

The Unusual Tales of Arch & Nape

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DOI: 10.36347/sjams.2020.v08i01.006

| Received: 24.12.2019 | Accepted: 07.01.2020 | Published: 13.01.2020

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Abstract

Original Research Article

Introduction: Cancer is among the ten leading causes of death in India, and head and neck neoplasia in India accounts for 23% of cancers in males and 6% in females. They encompass a diverse group of uncommon tumors that frequently are aggressive in their biologic behavior. **Aims & Objectives:** Considering the magnitude of the problem in India and the associated high morbidity and mortality we undertook this study to focus on the rare and unusual head and neck malignancies. **Material and methods:** A retrospective observational study of unusual malignancies diagnosed in head and neck swellings. **Result:** A spectrum of rare malignancies was encountered including chondrosarcoma of the maxillary sinus and ocular surface squamous cell neoplasia. Anaplastic large cell lymphoma presenting as an infra clavicular swelling, polymorphous low grade adenocarcinoma of the hard palate and clear cell carcinoma of minor salivary gland were some of the other rare tumors reported. Rare thyroid malignancies included poorly differentiated carcinoma and extramedullary plasmacytoma of thyroid. Few uncommon benign tumors were also reported including schwannoma of left side neck and plexiform neurofibroma of the lip. **Conclusion:** The need of the hour is to prioritize control and early detection of head and neck cancer in India with special emphasis on the uncommon entities. An awareness of rare entities is important to implement cancer control activities and fast track the diagnosis and management so as to deliver utmost benefit to patients.

Keywords: Head and neck, rare malignancies, spectrum.

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INTRODUCTION

Head and neck lesions are encountered in patients across all age groups commonly.¹The lesions have varying histology ranging from a spectrum of benign, inflammatory at one end to malignant on the other end [2]. Cancers of head and neck constitute a significant number of 23% diagnosed malignancies. India sadly has the highest incidence of these neoplasms in women. The anatomy of the head and neck is quite complex with various divisions into sites and subsites, encompassing a plethora of uncommon tumors that frequently have aggressive biological behavior [1].

The major risk factors for head and neck malignancies are preventable namely smoking, tobacco chewing and alcohol consumption. In various studies malignant cases were more common in males than females [3]. The five year survival rate of head and neck neoplasia varies from 20-90% depending on the site of origin, clinical extent and the biological behavior of the malignancy. The spectrum of these malignancies has a varied geographical distribution in the country and

site specific data can help in establishing the etiological factors responsible for this variation [4].

The present study of unusual malignancies diagnosed in head and neck swelling was undertaken considering the magnitude of the problem in India and the associated high morbidity and mortality with it. Numerous studies are present with emphasis on histopathological and cytological spectrum including inflammatory, benign and malignant lesions of this region with a scarcity of data on the rare entities diagnosed in this region. This study therefore focusses on the rare and unusual head and neck malignancies and diagnostic problems related with the same.

MATERIALS & METHODS

The present study was undertaken in the Department of Pathology, Hamdard Institute of Medical Sciences and Research and Hakeem Abdul Hameed Centenary Hospital, New Delhi between January 2011 to December 2017. Those patients who presented with head and neck lesion in Medicine, Surgical,

Dermatology and Dental OPD or admitted in hospital and underwent fine needle aspiration cytology (FNAC), a biopsy or resection with an uncommon diagnosis were considered as the study group. This retrospective observational study included 15 cases of head and neck swellings having a rare and unusual diagnosis over a 7 year time period.

The FNAC samples and the harvested material was transferred on to a glass slide taking care that minimum damage to cells is incurred. The smears were stained in the cytology laboratory of the department. The post-operative surgical specimens were received in the histopathology laboratory fixed in 10% neutral formalin and subjected to gross examination, processing, paraffin embedding, section cutting,

staining by Haematoxylin & Eosin and mounting by DPX. H & E Stain uses hematoxylin solutions for nuclear staining and eosin solutions for cytoplasmic staining. Specific Immunohistochemistry wherever required was performed.

The clinical presentation and the histological features were analyzed and the results were compared with those in the literature.

RESULTS

The following unusual malignancies encountered have been tabulated along with their sites and subsites in Table I and case details in Table II

Table-I: Distribution of cases according to site

Location	Unusual diagnosis
Eye	Conjunctival squamous cell carcinoma Sudoriferous cyst
Thyroid	Poorly differentiated carcinoma thyroid Extramedullary plasmacytoma
Larynx	Spindle cell carcinoma with tuberculosis
Salivary gland	Clear cell carcinoma Oncocytoma
Hard palate	Polymorphous low grade adenocarcinoma
Maxillary sinus	Chondrosarcoma
Tongue	Lingual schwannoma
Lip	Plexiform neurofibroma
Face	Adenoid basal cell carcinoma (nose) Malignant melanoma & basal cell carcinoma
Floor of mouth	Adenosquamous carcinoma
Infra auricular region	Extranodal anaplastic large cell lymphoma
Nose	Sinonasal Teratocarcinoma

Table-II: Distribution of cases according to their case details

Case	Location	Clinical presentation	Ancillary investigations	Gross	Histopathological findings	Diagnosis	IHC
1.	Eye	42year/ Male Upper eyelid cyst for 1 year.	Not done	Cyst, 1cm diameter	Cyst lined by double row of columnar cells with eosinophilic cytoplasm. In focal areas the cyst wall is thrown into papillary infoldings.	Sudoriferous cyst	-
2.	Eye	78years / Male Conjunctival mass for four months	Not done	0.8 x 0.8 cm soft tissue mass	Acanthotic squamous epithelium showing parakeratosis, dyskeratosis and marked atypia.	Conjunctival Squamous cell carcinoma	-
3.	Thyroid	42 year/ Female, Presented with goitre: 10 months back. The swelling was increasing in size.	FNAC done twice, suggested Colloid Goitre Computerized tomography showed a large complex thyroid with neck nodes	A specimen of total thyroidectomy with central neck node dissection. Grayish white, firm lesion with focal colloid collection. Few colloid cysts also seen.	Solid nests of follicular cells nodular growth pattern along with peritheliomatous pattern, extending into the surrounding tissue. Sheets and nests of large atypical cells with marked pleomorphism, Large areas of necrosis was seen in the intervening stroma.	Poorly differentiated carcinoma of the thyroid	Thyroglobulin positive

4.	Thyroid	53 year/ male Presented with a left-sided thyroid swelling of six months duration.	Thyroid profile showed slightly increased TSH (5.92 mIU/mL) Decreased T4 (3.65/dL. Well-defined mass on CECT, 6x6x10 cm in the left thyroid lobe. FNAC suggested Plasmacytoma thyroid in a background of thyroiditis. Serum kappa-free light chain level was increased (24.07 mg/L), while the level of lambda free light chain, kappa/lambda ratio were in normal limits	A globular soft tissue measuring 8.5x6x5 cm. The external surface was nodular with an adherent capsule and showed prominent blood vessels. Cut surface was gray, firm and fleshy.	Sheets of plasma cells infiltrating the thyroid parenchyma with few entrapped atrophic thyroid follicles. These cells were seen having eccentric, round nuclei with abundant eosinophilic cytoplasm. Sporadic binucleated and multinucleated plasma cells were also observed. Many lymphoid follicles with germinal centers were also present.	Extramedullary plasmacytoma of the thyroid in a background of Hashimoto's thyroiditis	CD138-positive tumor cells while CD20 was negative, Kappa light chain showed strong cytoplasmic positivity in the tumor cells while that of the lambda light chain was negative
5.	Larynx	40year male with hoarseness of voice for two years. Laryngeal growth excised. Recurrence after 5 months.	Laryngeal growth showed well differentiated squamous cell carcinoma. Recurrence of laryngeal growth after 5 months. showed spindle cell carcinoma of the larynx on biopsy	Total laryngectomy with selective neck dissection, right side	Sheets of spindle cells with focal myxoid and neural differentiation, involving the glottic and subglottic region. Cervical lymph nodes showed granulomatous lymphadenitis with no metastatic deposits. Acid fast bacilli were demonstrated on Ziehl Neelsen staining Nested PCR assay for Mycobacterium tuberculosis on lymph node tissue was positive.	Spindle cell carcinoma larynx (pT3N0) with tuberculosis	: tumor cells - Positive for AE1/AE3, Vimentin, S-100. - Negative for ALK-1
6.	Salivary gland	36years female with a slow growing painless mass in the tonsillar region for four years. Previous biopsy: squamous cell carcinoma	CECT: large ulceroproliferative lesion involving the left tonsil, floor of the mouth and left side of tongue, ipsilateral cervical lymph nodes of Level II and III were present.	Left tonsillar commando with RND Firm grey white solid tumor mass 4.5x3x 1.8 cm involving the base of tongue, left tonsillar sulcus and the tonsil.	Clear cell tumor with cells arranged in sheets, nests and trabeculae formation. Cells were polygonal with monotonous morphology, distinct cell borders, clear cytoplasm and a centrally placed small, round nuclei, infiltrating muscle. Intervening fibrous stroma was seen separating nests of cells. Minimal mitosis and focal areas of hyalinisation were also seen. Lymph node status: 4/21 positive	Clear cell carcinoma (NOS) of minor salivary gland origin	-
7.	Salivary gland	17years male with gradually increasing painless swelling left submandibular region for 1 year. O/E: 5X4 cm firm non-tender mobile mass in the left submandibular region	FNAC: cellular smears composed of epithelial cell clusters with a population of oncocytic cells in sheets and papillary clusters alongwith abundant single cells. Moderate to abundant densely stained granular cytoplasm with round nuclei and nucleoli were seen. Diagnosed as Oncocytoma of the submandibular gland	A firm nodular ovoid mass was received which had a uniform grey white appearance on cut section	Sheets of polygonal cells with small rounded nuclei and intensely staining eosinophilic granular cytoplasm thus confirming the cytological diagnosis of oncocytoma.	Salivary gland oncocytoma	-

8.	Hard palate	55years male with an asymptomatic swelling on the hard palate (1 yr)	FNAC: performed showed cohesive clusters of ductal cells and fragments of chondromyxoid stroma with a fine fibrillar structure. Diagnosis: Pleomorphic adenoma	Surgical excision with adequate margins was done	Showed a low-grade tumor with tubular, papillary and papillary-cystic areas. One single cell type was seen forming these structures with round pale nuclei, fine evenly distributed chromatin, indistinct nucleoli and no mitotic activity. Necrosis was absent and the stroma was eosinophilic and hyalinized in nature.	Polymorphous Low Grade Adenocarcinoma	S-100 protein and Vimentin positive
9.	Maxillary sinus	25years male presented with facial swelling and nasal stuffiness	Not done	Specimen consisting of left side of maxilla with a fungating tumor measuring 4X4.5X2.2 cms Cut section showed a grey white solid, lobulated tumor. Maxillary bone was seen to be infiltrated by tumor grossly.	Chondromyxoid lobulated tumor comprising of spindly and stellate cells lying in a myxoid background. Tumor cells show moderate pleomorphism, nuclear atypia & few mitotic figures.	Chondrosarcoma left maxillary sinus	-
10.	Tongue	25years male painless, nodular swelling on tongue	FNAC: moderately cellular, micro fragments of spindle cells with wispy cytoplasm uniform, elongated, wavy nucleus with fine chromatin, inconspicuous nucleoli. Background showed eosinophilic fibrillar material. Occasional Verocay body-like appearances were seen with nuclear palisading. Strongly suggestive of schwannoma.	0.5 X 0.5 cm soft tissue piece received	Histological section showed an encapsulated tumor composed of spindle cells with alternating hypercellular and hypocellular areas. The hypercellular areas showed bundles of spindle cells, with nuclear palisading around eosinophilic fibrillar material, Verocay body. Histological features thus confirmed the preliminary cytological diagnosis of schwannoma.	Lingual schwannoma	-
11.	Lip	6-year-old boy with single diffuse swelling(5X4cm) on upper lip extending to the buccal mucosa, progressively increasing in size. O/E: A lobulated firm, non tender, irreducible, non-pulsatile swelling with folding on the external surface. No café-au-lait spots were present over the body.		Reduction plasty specimen was received	Myxoid neurofibromatous lobules with central nerve bundles	Plexiform neurofibroma of lip	-
12.	Face (nose)	55/M with a slow-growing nodule on the right side of the nose for 3 years. O/E: ulceration		3 X 0.5 cm fungating, ulcerated soft tissue piece was received	Showed a predominant adenoid pattern with thin strands of basaloid cells in a reticulate arrangement, many tubules and few	Adenoid basal cell carcinoma	Anti-bcl-2 antibody showed homogenous cytoplasmic staining

		of the nodule and superficial bleeding since the last 10 days. H/O long hours of sun exposure. (labourer)			cystically dilated spaces containing mucin seen. Peripheral palisading of the basaloid cells was seen in some of the islands along with retraction artefact of the surrounding stroma and connection to the overlying epidermis was evident.		throughout the lesion. Staining for S-100, epithelial membrane antigen (EMA), carcinoembryonic antigen (CEA) and cytokeratin (CK-7) were found to be negative.
13.	Face	20-year-old male presenting with three papillomatous growths on the face, localized over the left frontotemporal region (1.2×1×0.5 cm), below the right eye (1.5×1 cm) and over the right eyebrow		Growth was covered with the hair-bearing skin and revealed a black nodule on cut section	Left fronto-temporal mass Growth showed a tumor arising from the epidermis, infiltrating the dermis. Nests and sheets of cells with a high nuclear-cytoplasmic ratio, eosinophilic macronucleoli, abundant cytoplasmic melanin pigment were seen. The tiny growth over the right eyebrow also showed similar morphological and immunohistochemical profile as that of the left fronto-temporal mass, consistent with a diagnosis of MM thus confirming the metastasis from malignant melanoma Papillomatous growth below the right eye revealed an atypical proliferation of basophilic cells arising in the epidermal basal cell layer and infiltrating the underlying dermis in nests, cords, and solid nodules. Deposits of the melanin pigment were scattered throughout the lesion. The stroma was fibrous and retraction clefts were present at the periphery of the tumoral nests.	Collision tumors of malignant melanoma and pigmented bcc.	cytoplasmic immunoperoxidation for HMB-45 Tumor cells revealing cytoplasmic immunoperoxidation for Melan-A Expression of bcl-2 in BCC
14.	Floor of mouth	57/M presented with a swelling in the floor of mouth		Wide Local Excision with marginal mandibulectomy specimen was received	The lining epithelium showed features of severe dysplasia and keratinizing SCC along with foci of adenocarcinoma in the deeper portion	Adenosquamous Carcinoma-floor of mouth	-
15.	Infraauricular region	A 16-year-old female patient with complaints of right infra-auricular mass for 5 months. On examination, an ulceroproliferative growth was seen with non-healing ulcer of the	FNAC reported it as undifferentiated carcinoma. Imaging modalities revealed a 6 × 6 cm mass lesion involving the right parotid region	Specimen received: Wide right neck composite resection with excision of the mass. Specimen was a gray-white, globular, soft-tissue mass, partially covered with skin, and	Large atypical cells arranged in sheets with bizarre, anaplastic forms, along with numerous multinucleated giant cells. Typical hallmark tumor cells with lobulated (embryo-like) nuclei and a prominent nucleolus. Mitosis was brisk with several atypical form	Extranodal anaplastic large cell lymphoma	Nuclear and cytoplasmic ALK positivity in tumor cells along with CD30 positivity

		overlying skin of the node involvement was seen.		measured 7 × 6 × 4 cm, ulcerated on the surface. Cut surface had a variegated firm yellow white tumor infiltrating the skeletal; muscle			
16	Maxillary sinus	A 26 year old male patient presented with prolonged nasal obstruction, difficulty in breathing and pain in eye, examination and imaging showed a nasal mass which extended into maxillary sinus, orbit and ethmoid sinus with presence of proptosis		Total maxillectomy with orbital extenteration was performed with excision of mass from nasal cavity upto cribriform plate and complete specimen. A 12x5x3.5 cm mass , grey white in color with variegated areas, friable in consistency with a papillary appearance.	Tumor showed a variegated lesion with carcinomatous, sarcomatous and teratoma like elements. The carcinomatous elements comprise of malignant glandular structures (adenocarcinoma) along with spindle cell areas which showed atypical changes with multinucleated osteoclast like giant cells. Neuroepithelial areas with presence of both pseudo and true rosettes and primitive blastemal-like cells in sheets were present in between adenocarcinomatous areas.. The tissue attached to orbit and optic nerve showed tumor with similar features. These histologic features suggested the diagnosis of sinonasal teratocarcinoma.	Sinonasal teratocarcinoma	Epithelial components stained positive with Cytokeratin while mesenchymal component was positive for Vimentin.

LEGENDS Adenosquamous Carcinoma- floor of mouth

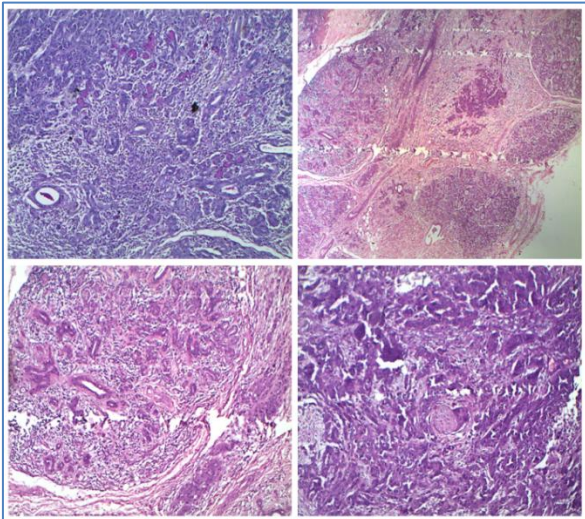


Fig-1: Microphotograph showing area of keratinizing stratified squamous cell carcinoma in the superficial part along with area of adenocarcinoma in the deeper portion (H&E)

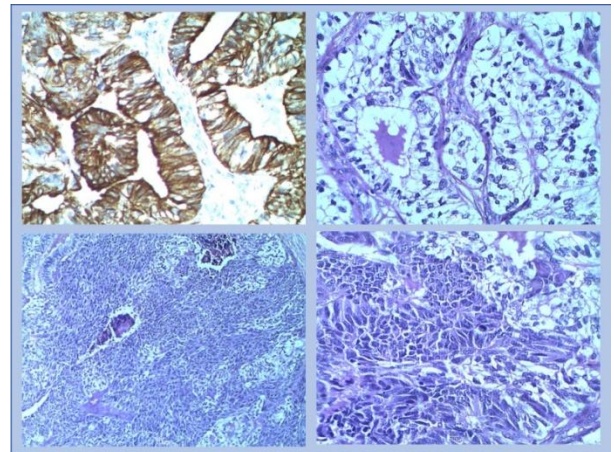


Fig-2: Poorly differentiated Carcinoma thyroid

Microphotograph upper left showing tumour area with adjacent normal thyroid tissue (H&E 10X), upper right shows peritheliomatous pattern of growth with areas of necrosis (H&E 10X), lower left & right shows large atypical cells with high N/C ratio and marked pleomorphism (H&E 40X).

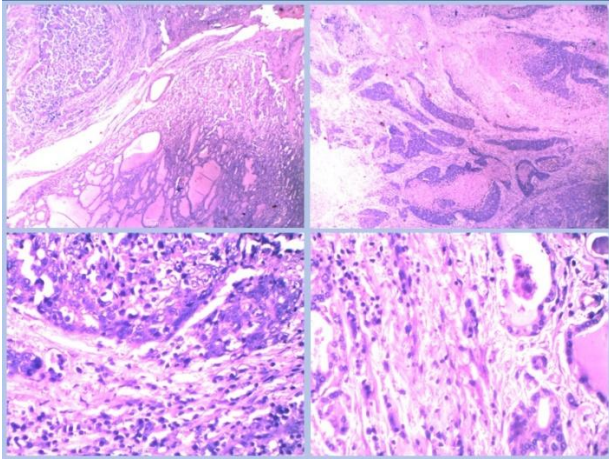


Fig-3: Sinonasal Teratocarcinosarcoma

Microphotograph shows malignant glandular elements, spindle cell areas and Pseudo and true rosettes formation. On IHC cytokeratin positivity seen

DISCUSSION

Head and neck neoplasias account for a significant number of cases associated with high morbidity and mortality, they encompass numerous rare malignancies posing a diagnostic challenge to the pathologist. In this surfeit of cases there was a varied age range ranging from 6 year old to 78 years, with significant male to female preponderance having 12 cases diagnosed in males and only 3 in females.

Case 1

Worldwide, conjunctival SCC is an uncommon disease, the incidence of which varies geographically from 0.02 to 3.5 per 100 000[5]. The term ocular surface squamous neoplasia (OSSN) was introduced to encompass the spectrum of conjunctival and corneal intraepithelial neoplasia (CIN) and SCC. Squamous cell carcinoma is the end stage of this spectrum; it has deleterious health problems ranging from loss of vision to death of the patient in severe cases. The disease progenitor cells appear to be the limbal epithelial cells. OSSN is a significant ophthalmic health problem [6] Histopathology remains a gold standard for its diagnosis and timely intervention with early diagnosis, surgery with excision of margins and adjuvant cryotherapy can prevent major eye damage with a favourable outcome[5].

Case2

Hidrocystoma also known as cystadenoma, a Moll's gland cyst, and a sudoriferous cyst is an adenoma of the apocrine sweat glands. It is an uncommon lesion accounting for 5% of eyelid masses and is a choriostomatous lesion. The cause is obstruction of excretory ducts with retention of clear fluid [7]. Eyelids are the site of presentation as in our case. On histopathology a bilayered cuboidal to columnar epithelium having eosinophilic cytoplasm and focal

apical snouting is seen, the microscopy of this report had typical features of a sudoriferous cyst.

Case3

The thyroid malignant tumors present as a spectrum having the lethal anaplastic thyroid carcinoma (ATC) at one end and the indolent well differentiated thyroid carcinoma (WDTC) at the other end [8]. Based on morphology, behavior and prognosis Poorly Differentiated Thyroid carcinoma (PDTC) has an intermediate behavior between WDTC and ATC [9]. The mean age at diagnosis is 5th and 6th decade and is slightly more common in females although the age at presentation in this case was younger (42years) than the defined age in literature. PDTC cytological diagnosis is a difficult one because of its subtle cytological features, rarity and inexperience as in our case on cytology it was diagnosed a colloid goiter and lymphocytic thyroiditis on two FNACs [10]. PDTC arises from the follicular cells, having an insular or trabecular growth pattern and a similar pattern was seen in the microscopy of this case. It may arise from a follicular or papillary carcinoma and according to WHO classification it falls under the category of morphological variant of follicular carcinoma. There was always a diagnostic challenge due to lack of an equivocal diagnostic criteria and rarity due to which a consensus meeting was then held in Turin, Italy, where an agreement was reached concerning the diagnostic criteria for PDTC so as to differentiate, ascertain and recognition of this category in order to provide appropriate management and have a better prognosis. The criteria includes

- (1) Presence of a solid/trabecular/insular pattern of growth,
- (2) Absence of the conventional nuclear features of papillary carcinoma, and
- (3) Presence of at least one of the following features:
Convolved nuclei;
Mitotic activity $\geq 3 \times 10$ HPF; and
Tumor necrosis [9]

PDTC's are immunoreactive for thyroglobulin with a predominant dot-like paranuclear pattern, and for malignancy-related markers such as HBME-1[11]. The three main differential diagnoses are Medullary Carcinoma, ATC and Carcinoma showing thymus like differentiation (CASTLE). Medullary carcinoma is Congo red positive due to amyloid in stroma, calcitonin positive and thyroglobulin negative. ATC lacks the insular growth pattern and may show giant, spindled or squamous cells. CASTLE on the other hand is TTF-1, thyroglobulin and calcitonin negative. In our case thyroglobulin positivity was present besides fulfilling the morphological diagnostic criteria thus confirming our diagnosis.

CASE 4

Extramedullary plasmacytomas (EMP) are localized plasma cell neoplasms with origin in tissue

other than bone. It is a very rare entity and more so thyroid gland is one of the rarest sites for its occurrence. Its diagnosis is made at a particular site once Multiple myeloma diagnosis has been ruled out through appropriate investigations like normal bone marrow, absence of osteolytic bone lesions on X ray and normal serum protein levels on electrophoresis. Galieni *et al.* has established diagnostic criteria for EMP:

- monoclonal plasma cell histology on tissue biopsy,
- plasma cells in the bone marrow representing < 5% of all nucleated cells,
- absence of lytic skeletal lesions or other tissue involvement,
- lack of hypercalcemia or renal failure, and
- a low level of serum M protein, if present[12]
- All these criteria were met in our case and thus a diagnosis of EMP was given.

CASE 5

Spindle cell carcinomas (SPC) of the larynx are very rare and unusual tumors and due its rarity are often misdiagnosed as mesenchymal malignancies or reactive lesions. SPC's are a highly malignant variant of squamous cell carcinoma with an incidence of 2-3% of all laryngeal malignancies. According to WHO classification these are biphasic tumors, comprising of a squamous cell carcinoma, in situ or invasive and malignant spindle cell component with mesenchymal appearance [13]. Glottic involvement and polypoid/pedunculated appearance are seen in majority of cases, the finding concomitant with our case. The diagnosis relies on histological demonstration of squamous cell component and spindle cells with sarcomatous appearances with immunohistochemically confirmation of the epithelial and mesenchymal markers. Epithelial markers include keratin (AE1/AE3, CK1, 8, 9), epithelial membrane antigens, KI, and K18. Mesenchymal markers include vimentin, desmin, S-100, Osteopontin, and BMP (2, 4)[13]. In this case as well tumor cells were positive for AE1/AE3, Vimentin, S-100, however they were negative for ALK-1 confirming our diagnosis. The interesting fact in this case which it even more unusual was the coexistence with tuberculosis in the cervical lymph nodes. Thus emphasizing on the point that tuberculosis is ubiquitous & in endemic regions, enlarged regional lymph nodes in malignancy are not always indicative of metastatic disease [14].

CASE 6

Clear cell carcinoma (CCC) of the salivary glands is a very rare carcinoma, accounting for 1% and is a recent addition to WHO classification of salivary gland tumors [15]. It can occur at various sites including tonsillar sulcus, oral mucosa, parotid and jaw but the most common site of occurrence is minor salivary gland as seen in our case as well. CCC is a low grade carcinoma which grows slowly and is defined as

a malignant epithelial neoplasm having single monomorphic population of cells with clear cytoplasm [16]. Clear cells are seen in numerous benign as well as malignant salivary gland lesions thus the differential diagnosis is a must, these include oncocytoma, mucoepidermoid carcinoma, acinic cell carcinoma, sebaceous adenoma, sebaceous carcinoma and epithelial adenoma [15]. The cytoplasmic clearing is due to glycogen and thus reacts with periodic Acid Schiff (PAS) stain, these clear cells are thought to originate from the myoepithelial cells or the intercalated duct cells. CCC's diagnosis is mainly made by histological features itself but special stains and IHC too are contributory for ruling out the differential diagnosis. Mucoepidermoid carcinoma shows Alcian blue and mucicarmine stain positivity as the clearing is due to hydropic degeneration in the squamous component, however the clear cells with foamy cytoplasm are seen in Sebaceous Adenoma and they are positive for fat stains as the clearing is due to lipid droplets. Oncocytomas on the other hand are well circumscribed with only foci of oncocytic cells and the clearing is due to glycogen [17]. CCC usually reaches large sizes before being diagnosed as they are slow growing and mainly asymptomatic, similar finding was present in our case the size increased considerably before being diagnosed. CCC is an extremely rare and a recent addition to the classification with varied differentials therefore a thorough histological examination and its awareness is essential in achieving a final diagnosis.

CASE 7

Amongst salivary gland neoplasms Oncocytomas constitute a miniscule of approximately 1.5% of all salivary gland tumors [18]. It is a benign tumor predominantly seen in the parotid (80%) and are quite rare in submandibular gland (9%) making this case an even more rarity [19]. The predominant affected age group is 5th to 7th decade making our case a very unusual one as in this occurrence was seen in a 17 year old male. FNAC smears of oncocytoma shows cohesive clusters of cell with granular cytoplasm; the arrangement is in sheets and papillary clusters. The uniqueness of the case lies because of the rae location and the younger age of the patient.

CASE 8

Polymorphous low grade adenocarcinoma (PLGA) is an indolent malignant tumor affecting the minor salivary glands almost exclusively. It is characterized by low metastatic potential, infiltrative growth pattern, cytological uniformity and morphological diversity[20] PLGA comprises 9-26% of all salivary gland malignancies, constituting as the second most common primary minor salivary gland malignancy after mucoepidermoid carcinoma.²¹ PLGA is found to have myoepithelial differentiation and thus it is of utmost significance to differentiate it from other salivary gland tumors having myoepithelial differentiation like pleomorphic adenoma (PA) and

adenoid cystic carcinoma(ACC)[20]. There lies a problem in distinguishing PA with PLGA due to presence of infiltrative margins where the capsule is incomplete and also presence of hyaline in the stroma in a limited PLGA specimen can be mistaken for mucomyxoid matrix of PA and a wrong diagnosis can be rendered, a similar thing happened in the present case[22]. IHC stands as an important tool in its differential diagnosis, PLGA cells show positivity for CK 7, Vimentin and S100 while being negative for glial fibrillary acid protein (GFAP), however PA is positive for GFAP and no immunoreactivity to Vimentin [20]. To conclude the pathologist should be aware of the characteristic morphologic and IHC findings of PLGA

in order to differentiate it from PA, thus an inquisitive work up with adequate size biopsy and IHC are ideal for its diagnosis.

CASE 9

Chondrosarcoma (CS) constitute second largest group of bone tumors accounting for less than 10% occurring in the head and neck region and a miniscule of 0.1% of total head and neck cancers. It is malignant cartilage forming tumor, mostly arising in flat bones and peripheral long bones. They have been divided based on their location, histological features and degree of cellular differentiation:

Criteria	1.	2.	3.	4.
Location	Central	Peripheral	Juxtacortical	
Histological features	Mesenchymal	Clear cell	Myxoid	Dedifferentiated
Cellular differentiation	Low grade	Medium grade	High grade	

Evans *et al.* devised a histological classification segregating CS into grades:

Grade I: Low grade/ Well differentiated – Having small densely stained nuclei often containing multiple nuclei within one lacuna

Grade II: Moderately differentiated – having increased cellularity, significant proportion of cells with moderately sized nuclei but having a low mitotic rate (< 2 mitoses/field)

Grade III: Poorly differentiated- having >2 mitoses/HPF and nuclear size greater than grade II including dedifferentiated tumor [23].

CASE 10

Lingual (Intraoral) schwannoma is a rare finding and on extensive review of literature it was found to constitute an incidence of less than 1%. Numerous differentials like traumatic fibroma, neurofibroma, leiomyomas, granular cell tumors, hemangiomas, rhabdomyomas, lymphangiomas, pyogenic granulomas and benign salivary gland neoplasm should be excluded and differentiated from Schwannomas.²⁴ However this being an unusual site a diagnosis of lingual schwannoma should be kept in mind while diagnosing painless tongue nodules as in the present case scenario.

CASE 11

Neurofibromatosis (NF) an autosomal dominant disorder has been divided into two categories dermal (single peripheral nerve origin) and plexiform (multiple nerve bundles) by World Health Organization. Plexiform Neurofibroma (PN) is characterized by proliferation of Schwann cells that too from the inner aspect of nerve sheath and these results in a distorted, irregularly thickened and a tortuous structure. Oral manifestations are seen in a miniscule of 4-7% of

patients suffering from NF, amongst that too gingiva as a location was found in only one other case report [25]. Solitary PN should therefore be kept as a differential by pathologist in diagnosing asymptomatic lip swellings.

CASE 12

Basal Cell Carcinoma (BCC), a common cutaneous tumor has numerous morphological variants, namely adenoid, morphoeiform, cystic, infundibulocystic, clear cell, pigmented, signet ring cell, adamantoid, giant cell, schwannoid etc all together constituting less than 10% of all BCC's[26]. A close perusal of literature showed an incidence of only 6.67 % of the adenoid variant amidst all the histological subtypes, the occurrence in its pure form is even more rarely seen [27]. The two important differentials are trichoepitheliomas and adenoid cystic carcinoma. In adenoid BCC homogenous cytoplasmic staining is seen throughout the lesion with anti-bcl-2 monoclonal antibody helping in differentiating from tricoepitheliomas. To exclude adenoid cystic carcinoma as a diagnosis S-100, EMA, CEA, CK-7 were done and found to be negative[26]. There should be an awareness of entities like cutaneous Adenoid cystic carcinoma and primary cutaneous apocrine carcinoma are the histological mimics and IHC along with clinical findings helps in achieving an accurate diagnosis.

CASE 13

Two contrasting primary malignant neoplasms presenting in a same patient simultaneously is an uncommon finding, here we report a case of both malignant melanoma (MM) and BCC in the same patient with the presence of metastasis from the melanoma focus at the same site. The abutment of BCC and MM is a very rare and uncommon finding and thus in such a cases while rendering a diagnosis the possibility of conjunction of two separate neoplasms should be kept in mind [28].

CASE 14

According to the WHO classification Adenosquamous Carcinoma (ASC) is a variant of squamous cell carcinoma. Its occurrence is usually as a mucosal carcinoma and is an uncommon malignant tumor of the head and neck region. For the histopathological diagnosis of ASC Alos *et al.* has stated the following diagnostic criteria:

- the most common component is usually keratinizing SCC;
- the second component is adenocarcinoma in the deeper portion; and
- Severe dysplasia or carcinoma in-situ (CIS) is seen in the surface epithelium [29].

All the three criteria were fulfilled in our case thus confirming the diagnosis.

CASE 15

Primary systemic anaplastic large cell lymphoma (ALCL) is designated as the neoplastic proliferation of lymphoid cells having an anaplastic appearance. It is an aggressive lymphoma characterized by presence of large pleomorphic “hallmark” cells having a kidney or horse shoe shaped nuclei with a constant CD30 expression on all neoplastic cells [30]. On extensive search of literature a report by Gustafson *et al.* was found of a 6 year old girl which was initially diagnosed as a neuroblastoma, they did not assess CD30 and ALK as there was no histological suspicion of ALCL but later on perusal it was considered and CD30 and ALK positivity confirmed the diagnosis. ALCL has been found to mimic non lymphomatous soft tissue tumors like rhabdomyosarcoma due to its varied spectrum of morphology presenting as diagnostic challenge to the histopathologist[31]. To conclude with while diagnosing pediatric soft tissue tumors ALCL should be considered as a differential and IHC should be applied in order to achieve the diagnosis without delay.

CASE 16

Sinonasal teratocarcinosarcoma is an extremely rare and aggressive neoplasm of paranasal sinuses. Characterized by varied histology, it comprises of combined histological features of teratoma and carcinosarcoma. Sinonasal teratocarcinosarcoma is a highly malignant neoplasm with poor long term survival. It is a rare tumor and extensive sampling is necessary to avoid misdiagnosis [32]. It occurs almost exclusively in nasal cavity and paranasal sinuses with occasional involvement of orbit, oral cavity and pharynx. This neoplasm was first described by Shanmugaratnam *et al.* as carcinosarcoma in 1983[33]. Histologically, the tumor show diverse morphology, comprising of carcinosarcoma with teratoid elements [32]. The most common is the epithelial component comprising of glandular or ductal structures and squamous epithelium, which may show fetal like clear cells. This immature fetal like squamous component is

quite characteristic of this tumor. The mesenchymal component may show fibroblastic or myofibroblastic cells with mature and immature osteoid, cartilaginous areas, smooth or skeletal muscle, these may show variable degree of atypia. The teratoid component includes fetal like clear squamous cells, organoid structures and neuroepithelium which may show rosettes or immature blastemal like cells.

CONCLUSION

The purpose of this article is to bring an awareness of these rare and uncommon entities so as to achieve cancer control activities and expedite the diagnosis and management to give utmost patient care. The need of the hour is to prioritize, control and early detection of head and neck cancer in India with special emphasis on these uncommon entities.

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