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Ganglion Cell Tumor of Nasal Origin
A Rare Clinical Entity

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Abstract: Ganglion cell tumors occur rather infrequently and usually arise from the central nervous system. The most common site is the temporal lobe (65%). The origin of Ganglion Cell Tumor outside the central nervous system is very rare. This report presents a 25 year old female who presented with a large left nasal cavity ganglioneuroblastoma with blockage of the maxillary sinus. The tumor was removed surgically though a lateral rhinotomy approach. Immunohistochemical analysis was positive for chromogranin and Negative for the S-100 protein.

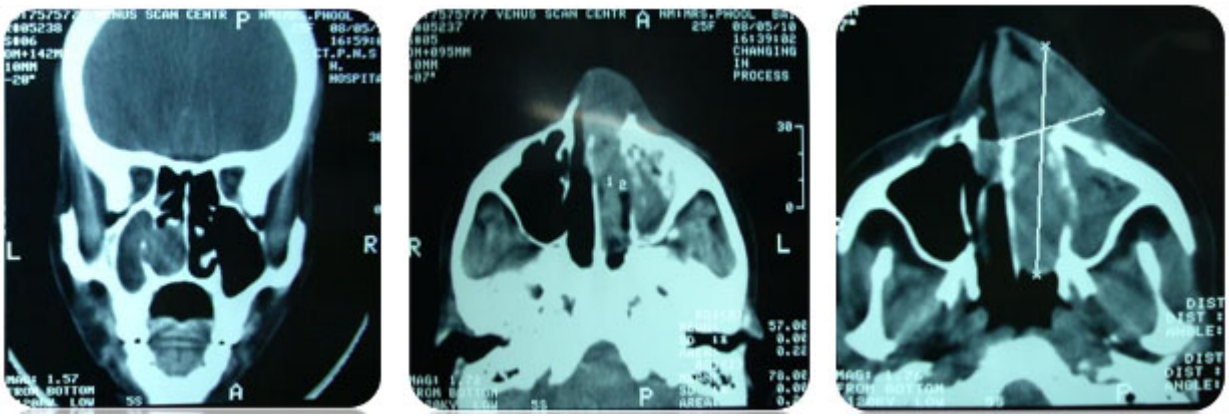
Introduction: In broadest sense, these are neoplasms composed of neoplastic neuronal cells, a gangliocytoma, or a combination of neoplastic neurons and neoplastic glial cells, a ganglioglioma. These tumors are more common in children and young adults with a predilection for the temporal lobes. These tumors are very slow-growing. Here we report a rare case of Sino Nasal Polyp with histopathological characteristics of a Ganglion Cell Tumor.

Case Presentation: A 25 yr old female presented to our department with chief complaints of mass in left nasal cavity along with nasal blockage and difficulty in breathing for 3 months, occasionally associated with blood tinged discharge. No other head and neck complaints were reported. There was no history of seizures.

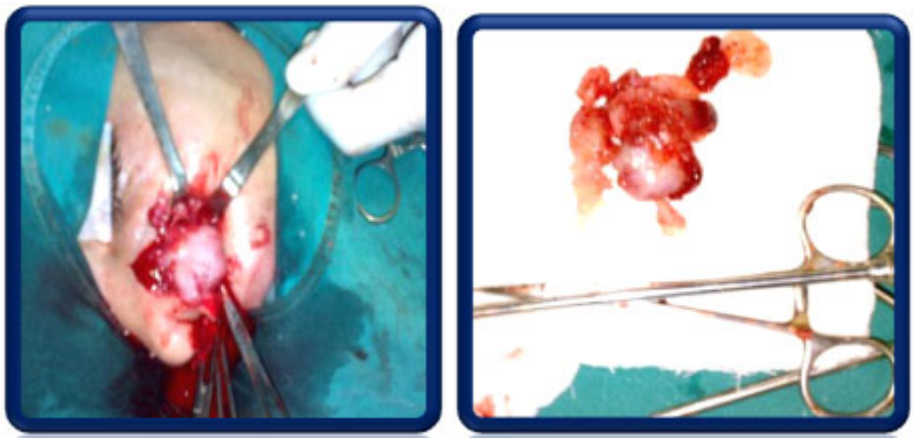
External examination revealed the obliteration of the left nasomaxillary groove while on anterior rhinoscopy a soft and smooth fleshy pink mass was seen, filling the entire left nasal cavity and slightly protruding through the naris. On Probing, the probe was passed all around except laterally and the mass was sensitive to touch and thee was no evidence of bleeding. The mass was not visible on posterior rhinoscopy. The remainder of head and neck examination was unremarkable.

Nasal Endoscopy was attempted both pre- operatively and intra operatively but couldn't be passed due to the size of the mass. Nasal Endoscopy on the opposite side was normal.

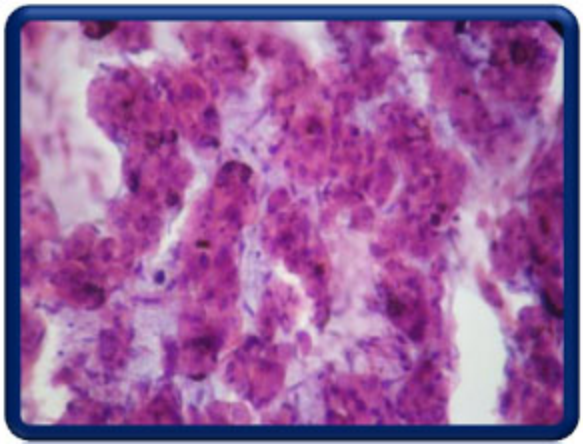
Imaging studies revealed a soft tissue density completely filling the left maxillary sinus and the left nasal cavity, which was suggestive of a left antrochoanal polyp. Biopsy was performed for diagnosis. The pathology showed the presence of polypoidal fragments partially covered by respiratory epithelium and partly by respiratory mucosa. The stroma in focal areas showed irregularly distributed large cells resembling ganglion cells in delicate fibrillar matrix, suspicious of a ganglion cell tumor.



Above are CT Scans showing a soft tissue mass completely filling the nasal cavity and maxillary sinus.



Although the Author prefers the removal of smaller masses via Endoscopic technique, in this case the mass was excised through a lateral rhinotomy approach considering the size of the mass. Intraoperatively, a greyish white colored globular friable mass about 3 by 4 by 4 cubic cm in size was resected. The mass was found limited to the left nasal cavity with sticky mucoid discharge filling the left maxillary sinus. Normal anatomical landmarks of the lateral nasal wall were found to be distorted. There was no breech of the cribriform plate.



Histopathological diagnosis of the postoperative specimen was consistent with ganglion cell tumor. The specimen was also sent for immunohistochemical analysis. The following results were obtained: Positive for chromogranin and Negative for the S-100 protein.

To the right is a picture of a slide stained by markers of ganglion cells.

Discussion: The origin of ganglion cell tumor outside the central nervous system is very rare. These tumors are more common in children and young adults with a predilection for the temporal lobes. Ganglion cell tumors usually arise from the central nervous system, most common site being the Temporal lobe (65%). These tumors are very slow-growing.

Ganglion cell tumors occur rather infrequently and account for 0.4% to 1.3%[1] of all adult brain tumors, and 4.3%[2] to 10.7%[3] of pediatric brain tumors, occurring over a broad range of age with the median age varying amongst studies between 10-25 yrs and are very rare in other sites especially in the nose. Over 90% of ganglion cell tumors are benign. Gangliogliomas are mixed tumors of neoplastic mature neuronal cells and neoplastic glial cells. Usually the histopathological grading of the gangliogliomas depends on the glial component while the neuronal component is almost always benign. In broad aspect, these neoplasms are composed of neoplastic neuronal cells, a gangliocytoma, or a combination of neoplastic neurons and neoplastic glial cells, a ganglioglioma.

The diagnosis of gangliogliomas is occasionally difficult when the neurons are minor components. Wolf et al described that 15% of gangliogliomas contain areas of purely glial cells and emphasized the need for thorough sampling of tumors by surgeons and pathologists.[4] However, the possible occurrence of malignant transformation of the neuronal component in ganglion cell tumors remains controversial.[5]

Some investigators consider that aggressive histological features are not a definite indication of malignancy. Rather, tumor location and resectability are more important determinants of pathological behavior.[6] The pathogenesis of ganglion cell tumors is unknown. Wolf et al. also suggested that the neuronal component of ganglion cell tumors is malformative rather than neoplastic.[4] Ganglion cell tumors

may arise from glioneuronal hamartomas through a neoplastic transformation of the astrocytic component. Another theory is that the ganglion cell tumor may originate from ectopias of the peripheral autonomic nervous tissue in the central nervous system.[7,8] A third theory is that ganglion cell tumors may arise from a single stem cell that differentiates into both glial and neuronal cell lines.[9,10] Further study is needed to understand the pathogenesis of ganglion cell tumors. The differential diagnosis includes:

a) Olfactory neuroblastoma – very rarely it has to be differentiated from an actual ganglion cell, shows the S-100 protein positive focally and is positive for NSE (nonspecific endolase).

b) Ganglioneuroma – found in older age group. The most common location is the retroperitonium and posterior mediastenum. microscopically numerous collection of abnormal fully mature ganglion cells having more then one nucleus and ganglion cells are surrounded by Schwann like cells.

c) Ganglioglioma – glial cells are found, occur in first decade fibrillary material is found.

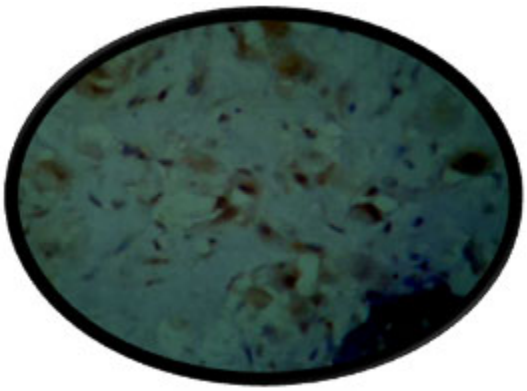
d) Gangliocytoma- composed mainly of neuronal component.

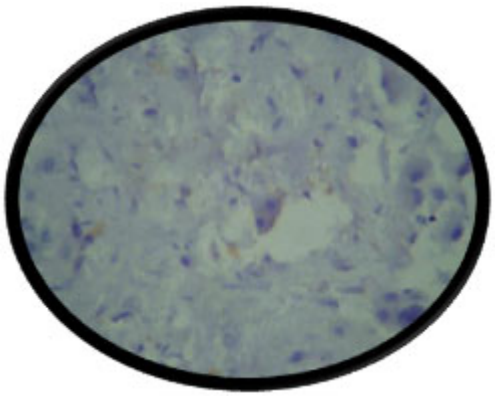
e) Ganglioneuroblastoma –It has primitive neuroblast along with maturing ganglion cells. The number and cell arrangement varies, so tumor assume a wide range of appearance. Generally it follows the distribution of sympathetic gangli, found in paramedian position at any at point between the face and the skull, and the pelvis. The amount and arrangement of the stroma determine the subclassification. The stroma may be:

- Nodular.
- Ganglioneuroblastoma with focal complete differentiation.
- Intermediate.
- Borderline.

Most common location is abdominal (68%) and in 5% the site is unknown. Composed of immature ganglion cells with distinct cell borders, heightened cell cytoplasm eosinophilia & eccentric nucleus. Antibody positivity for chromogranin, NSE - neuronal marker, and to the S-100 protein predicts a good prognosis as the glial component will be increased.. ganglion cells multinucleated, immature with fine fibrillary network is seen between mass of cells.

Our patient's most probable diagnosis is a ganglioneuroblastoma as as indicated by his H & E staining and tumor markers.





Chromogranin Stain -- Positive **S-100 Stain - Negative**

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