



PLEXIFORM SCHWANNOMA OF INFRAORBITAL NERVE - A RARE CASE REPORT

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Abstract: A thirty year old female presented with a painless solitary swelling at right cheek since last one year. Fine needle aspiration cytology was non conclusive. CT-Scan was done which revealed a non-enhancing, multinodular mass in the inferior orbit, pterygopalatine fossa and posterior part of right maxillary sinus with no obvious bony erosion. There was no significant orbital extension. A probable diagnosis of benign schwannoma of infraorbital nerve was made. Sublabial approach was taken and the tumor was excised with preservation of the infraorbital nerve from where it was originating. The mass was multinodular and therefore diagnosis of infraorbital nerve Plexiform schwannoma was confirmed by histopathology. Isolated infraorbital nerve Plexiform or multinodular schwannoma is rare and therefore its literature is reviewed and discussed.

Keywords: Infraorbital Nerve, Plexiform Schwannoma, Schwann Cells.

INTRODUCTION

Schwannomas are benign peripheral nerve sheath tumors that present as slowly progressing, well-defined, unilateral orbital masses.^[1] Schwannomas of the orbit are rare and account for about 1-6% of all orbital tumors.^[2] Orbital schwannomas may arise from supraorbital, infraorbital, oculomotor and trochlear nerves. Orbital schwannomas should be excised as early as possible to avoid complications. We report a rare case of plexiform or multi nodular schwannoma of infraorbital nerve presenting as painless mass in the right cheek, diagnosed on histopathology.

Case History:

A thirty year female, presented with painless solitary swelling near right cheek region since last one year. The swelling was progressive in nature and becoming more obvious in last two months. There was no history of facial numbness or paresthesias. On examination the swelling was painless, firm to hard in consistency and was freely mobile. There was no proptosis and vision was normal. CT-Scan revealed a 5.2 x 3.1 cm mass in the right inferior orbit, pterygopalatine fossa and posterior part of right maxillary sinus. General and systemic examinations were within normal limits. Fine needle aspiration cytology was non conclusive. Written consent was taken. The Sublabial approach was considered as the tumor was not having extensive lateral and orbital extension. A right sublabial incision was given and excision of the tumor with preservation of the infraorbital nerve, from where it was originating, was done under general anaesthesia (figure 1, 2). It was a multi nodular solid mass measuring about 5x3x2 cm in size (figure 3). Postoperative period was uneventful.

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The histopathological examination showed a tumor composed of closely packed spindle shaped cells with occasional nuclear palisading (Antoni A pattern with Verocay bodies) and less cellular loosely textured areas (Antoni B pattern), confirming it to be a multinodular or a plexiform schwannoma (figure 4,5). There was no evidence of malignant transformation. One year follow-up showed no signs of recurrence.



Figure.1:

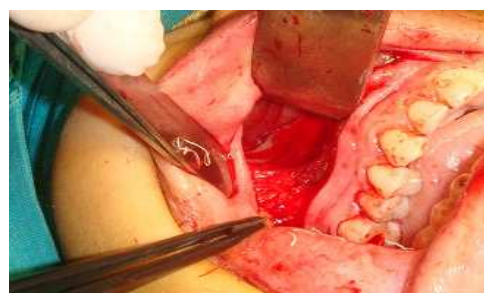


Figure.2:





Figure.3:

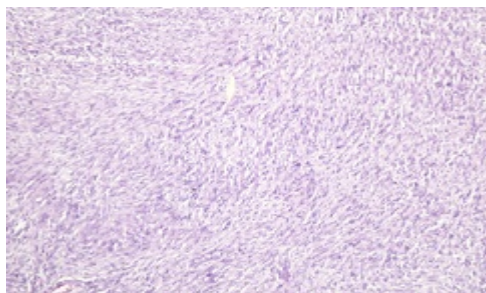


Figure.4:

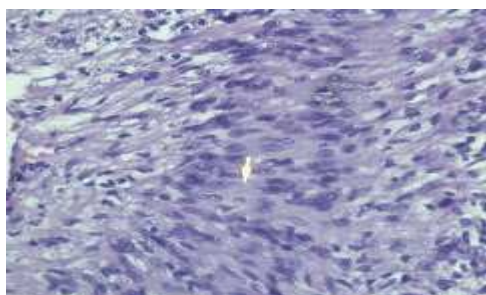


Figure.5:

DISCUSSION

Schwannomas arise from schwann cells present in the nerve sheaths of the originating nerve. Olfactory and optic nerves lack Schwann cells in their sheath and therefore except these two, all other cranial nerves can develop schwannomas. Extra cranial trigeminal nerve schwannomas are less frequent than cranial nerves IX, X, XI and XII. [3,4] Orbital schwannoma usually arise from sensory branches of ophthalmic division of the trigeminal nerve. The supratrochlear and supraorbital nerve are more commonly affected than infraorbital nerve. Garg et al reported a case of infraorbital schwannoma in 2008. [5] Plexiform schwannoma is defined as schwannoma growing in a multinodular manner. [6] Presumably involving a nerve plexus, the vast majority arise in skin or subcutaneous tissue of head and neck or trunk. The tumor is associated with NF2 but not with NF 1 and has been noted in non NF2 patients with multiple schwannomas. [7,8] Cranial and Spinal nerves are usually spared.

Rarely schwannomas may present with numbness in the distribution of trigeminal nerve or with pain or may mimic sinusitis. [9] Larger tumors may produce diplopia, particularly when they arise from orbital parts

of III, IV and VI cranial nerves. Lesions near orbital apex may simulate retrobulbar neuritis. Schwannomas are usually unilateral, although a case of bilateral schwannoma has been reported in literature. [10]

CT-Scan clearly demonstrates the location, extent and bony invasion of tumor. In CT-Scan, schwannomas appear as smooth, solitary mass with their long axis in the direction of nerve. The imaging feature does not clearly distinguish benign from malignant schwannoma of trigeminal nerve. Malignancy is suggested in case of rapid growth or extensive nerve involvement. Confirmation requires tissue diagnosis. Highly cellular tumors have greater chances of recurrences and malignant transformation. Malignant schwannoma of trigeminal nerve has been reported in literature.

A tumor composed of spindle shaped neoplastic schwann cells with alternating areas of compact, elongated cells with occasional nuclear palisading (Antoni A pattern with Verocay bodies) and less cellular, loosely textured often lipidized tumor areas (Antoni B). The Verocay bodies are formed by roughly parallel arrays of tumor cell nuclei separated by dense closely aligned cell processes and basement membrane which are hypereosinophilic. Schwannoma vasculature is typically thick walled and hyalinised.

The treatment of extracranial head and neck schwannoma is exclusively surgical. The appropriate surgical approach is dictated by the extent of the tumor. In our case the tumor was present in inferior orbit extending to posterior part of right maxillary sinus and pterygopalatine fossa. Various surgical approaches have been described in the literature and we attempted Sublabial approach because of limited orbital and lateral extension of the tumor [3]. This approach prevents external scar mark and other possible facial disfigurements which may be seen with other approaches.

CONCLUSION

Infraorbital nerve Plexiform schwannomas are not very common and we present this case as an addition to the other reported cases of schwannomas in various other literature. The tumor was resected using Sublabial approach which provided appropriate exposure with no aesthetic compromise.

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