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CASE REPORT

Recurrent Trigeminal Schwannoma with Parapharyngeal Extension: A Rare Cause of Facial Pain

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ABSTRACT

Trigeminal Schwannoma is a rare benign tumor of less than 1 % of all intracranial tumors. Trigeminal nerve involvement is the second most common in the head and neck region. They are slow growing and usually arise from sensory divisions of the trigeminal nerve. Orofacial pain and swelling are key clinical features of motor loss in advanced disease. Magnetic resonance imaging (MRI) is diagnostic. Surgery with wide margins is the mainstay of treatment along with adjuvant radiotherapy. We report a case of recurrent Trigeminal Schwannoma in a young female presented as recurrent orofacial pain and swelling. She had undergone surgery a few months age without adjuvant radiotherapy. MRI showed an abnormal signal area, which was very locally advanced reaching to the oral cavity, para pharyngeal space and infratemporal fossa. She was advised against resurgery and definitive radiotherapy was suggested but patient did not pursue due to unknown reasons.

Keywords: Orofacial Pain, Para Pharyngeal Space Extension, Trigeminal Schwannoma.

INTRODUCTION

Trigeminal Schwannoma is a rare benign intracranial neoplasm and its incidence approximately ranges between 0.07-0.36 percent of all intracranial tumors.[1] Like any schwannoma elsewhere in the body, trigeminal schwannoma arises from Schwann cells mostly from perineural sheath of the nerve. It is more common in head and neck region. Vestibulocochlear nerve is the commonest nerve in the head and neck involved by the schwannomas while trigeminal nerve is the second most common nerve to be involved but still very rare.[2] Infrequent malignant transformation in this benign tumor has been reported in the literature and its outcome is associated with dismal prognosis. Trigeminal schwannomas commonly arise from sensory divisions of the nerve like ophthalmic and maxillary divisions and they are slow growing and invade locally in the adjacent tissues or organs.

Oro-facial pain involving the skull, face and cervical region is the commonest clinical presentation due to the rich nerve supply of Vestibulocochlear nerve.[3] Motor functions loss may be the consequences of advanced disease.

The preferred modality of imaging is Magnetic Resonance Imaging (MRI) for this tumor. MRI shows an isointense mass when compared to the brain in T1-weighted images, with homogenous enhancement after the administration of contrast medium, while in T2-weighted images the lesion is hyperintense.[4]

The mainstay of the treatment is early surgery with wide margin with or without post-operative radiotherapy. Definitive radiotherapy via 3D Conformal Radiotherapy (3DCRT), Intensity Modulated Radiotherapy (IMRT) or radiosurgery may be applied in very locally advanced inoperable cases to achieve local control and improvement in survival. The primary goal of surgery is to completely remove the tumor by intracranial and intranasal approach, however small tumors localized in Meckel cave may be excised via endoscopic approach. As a response to radiotherapy, this benign tumor continues to regress slowly after radiotherapy for about 3 to 4 years to complete resolution.

CASE REPORT

We report a case of 38 years old woman with no known comorbid presented with gradually increasing dull type of mild to moderate pain in the left half of the face which was non-radiating and continuously present involving the cutaneous innervation of all three divisions of trigeminal nerve along with off and on tingling sensation in the same area, swelling and electrifying sensations with few symptoms free intervals.

Past history was consistent with recurrent left trigeminal Schwannoma. She first presented with only left side facial pain in September 2020 and excision was done in October 2020. Histopathology was trigeminal schwannoma and 1.1 cm tumor was excised. Margins were not commented due to fragmented nature of specimen. No adjuvant treatment was received at that time. She lost follow up after surgery and remained symptom free. In February 2021 she had recurrence of left sided facial pain and she underwent second craniotomy and excision in April 2021 in a government hospital neurosurgery department. Post-operative recovery was uneventful. After surgery, patient had second recurrence of symptoms and presented in our hospital OPD with a post-operative MRI scan brain.
No history of addiction. No systemic symptoms or signs. She is a house wife with 2 normal kids and poor socioeconomic status.

Examination

On examination she was vitally stable. Well oriented with time, place and person. There was asymmetry of the face due to a soft non-tender swelling involving left temporal region, left upper and lower face. Mouth opening was 3 cm and there was intra oral hard swelling 2 x 3 cm arising from left side of maxilla protruding in the oral cavity without mucosal erosion. There were Impalpable lymph nodes in the neck bilaterally. Vision was normal. No signs regarding cranial nerves palsies were found. Patient picture is shown in Figure 1 showing left face swelling. No cutaneous manifestation of neurofibromatosis was seen. Systemic examination was unremarkable.

Histopathology

Her second surgical histopathology revealed multiple pieces of tumor tissue aggregately measuring 2.4 x 1.2 cm. Immunohistochemical stains were performed and S-100 and epithelial membrane antigen were positive while glial fibrillary acidic protein and PR were negative. It was concluded as trigeminal schwannoma. No comments over margin done due to the piecemeal specimen.

Imaging

MRI brain after second surgery with contrast done on 08.06.21 showed heterogeneously enhancing abnormal signal mass involving the parasellar region and Meckel's cave inseparable from cavernous sinus. Mass was causing effacement of left temporal lobe, posteriorly extending to cerebello-pontine angle and compressing the pons, effacement of cerebral aqueduct extending to base of skull with effacement of infratemporal fossa and displacement of posterior maxillary wall also extending to para-pharyngeal space and narrowing of nasopharyngeal lumen. It was hypointense on T1 weighted images as in Figure 2 showing tumor extension into para-pharyngeal space and infratemporal fossa and hyperintense on T2 weighted images with post contrast enhancement as in Figure 3 showing tumor in close proximity with the brain stem. It measured 5.7 x 4.4 x 7 cm and showed disease progression as compared with previous scan of April 2021. Figure 4 also shows T1 weighted MRI coronal section with tumor extension up to the oral cavity.

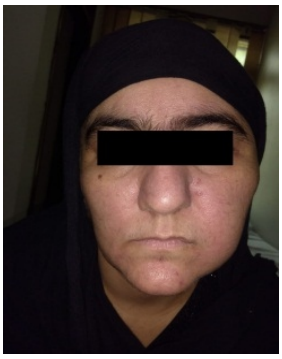

Figure 1: Patient picture showing swelling of left side of face

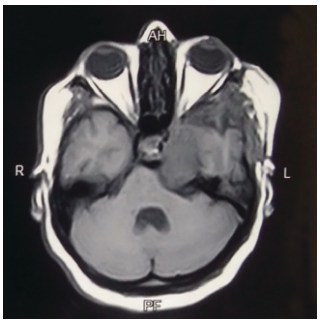

Figure 2: T1 weighted MRI image showing tumor invasion into para-pharyngeal space and infratemporal fossa

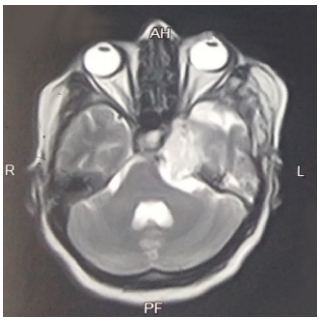

Figure 3: T2 weighted MRI image showing tumor close proximity with brain stem

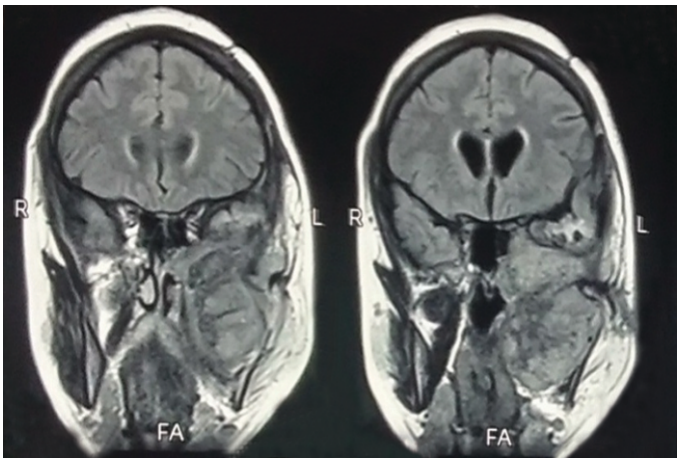

Figure 4: T1 Weighted MRI Coronal section showing tumor extension up to oral cavity

After this MRI scan, this patient was seen in the Oncology OPD. This patient was discussed in the multi-disciplinary team Tumor board. Surgery was not possible due to tumor close proximity with brain stem and cavernous sinus. Patient was referred to radiotherapy facility for radiation therapy but she left without treatment and lost follow up. She was kept on oral medications of amitriptyline 50 mg BD, Pregabalin 50 mg BD, Naproxen 500 mg TDS and Serrapeptase 10 mg TDS. These drugs mildly improved her facial pain.

DISCUSSION

The trigeminal nerve is a mixed nerve and supplies the face mainly. Trigeminal Schwannoma affecting trigeminal nerve is rarely reported and exhibits slow, insidious and benign clinical course, however tumor growth and its close proximity with critical structure makes it locally destructive and sometime inoperable when large mass has been developed. It affects middle to old age people, slight predominance for women and symptoms are related to the nerve compression. Studies report that neoplasms involving the trigeminal ganglion usually present with continuous pain, whereas neoplasms involving their roots are asymptomatic which would justify further investigation.[5] This pattern of symptoms was not observed in our patient despite tumor involvement of the root and its branches.

In many of the studies, MRI is the preferred tool of diagnostic imaging and it shows abnormal signal on T1 and T2 weighted images with variable contrast enhancement predominantly cystic component is seen with well-defined tumor margins.

The origin of the tumor is variable and reported cases shows that it arises from trigeminal root, gasserian root or three peripheral branches. Our reported case had few symptoms relatively disproportion of the size of the mass although tumor was inseparable from cavernous sinus and prepontine cistern but no cranial nerve palsy was observed which is an unusual phenomenon.[6] Jefferson gave three categories as following. A: limited to middle cranial fossa, B: limited to posterior fossa, C: both middle cranial fossa and posterior fossa involvement. Day and Fukushima added a fourth D category of extra cranial extension which is in our case. Extra cranial extension of this intracranial neoplasm is a very rare phenomenon seen in our case.

There are various rare clinical presentations of this tumor. These rare clinical presentations include the painless malocclusion and unilateral masticatory weakness, association with primary Sjogren's syndrome, trigeminal motor neuropathy in neurofibromatosis Type II and Unilateral atrophy of the masticatory muscles and mandibular ramus due to pure trigeminal motor neuropathy.[7]

In our case, para-pharyngeal space was extensively involved by the tumor and some studies showed that schwannomas involving the mandibular division of the trigeminal nerve and localizing exclusively in the para-pharyngeal space (PPS) is extremely rare and a surgical approach to such tumors has not been well established.[8] In such cases, the combined trans cervical-trans mandibular approach is preferred for better exposure and facilitates complete resection in some studies but still consensus has to be developed.[9] A locally advanced disease due to extensive para-pharyngeal space involvement render these tumors inoperable as shown in many studies such as by Inderdeep singh series of cases of extensive Para-pharyngeal space involvement stating increased morbidity and mortality due to surgery.[10] Post-operative radiotherapy by stereotactic radiosurgery (SRS) or fractionated stereotactic radiotherapy (FSRT) with minimal toxicity may be a good choice for mild to moderate size tumors with excellent local tumor control. In few studies SRS and FSRT are applied up to 10 ml volumes of tumor and flare phenomenon of symptoms was seen mainly with FSRT.

In our case, complete surgical resection with wide margin should have been done earlier in first surgery and the failure to achieve this goal was the primary cause of recurrence/progression of the disease and development of advanced disease later on. Failure of patient compliance with the treatment is another important cause of recurrence or progression, however effective counseling and patient social and logistic supportive services could overcome this barrier. Also, adjuvant treatment in the form of radiation therapy was

important when microscopic margins are close or positive in case of R1 resection.

CONCLUSION

The primary aim to present this case of trigeminal schwannoma was to inform the readers about the uncommon features of this disease like unusual pattern and presentation disproportionate to the size, origin of the tumor and extensive para pharyngeal space involvement. Such cases require effective Multi-disciplinary team management especially effective joint collaboration of neurosurgeon and head and neck surgeon for a well-planned surgery along with radiation oncologist input as an adjuvant management and last but not the least is patient ownership, which is of paramount importance.

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