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Article

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Primary sinonasal undifferentiated carcinoma with intracranial extension presenting postoperatively as garcin syndrome: a rare case report and review of literature

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Abstract: Sinonasal undifferentiated carcinoma (SNUC) is an aggressive malignancy originating in the mucosa lining the walls of the nasal cavity and paranasal sinuses. It is a rare tumor that usually presents in an advanced stage, is highly invasive, and is traditionally reputed to be refractory to even the most radical therapy with an attendant poor prognosis. This is particularly true when it transgresses the cranial base.

Key words: Sinonasal undifferentiated carcinoma,intracranial extention, Garcin syndrome

Introduction

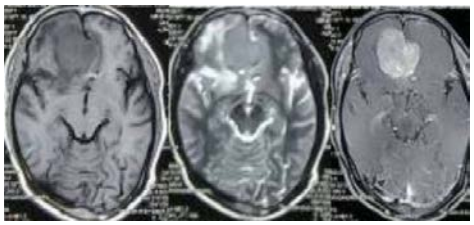
SNUC was first described in 1986 as a rare and highly aggressive neoplasm arising from the nasal cavity and paranasal sinuses[1]. These tumors tend to be large and expansile, with widespread involvement of the nasal cavity and paranasal sinuses. Bone destruction and invasion of adjacent structures such as the orbit, cranial vault, and skull base is frequently seen. The median age of presentation is the sixth decade and men are more commonly affected (male to female ratio of 2.3:1) The vast majority of affected patients present with epistaxis, rhinorrhea, nasal obstruction, and facial pain[2,3]. Very rarely these tumour present as Garcin syndrome. Garcin syndrome consists of unilateral palsies of almost all cranial nerves without either sensory or motor long-tract disturbances and without intracranial hypertension, and it is caused by a malignant osteoclastic lesion at the skull base. The underlying cause of Garcin's syndrome is usually a sarcoma, lymphoma, metastasis, chemodectoma, or carcinoma of the skull base. The literature on Garcin's syndrome is limited, and this makes the present case rare and interesting. The natural history, clinical course and histological features of SNUC are poorly understood[4] Overall survival of patients with SNUC is poor in most reported series. Although no clear consensus exists regarding treatment, a multidisciplinary approach combining surgery (craniofacial resection) with chemotherapy and radiation may be used

to control local disease[2,3]. The extent of disease at diagnosis represents the most sensitive predictor for survival. The purpose of this report is to present a case of SNUC and to review the current concepts of diagnosis, differential diagnosis and management of this rare disease.

Case report

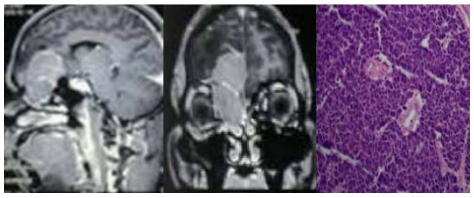
43 years old patient presented to ous with complain of bifrontal headache and nasal obstruction from last 3 month. On examination patient was conscious and oriented to time place and person. His cranial nerve examination was normal except that there was decrease smell sensation on right side. Motor, sensory and cerebellar examination was normal and bladder bowel was not involved. Patient was investigated properly and MRI brain with contrast was done [figure 1, figures 2A and 2B]. Patient was planned for surgery and right sided extended frontal craniotomy was done. Tumour present in basifrontal ethmoidal region was removed, part of tumour attached to medial wall of orbit was left [Figure 3]. Dura was repaired and frontal sinus was exteriorized. Postoperatively patient was conscious and oriented. On day 5 patient develop headache and on subsequent days right sided ptosis was noticed. Initially it was partial ptosis but in 2-3 days it become complete ptosis and it also develop on contralateral side. On examination we found that there is also involvement of 4th and 6th cranial nerve. Within next 5-6 days there is also involvement of 5th, 7th, 9th, 10th and 12th cranial nerve on ipsilateral side. Patient also develop pain in the back region. Inspite of involvement of multiple cranial nerve patient had no motor and sensory impairment and his bladder and bowel was intact.

We report this case as SNUC is very very rare and involvement of multiple cranial nerve in step by step fashion without impairment of motor and sensory systems (Garcin syndrome) is extremely rare. So this case become an interesting case and only few such cases are reported in the literature.



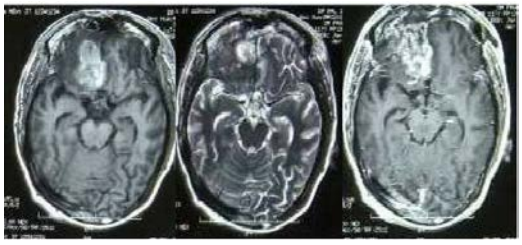
A B C

Figure 1A - MRI T1W axial image shows hypointence mass present in right frontal region
Figure 1B - MRI T2W axial image shows iso to hypointence mass present in right frontal region
Figure 1C - Contrast MRI axial image shows contrast enhancing mass present in right frontal region



A B C

Figure 2A - Contrast MRI saggital image shows contrast enhancing mass in right basifrontal region with extension into nasal cavity
Figure 2B - Contrast MRI coronal image image shows contrast enhancing mass in right basifrontal region with extension into paranasal sinuses
Figure 2C - H&E stained sections show a tumour disposed in nests with interspersed lymphocytes and plasma cells. The tumourcells are round to oval with high nuclei-cytoplasmic ratio, hyperchromatic nuclei and scant cytoplasm. Numerous mitotic figuresare evident



A B C

Figure 3A - Post ope MRI T1W axial image shows hyperintence changes in right frontal region

Figure 3B - Post ope MRI T2W axial image shows iso to hyperintence changes in right frontal region

Figure 3C - Post ope contrast MRI axial image shows small residual tumour in right frontal region

Discussion

SNUC is an aggressive, highly malignant tumor of the mucosal lining of the nasal and paranasal sinuses. It was first described as a distinct and separate entity by Frierson et al in 1986 [5].The prognosis of the patient is very poor with high frequency of local recurrence and distant metastasis. These tumour generally present with epistaxis, nasal obstruction and headache. Very rarely these tumour present as Garcin syndrome. Garcin syndrome is an ipsilateral step-by-step deterioration of all 12 cranial nerves, first described in 1927. This rare progressive condition is generally associated with skull based malignant osteoclastic lesions but has also been described with sarcoma, lymphoma, metastasis, chemodectoma, or carcinoma of the skull base.

Cranial nerves involvement in these tumour occurs in many ways. First, the tumor may extend superiorly through foramen lacerum, which is an unimpeded pathway near the fossa of Rosenmuller into the cranium, thus involving the cranial nerves in the middle cranial fossa and cavernous sinus. Cancer may break through the pharyngo-basilar fascia and spread along vascular sheaths, that is, facial planes surrounding the jugular vein and carotid artery. Lateral and posterior extension of the primary growth itself may involve the lower cranial nerves exiting from jugular and hypoglossal foramina.

Confirming the diagnosis of SNUC at the light microscopic level can be challenging, since the microscopic differential diagnosis include olfactory neuroblastoma, rhabdomyosarcoma, undifferentiated nasopharyngeal carcinoma (lymphoepithelioma), malignant lymphoma, malignant melanoma and neuroendocrine carcinoma. SNUC can be differentiated from these other neoplasm by correlating clinical, light microscopic, immunohistochemical and ultrastructural charecteristics. [6] The tumour is positive for pan-cytokeratin, epithelial membrane antigen, CD99, cytokeratin7 with scattered p-40 expression on immunohistochemistry.

Olfactory neuroblastoma typically present as a polypoidal mass protruding from the roof of nasal cavity. Immunohistochemistry demonstrate staining for neuroendocrine markers such as neuron specific enolase (NSE), synaptophysin and chromogranin. S-100 is usually positive in a sustentacular pattern [7].

Differential diagnosis of SNUC from olfactory neuroblastoma and neuroendocrine carcinoma may be difficult because a small proportion of SNUC may express neuroendocrine markers including NSE and

chromogranin. However SNUC lacks S-100 and does not show glandular differentiation or rosettes. SNUC has more extensive cytological atypia and necrosis. Ultrastructurally neuroendocrine markers tend to be focal and no neurosecretory granules are evident in SNUC. [8] Melanoma and lymphoma are easily differentiated on the basis of immunohistochemistry. Melanoma stain positively for S-100, HMB 45 and melan A, while lymphomas are invariably positive for CD19 and CD20 as well as other B cell lineage markers [9].

Due to rarity of the disease the proper diagnostic and management criteria could not be defined. On reviewing the literature we found that we can differentiate this tumour from others by histological and immunohistochemical properties. SNUC could be treated by craniofacial resection followed by adjuvant radiotherapy and or combination of chemotherapy. Inspite of these multimodality treatment these tumour recur in one year and most of these patient died. [10]. So early diagnosis and prompt treatment can improve the survival of these patient.

Conclusion

Sinonasal undifferentiated carcinomas (SNUC) are highly aggressive lesions arising in the superior nasal cavity and paranasal sinuses. The differential diagnosis may be wide since a range of similar lesions may present at this site. Recently the criteria for diagnosis of SNUC has been clarified. On the basis of histological and immunohistochemical charecteristics we can differentiate it from other lesions.

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