Essentially treatment depends on the physician preference, available expertise and patient preference. Surgery is used to treat nearly two thirds of the patients. Endoscopic removal is also considered for selected cases with preoperative embolization to control bleeding at the time of removal[6,7]. Radiotherapy offers a very good local control rate, as high as 80-85% with the tumors regressing very slowly over two to three years. McAfee et al. have reported 91% control with a dose of 30-36 Gy and have not seen any complications of radiotherapy in a follow up of two to five years[8]. Nasopharyngeal angiofibroma with advance stage disease is treated with radiotherapy. Tumors invading cavernous sinus considered inoperable because of bleeding from cavernous venous plexus and potential injury to the internal carotid artery or cranial nerves. Surgical defects in the dura and possibility of uncontrollable cerebrospinal fluid leak. A dose of 36-46 Gy is recommended over a period of 4-5 weeks. Endocrine hypo function, cataract formation and second malignancies, the possible side effects of radiation have to be kept in mind[8] and hence radiotherapy needs to be used only when surgery is not possible or in residual and recurrent lesions. State of art radiotherapy in the form of 3DCRT, IMRT and proton therapy may help to reduce the late radiation toxicities. In this case radiotherapy is used as prime treatment. Patients stage of tumor was changed, making it possible for us to remove it surgically as well.

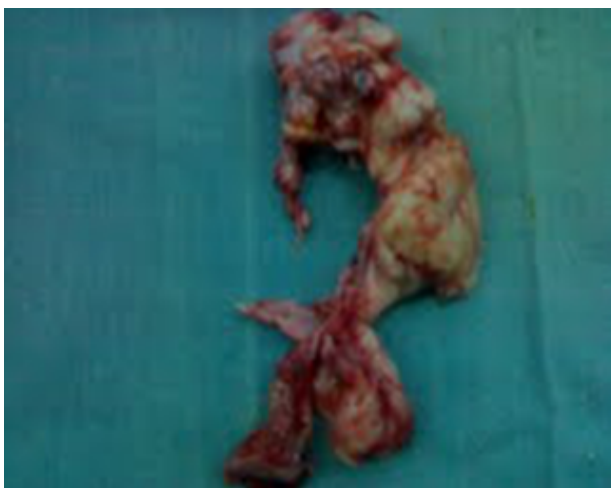

Figure-3 : Tumor excised via lateral rhinotomy apporach.

CONCLUSION: We presented this case of invasive angiofibroma stage 4-b.Which is usually inoperable, but by using the radiotherapy modality the scenario was changed and disease became curable with surgical removal. This benign vascular tumor is having a tendency to invade locally. There are multiple treatment options. In this case we have used the radiotherapy modality to down stage the disease and change the state of patient.By using radiotherapy we find out that inoperable stage of disease became operable.

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