INTRODUCTION
Carcinoma of the paranasal sinuses accounts for about 0.3% of all cancers. Adenosquamous carcinoma appears in the surface epithelium of respiratory tract. The most frequent known site is supra glottic larynx (almost 50%), followed by oral cavity and oropharynx (about 40%); 8% occur in hypopharynx while only 2% occur in nose and paranasal sinuses tumours.1 Adenosquamous carcinoma of head and neck is an aggressive malignancy. Only 7 cases of adenosquamous carcinoma of paranasal sinuses have reported in the literature. These aggressive tumours erode the vital areas early due to their proximity to skull base and become inoperable.2 As these tumours do not occur frequently so their various management modalities other than surgery like chemo-radiation and immunotherapy are yet to reveal and their effect on tumour need trials. Adenosquamous carcinoma rarely present as either T1 or T2 tumours.3,4

Primary ciliary dyskinesia (PCD) is very infrequent disease; its incidences estimated 1 in 32,000 only.5 This is autosomal recessive genetic disorder that causes defect in cilia lining upper and lower respiratory tract, paranasal sinuses, eustachian tube, middle ear and fallopian tube. Dysfunction of cilia results in poor mucociliary clearance. PCD when accompanied with situs inversus, chronic sinusitis and bronchiectasis is known as Kartagener syndrome.6,7

Kartagener syndrome and adenosquamous carcinoma have never reported in the same patient before in literature in addition to the AB negative blood group.

CASE REPORT
A 34 years old non-smoker male patient reported with growth of right maxillary region which on histopathology confirmed adenosquamous carcinoma of nose and paranasal sinus. Patient also had total situs inversus including dextrocardia, bronchiectasis and sinusitis. His blood group was AB negative. This association of Kartagener syndrome with adenosquamous carcinoma of paranasal sinuses has never been reported. Kartagener syndrome, AB negative blood group and adenosquamous carcinoma of paranasal sinuses all are extremely rare clinical conditions found in populations and the combination of all three in the same patient have never been reported to the best of authors' knowledge.

Key Words: Adenosquamous carcinoma. Paranasal sinuses. Situs inversus. Kartagener syndrome. AB negative blood group.
Specimen was in single piece of tissue of about 1.5 x 1 x 1 cm revealed skin exhibiting an infiltrating tumour cells. The tumour showed two patterns of Keratinizing squamous cells and the other glandular cells with clear cells containing mucin. Adenosquamous carcinoma was reported in histopathology.

He made an uneventful recovery. On his discharge, he was advised to take chemo-radiation and remain in regular follow-up. The patient did not get chemo-radiation neither he visit us till February 2011 when he became worried due to broadening of nose and eyes displacing apart. There was history of occasional episodes of epistaxis and frontal headache for last 2 - 3 months. On examination, there was 3 x 3 cm hard painless swelling at the root of nose that was extending to both medial canthi. This swelling was more on right side causing telecanthus and proptosis, however, vision was intact and no diplopia or extra ocular muscle palsy was found. Corneal and pupillary reflexes were intact. Sclera was exposed with ectropion of lower lid due to previous wound healing by forehead flap. Right nasal ala was displaced more laterally and inferiorly (Figure 1). On anterior rhinoscopy, growth was present on right nostril that bled on touch and sensitive on probing. Posterior rhinoscopy reflected reddish mass. Olfaction was found slightly decreased.

CT scan revealed a large soft density enhancing mass measuring 4.0 x 6.0 x 4.2 cm occupying the nasal cavity, right ethmoids and periorbital tissue. Mass eroded the cribriform plate and extending to anterior cranial fossa and sphenoid sinus (Figures 2 and 3). Enlarged lymph nodes of 9 mm identified at level-I bilaterally. MRI identified mass of abnormal signal intensity completely involving the ethmoid and nasal cavities frontal sinuses bilaterally with bulging of lamina papyracea bilaterally marked on right side. The lesion showed non-homogenous signals on T2 weighted images and isointense on T1 weighted images (Figure 4).

Case was discussed in ENT monthly meeting with the board of Head and Neck Surgeons from Karachi city held at Dow University of Health Sciences. In that meeting consensus was made to give maximum survival with less possible morbidities. It was decided that as the patient is young and may tolerate extensive craniofacial surgery, so he must be given chance of survival by removing this aggressively growing tumour as if no surgical intervention was done at this time, patient will die of this tumour. When the nature of surgery and its likely complications were explained to the patient, he refused for surgery and left the hospital.

After 2 weeks, he presented in emergency with marked swelling of nose. It measured 4 x 5 cm at the root of nose. There was profuse bleeding from nose which was controlled by applying anterior nasal packing. Proptosis became more marked accompanied with chemosis of eyes. He was operated in emergency. Incision was taken below both eye brows joining the right sided lateral rhinotomy incision. Elevation of the flap exposed tumour that was eroding the septum both nostrils extending up to ethmoidal, frontal and sphenoidal sinuses. Tumour was highly vascular and fleshy in appearance eroding the lamina papyracea. With the aid of Freer's elevator, tumour was separated from the wall of sinuses and facial bones. As it was resected from the anterior skull base, the dura was found breached by the tumour and was repaired with temporalis fascia. It was planned to give chemo-radiation therapy for residual tumour that had became inoperable. Surgery lasted for more than 5 hours. Bleeding was massive even after the removal of tumour, diffuse bleeding was continuous from walls of sinuses.

He was transfused during surgery to compensate blood loss exceeding 2 liters. Despite all measures, he expired on ventilator due to multi-organ failure on the day of surgery.

**DISCUSSION**

Kartagener syndrome, AB negative blood group and adenosquamous carcinoma of paranasal sinuses all are extremely rare clinical conditions found in general population and the combination of all three in the same patient have never been reported to the best of our knowledge. There were many more unusual sequelae in this man. He suffered with adenosquamous cancer of maxillary sinus at a relatively young age of 34 years. The reported incidence is in 5th - 6th decade of life.
Adenosquamous cancer (ASC) of the head and neck is an infrequent aggressive neoplasm. Only 7 cases of ASC of nose and paranasal sinuses have been reported in the literature. ASC originates in the surface epithelium of the upper respiratory tract and is found more commonly in males. ASC has two distinct histological components; the most extensive one is a keratinizing squamous cell carcinoma arising from the surface epithelium and the second component is adenosquamous carcinoma usually found in the deepest areas of tumour. Maxillary sinus cancers were mostly either squamous or adenosquamous that developed from lining mucosal epithelium or metaplasia.

Alos et al. studied 12 cases of adenosquamous carcinoma of head and neck and all patients were male. He concluded that peak age of the patients at time of presentation was found 60 years and the commonest site appeared larynx. Fifty percent of patients with ASC died after a mean period of 23 months. Management of adenosquamous carcinoma is controversial as it is an unusual tumour which is often misdiagnosed. Its prognosis is very poor due to high metastatic rate. This patient presented at the age of 34 years only and tumour appeared in the maxillary sinus first. Management of adenosquamous cancer of paranasal sinuses is challenging as this tumour become extensive and destructive when involving skull base. Only one case has been reported about an adenosquamous cancer of nose that was limited and removed via endoscope successfully. The patient was first presented with a limited lesion of maxilla. It can be assumed that if he had acted according to the advice to get postoperative chemo-radiation therapy after the first surgery, he might had different fate. The problem was patient approaching the study centre had origin from remote and rural areas and had inability to afford the expense of living in the city for treatment. Craniofacial resection of the lesion, when it is eroding the roof of ethmoids and cribriform plate, leads to morbidities like CSF rhinorrhea, orbital injury, and intracranial injury. Reconstruction of nose after wide excision is a big question as well. Adenosquamous carcinoma of nose and paranasal sinuses requires extensive surgical excision followed by chemo-radiation therapy.

REFERENCES