Case Report

# Isolated Infra-orbital Nerve Schwannoma

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**ABSTRACT**

***Introduction***

*A schwannoma is a benign tumor that develops from Schwann cells. It's known to occur more frequently in women than men, and about one third occur in the head & neck area. However, it is rarely associated with the trigeminal nerve, and especially, schwannomas related to the infraorbital nerve are rare.*

***Case Report***

*A 24-year-old male jute mill worker presented with painless swelling over left side of face for the last 12 years. On examination, localized, nontender, firm 3cm x 3cm swelling was found over left maxilla. Computed Tomography scan showed a well-defined heterogenous oval mass over the left maxilla extending over infraorbital foramen. Aspiration cytology of lesion showed good numbers of spindle cells in groups and dispersedly with fibrous strands. Diagnosis of schwannoma probably arising from terminal branch/trunk of infraorbital nerve was made. Surgical excision of lesion was done under general anesthesia. Histopathological examination of the excised lesion showed grossly firm and gray mass. Microscopically the lesion showed admixture of dense and loose areas referred to as Antoni A and Antoni B areas respectively.*

***Discussion***

*Infraorbital nerve schwannoma is a possible diagnosis in case of a swelling over the maxilla.*

***Keywords***

*Schwannoma; Infraorbital Nerve; Schwann Cells; Midface Swelling; Neurofibromatosis*

Schwannomas are benign tumors arising from neuroectodermal Schwann cell of cranial, intraspinal, peripheral and autonomic nerve sheaths.[1] Schwannomas are component of Neurofibromatosis 2 and even sporadic Schwannomas are commonly associated with inactivating mutations of Neurofibromatosis 2 gene on chromosome 22.[2] Lesions in head and neck region account for 25-45% of extra-cranial schwannomas.[1] However, it rarely involves infra-orbital nerve. They most commonly appear between second and fourth decades.[3] Surgical removal is curative.[4]

This paper describes a case of Schwannoma arising from infra-orbital nerve which presented as a nodular swelling over the left maxilla. The lesion was approached sub-labially and was completely resected. Only 10 cases of Infraorbital nerve schwannoma cases were reported previously.

## Case Report

A 24-year-old male jute mill worker with no known co-morbidities presented with a chief complaint of painless swelling over left side of face for 12 years which was insidious in onset, gradually progressive to reach the present size. There was no aggravating or relieving factors. There was no paresthesia over the swelling. On examination, localized, 3cm x 3 cm swelling over left maxilla extending from 2 cm lateral to ala of nose along the lateral aspect of nasolabial fold, superiorly below the infraorbital margin (Fig.1). Nasolabial fold was partially obliterated. The overlying skin appeared stretched and

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tensed with no punctum or ulceration. It was non tender, firm in consistency and was having mild mobility confirming all the inspection findings. The skin was pinchable over the lesion. Intra oral and anterior rhinoscopy examination yielded no swelling and was normal. There was no proptosis and vision was normal. The cervical lymph nodes were not palpable. Patient had a positive addiction history of chewing betel leaf. No significant past, allergic or drug history. Overall clinical presentation was suggestive of a benign sub cutaneous lesion with differential diagnosis of ossifying fibroma, epidermoid cyst and neurogenic tumor, Trichoepithelioma, sebaceous hyperplasia.

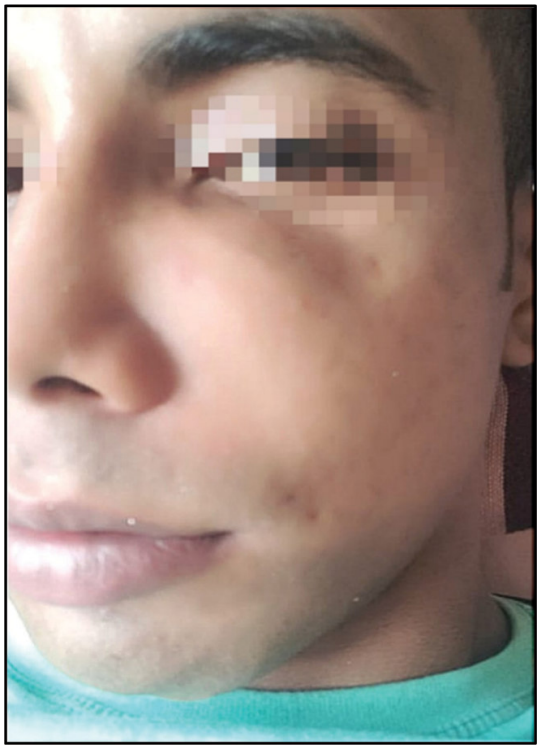

**Fig. 1. Lesion presenting as sub-cutaneous nodular swelling**

CT scan was advised to study the nature, size and extent of the lesion in relation to infraorbital foramen. Report illustrated a well-defined heterogenous oval mass over the left maxilla extending over infraorbital foramen. There was no evidence of bony erosion or extension of mass into oral or nasal cavity.

Aspiration cytology of lesion showed good numbers of spindle cells in groups and dispersedly with fibrous strands.

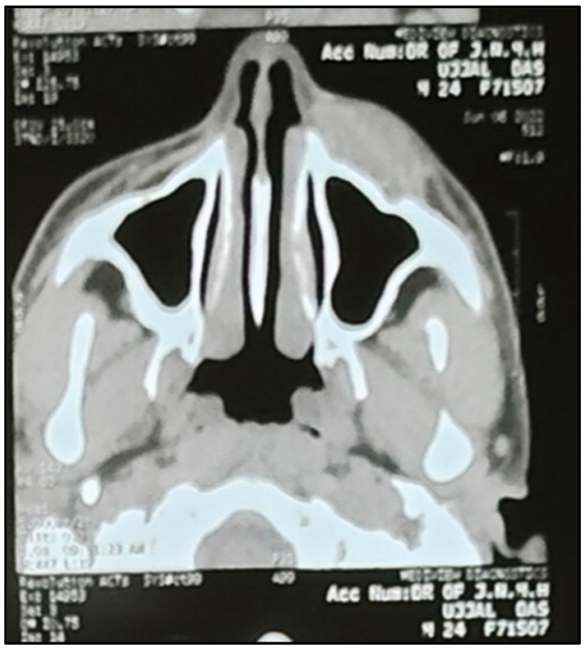

**Fig. 2. CT scan showing a heterogeneous oval mass Over left maxilla.**

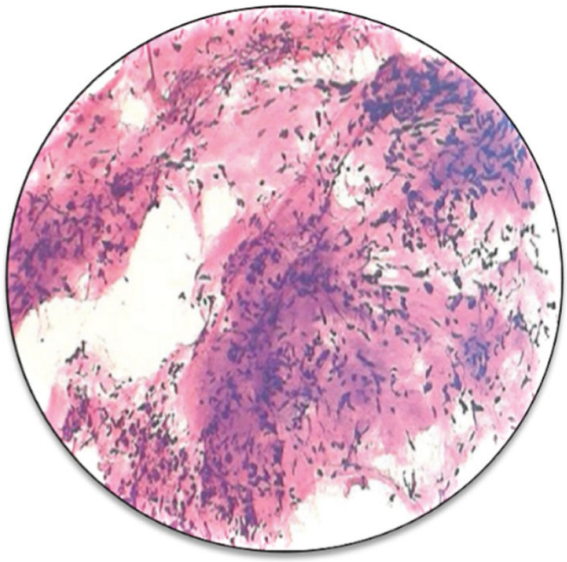

**Fig. 3. F.N.A.C. shows spindle cells in groups dispersedly with fiber strands. (H and E 100X)**

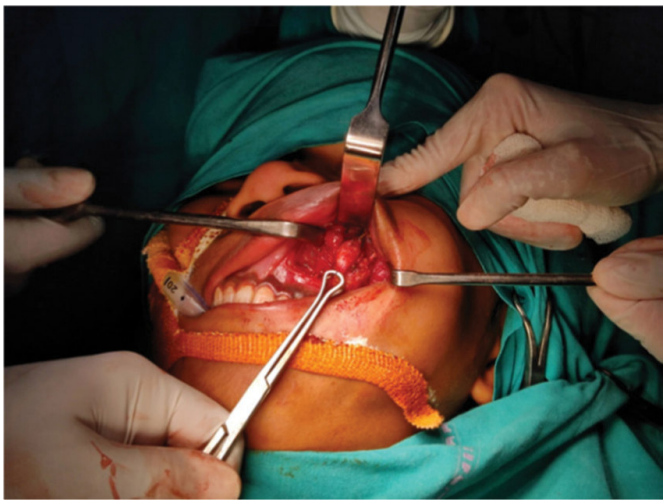

**Fig. 4. Sub-labial incision was made, periosteum elevated, tumor attached with the infraorbital nerve**

Diagnosis of schwannoma probably arising from terminal branch/trunk of infraorbital nerve was made.

Surgical excision of lesion was planned under general anesthesia. After infiltrating adequately with sterile water and adrenaline. Sublabial incision was made. Periosteum elevated, tumor was identified, separated off its margins. The lesion was found to emanate from the nerve trunk of infraorbital nerve which was dissected and preserved meticulously. Hemostasis achieved and wound closed with 3-0 vicryl. Patient withstood procedure well. Post operatively patient experienced paresthesia probably due to the manipulation of nerve while delivering the mass which improved in 2 weeks time.

Histopathological examination of the excised lesion showed grossly firm and gray mass. Microscopically the lesion showed admixture of dense and loose areas referred to as Antoni A and Antoni B areas with no evidence of dysplasia or malignancy respectively suggestive of Schwannoma.

## Discussion

Schwannomas are well differentiated, solitary benign tumors that originate from the Schwann cells of the nerve sheath.[5] It can involve any of the 12 cranial nerves, except the olfactory and optic nerves since they lack Schwann cells in their sheaths. Schwannoma arising from infra-

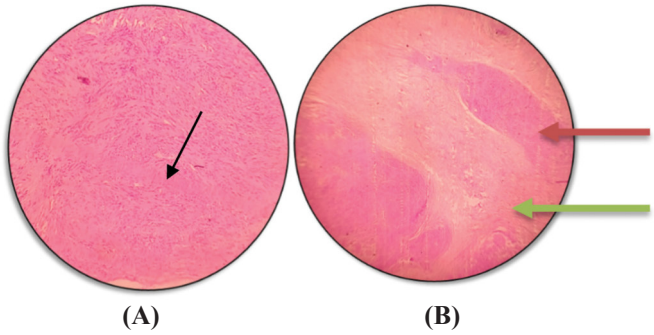


**Fig. 5. H and E stained excised section (A) smear showing Verocay bodies in black arrow (100X), (B) smear showing Antoni A (red arrow) and Antoni B (green arrow) areas (40X)**

orbital nerve (branch of maxillary division of fifth cranial nerve) is extremely rare.[6,7] To best of our knowledge, only ten cases of infra-orbital Schwannoma has been previously described in English literature. Schwannoma generally presents as slow growing painless sub-cutaneous or submucosal swelling.[1]

Depending on the site of nerve trunk or branch from which it arises, it may be located within the lower part of orbit, maxillary sinus or present as sub-cutaneous mass in infra-orbital region. Most of the Schwannomas generally develop from supraorbital or supratrochlear nerves producing downward displacement of globe, and less frequently from Infra-orbital nerves.[1] However, in our case, it was a well-localized painless mid-face sub-cutaneous swelling without any ocular, nasal, oral lesions or involvement of underlying osseous skeleton.

It is rare for schwannomas to present with numbness over the distribution of the nerve it relates which tally with our case with no paresthesia experienced.[8] However during the surgical dissection, lesion was found to emanate from the nerve trunk of infra-orbital nerve, which was dissected and preserved meticulously. Only 1 out of 10 previous reported cases had preoperative paresthesia.[1]

Fine-needle aspiration cytology, ultrasonography, magnetic resonance imaging and computed tomography image are diagnostic tools for Schwannoma. Fine-needle aspiration cytology has a diagnostic accuracy of 17.6% and is characterized by the presence of spindle cells and the finding was similar in our case.[9]

The treatment of schwannomas is exclusively surgical and the approach depends on the extent and location of the tumor. In our case, we did a sublabial approach in view of the cosmesis, making no compromise in the adequacy of exposure and clearance of tumor. Other approaches used include external approach with skin incision, Caldwell-Luc approach for tumor within the maxillary sinus, subciliary or eyelid crease incision, Weber Fergusson incision combined with osteotomy for larger lesions.[1]

Extracranial Schwannomas have good prognosis with exclusively surgical treatment.[10] Malignant transformation and recurrence of Schwannoma is very low, though highly cellular tumors have greater chance warranting early treatment to avoid complications.[11,12] In our patient we were successful in achieving timely and complete excision of the tumor.

## Conclusion

Incidence of Schwannoma arising from Infraorbital nerve is rare making it difficult to diagnose. Hence Schwannoma involving Infraorbital nerve be included in the differential diagnosis of a localized benign subcutaneous mid-face swelling. As a result they can often be resected without sacrificing nerve function.

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