Nasal Pleomorphic Adenoma

**KEYWORDS**
pleomorphic adenoma, lateral nasal wall, FNAC,

**ABSTRACT**
Pleomorphic adenomas are the most common benign tumor of the major salivary glands. In addition to the major salivary glands, they may also occur in the salivary glands of the hard and soft palate. Rare cases have been reported in the nasopharynx, oropharynx, hypopharynx, and larynx. Intranasal pleomorphic adenomas are quite rare and may be misdiagnosed because they have greater myoepithelial cellularity and few myxoid stromata compared to those elsewhere. We present a rare case of a pleomorphic adenoma of the nasal ala and discuss the pathologic findings and clinical management. It rarely originates in the nose, and is especially rare in the lateral nasal wall.

**CASE REPORT**
A 52-year-old female presented to an outside institution with a 5-year history of right-sided nose swelling and 2-month history of repeated pain. On seeing by naked eyes revealed a smooth, reddish, not easily hemorrhagic tumor causing pressing of the right nasal cavity. We admit her in 25/4/2016 and operate on 03/5/2016. Dr. Shamendra (+919414909984), Dr. Vijay Kumar Meena and Dr. Rajkumar Jain were team members.

Her head and neck examination showed no other lesions or lymphadenopathy. On FNAC report was pleomorphic adenoma.

The histology was typical of pleomorphic adenoma and comprised an admixture of epithelial, myoepithelial and stromal components with acini or ducts and a myxochondroid stroma (Figure 4). There was no invasion of the adjacent structure microscopically. The patient's postoperative course was uneventful. She healed well without evidence of residual or recurrent disease on follow-up.

**Introduction**
Pleomorphic adenoma is a benign mixed tumor mainly arising in the major salivary glands. Rarely, they can be found in the minor salivary glands of the upper aerodigestive tract, including the nasal cavity, pharynx, larynx, and trachea. Intranasal pleomorphic adenomas are rare and nasal septum is the most common site of origin. Here, we present and discuss an unusual case of pleomorphic adenoma arising from the lateral nasal wall.

Mixed tumour of the salivary gland or the pleomorphic adenoma is a benign tumour, arising mainly in the major salivary glands (65%), especially in the parotid and, less frequently, in accessory salivary glands (35%). Rare cases have been reported in the lip, 2 the hard and soft palate, 3 the lacrimal gland, 4 and the external auditory canal. 5 It is extremely rare to find these in the respiratory tract. 6-8 The incidence is even lower in the upper respiratory tract, such as the nasal cavity, maxillary sinus and nasopharynx. 8, 9 The largest reported case series of intranasal pleomorphic adenomas are those of Spiros et al. 10 with 40 cases, Compagno and Wong 11 with 40 cases and, more recently, Suzuki et al. 12 with 41 cases.

Intranasal pleomorphic adenomas generally arise in the nasal septal mucosa (reported incidence varies between 82.5% 12 and 90% 13), even though the seromucosal glands are mainly located within the lateral nasal wall, in particular in the turbinates. 13

Various theories have been proposed to explain this observation. According to Stevenson 14, remnants of the vomero-nasal organ, an epithelium-lined duct in the cartilaginous nasal septum degenerated in early foetus, could be the reason for the appearance of these tumours in this particular region. According to Ersner and Saltzman, in 1944, the precursors of the septal pleomorphic adenoma are ectopic embryonic epithelialised cells on the nasal septum mucosa, found during the migration of the nasal buds. 15 According to Evans and Cruikshank, it originates directly from the matured salivary glandular tissue. 16 Dawe, in 1979, proposed a viral aetiology from polyoma virus. 17

(Figure 1).
Postoperative photo was

Figure 2.

DISCUSSION
Nasal pleomorphic adenoma is seen predominantly in females usually between the third and fifth decades of life. There is no reported correlation with occupational exposure or inhaled toxic chemical compounds. It is generally known to be a slow-growing tumour and, therefore, clinical symptoms appear after a long silent period. Patients commonly present with gradual worsening of monolateral nasal obstruction and occasional epistaxis. Less commonly, when the tumoural mass reaches a relatively large size, to that of the nasal cavity, external swelling of the nasal pyramid as well as pain may be present.

Clinically, it appears as a polyloid, unilateral, sessile, translucent pinkish-grey mass, with smooth surface and soft consistency. Tumours can range in size from < 0.7 cm to > 7 cm.

The clinical features, such as absence of superficial ulceration, no bleeding either on touch or spontaneously and lack of invasion of surrounding structures suggest a benign nature of the mass.

Histologically, all pleomorphic adenomas have a collagenous thin capsule, with a clear-cut distinction of the tumour tissue from the surrounding normal connective tissue. The tumours consist of three main structures: tubuloductal structure, solid area, and myxoid area. The tubuloductal structure presents ducts with double cell layers: cuboidal-shaped epithelial cells at the inner layer, and spindle-shaped myoepithelial cells at the outer. Predominantly the solid areas consist of the spindle-shaped cells with high cellularity; the myxoid areas are characterised by their low cellularity.

The intra-nasal pleomorphic adenoma shows a predominance of epithelial rather than stromal elements, as compared with major salivary gland tumours. The epithelial cells are small, oval-shaped and often arranged in cords; sometimes, they are organized in small acinous structures.

Immunohistochemical stains prove positive for various cytokeratins, S100 protein, glyal fibrillary acid protein (GFAP), Vimentine, a smooth muscle actine (SMA). This describes the “mixed” nature of the tumour, namely, the stromal and the epithelial line.

Differential diagnosis of intra-nasal pleomorphic adenoma includes both malignant and benign tumours such as squamous cell carcinoma, adenocarcinoma, adenoid cystic carcinoma, mucoepidermoid carcinoma, melanoma, olfactory esthesioneuroblastoma, polyps, papillomas (including inverted papilloma), angiofibromas and osteomas.

Differential diagnosis can also be difficult in the presence of a “neuroestesioepithelioma” (even though the site of origin of such a lesion, i.e., the ethmoid plate, should suggest its nature, since the lateral nasal wall is extremely rare), as, in the early stage, it presents a small cell proliferation organized like a “rosette”, positive for S100 protein. Diagnosis is possible on account of the lack of an extracellular neurofibrillar structure, neurorubules, neurosecretive granules and due to the presence of mucinous material and the rarity of malpighian lobules disseminated on the pleomorphic adenoma. Regardless of where the lesion originates, the main treatment modality should be surgical. While complete excision of the tumour with histologically clear margins is mandatory, even if the tumour appears to be clinically benign and resected completely.

Recurrences are not frequent. Compagno and Wong reported 3 cases of local recurrences in 40 patients (7.5%) probably, as they thought, due to the amount of myxoidstroma of the tumour, which could be split into the surgical field.

The potential risk of malignant transformation of the pleomorphic adenoma is about 6% and is predominantly seen in the female patients. The risk is increased by delay in diagnosis.

A histopathologically confirmed case with adenoid cystic and squamous carcinomatous differentiation has been reported.

There has also been a report of metastasis to the submandibular lymph-node, in a recurrent sepal pleomorphic adenoma, 17 years after the first diagnosis. Even in this case, the microscopic features of both the primary and metastatic lesion were benign. In this regard, a iatrogenic theory has been proposed. This theory suggests that the metastasis occurs as a result of incomplete excision or inadvertent disruption of the tumour with consequent spread through haematogenous or lymphatic routes. Pulmonary, hepatic and bone metastasis have also been reported.

Long term follow-up is therefore necessary for early diagnosis of loco-regional recurrences by endoscopic examination followed by imaging (CT or MR) in case of clinical evidence of disease.

Conclusions
In the presence of a slow-growing unilateral mass of the nasal cavity, it is important to consider, among the various diagnoses, the presence of pleomorphic adenoma, even if it is not frequently encountered. Early diagnosis offers the possibility of a more complete excision with adequate care being taken not to disrupt the tumour in order to prevent local and distant spread of neoplastic cells. Long-term follow-up, to exclude malignancy is mandatory, even if the tumour appears to be clinically benign and resected completely.

References