

flattened, with weakly eosinophilic & markedly vacuolated cytoplasm. Nucleus is eccentric with fine chromatin and small, central nucleolus and little to no mitotic activity. Cells may form solid cords alternating with channels having dilated lumina *simulating vascular structures*. Cystically dilated gaping spaces with no evident lining, representing a necrotic tubular component, and smaller gland-like spaces (giving the appearance of vascular spaces) are major clues to the diagnosis. Stroma is *prominent and fibrous*, with abundant smooth muscle and elastic fibers. Additional stromal features may be *prominent lymphoid follicles*, extensive necrosis, abundant desmoplasia and *signet-ring like cells* (in clusters or individually scattered).

The first cytological description of adenomatoid tumors was given by Perez-Guillermo et al in 1989. Cytology reveals *sheets, cords, glandular patterns or multilayered clusters of monotonous, round to oval cells* with pale, vacuolated cytoplasm. Nucleus is spherical & eccentric with fine chromatin and small central nucleolus^{4,5}.

They are considered to be of *mesothelial* origin (as originally proposed by Masson et al), which is supported by immunohistochemical studies (positivity for HMBE1 & Calretinin) and genetic analysis of *Wilms tumor-1 gene expression*¹. *These tumors show positive immunoreactivity for Calretinin, Epithelial markers (AE1/AE3, EMA, Cam5.2, CK5/6, CK7), Vimentin, WT1 (Wilms' tumor 1), HMBE 1 (Mouse Anti-human Mesothelial cell marker), D2-40 (Podoplanin)*³ and are negative for Carcinoma markers (CEA, CD15, B72.3, MOC-31, Ber-ep4, LeA 135), Endothelial markers (FVIIIIRA, CD31, CD34), Germ cell tumor markers (OCT 3/4, Nanog, Sox-2, AFP, PLAP) and Inhibin (helps exclude adrenal cortical & sex cord-stromal neoplasias)⁶. Diagnosis is mainly based on histopathology,

aspiration cytology, IHC and ultrasonography. The differential diagnosis includes epithelioid hemangioma, malignant mesothelioma, metastatic adenocarcinoma, papillary cystadenoma of epididymis, large cell calcifying Sertoli cell tumor, Epididymal carcinoma, testicular rhabdomyosarcoma, and carcinoma of rete testis. No nuclear atypia suggesting malignancy was present in our case.

Adenomatoid tumor should be suspected in any intrascrotal mass lesion. Its separation from testicular tumors is important as paratesticular tumors have good prognosis compared to testicular tumors. We are highlighting the strong correlation between cytology and histology features. Both diagnostic modalities reveal monotonous proliferation of tumor cells with similar cellular features. FNAC, as a preoperative diagnostic tool, can help to plan surgery as complete local excision of this benign tumor is both diagnostic as well as therapeutic and has had no reported case of recurrence or metastasis after excision⁵. The main clinical consideration is accurate diagnosis in order to prevent unnecessary orchiectomy and preserve endogenous testicular function^{5,6}.

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Kartagener's Syndrome: A Rare Case Report with Review of Literature.

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Abstract: Kartagener syndrome is a subset of primary ciliary dyskinesia, an autosomal recessive condition characterized by abnormal ciliary structure and/or function leading impaired mucociliary clearance. The findings of CT thorax, abdomen and PNS support the clinical diagnosis of the Kartagener's Syndrome. We report the CT findings of this rare syndrome.

INTRODUCTION

Kartagener's syndrome (KS) is a rare disorder which is seen in nearly half of the cases of primary ciliary dyskinesia. Primary ciliary dyskinesia is an inherited autosomal recessive condition characterized by bronchiectasis, sinusitis and otitis media. When situs inversus is associated with primary ciliary dyskinesia then it is referred to as Kartagener's

syndrome¹. The prevalence of Kartagener's syndrome is 1 in 32000 live births². No gender predilection is recognized³.

CASE REPORT

A 24 year male patient presented with complaints of productive cough, rhinorrhea and headaches since childhood with episodic fever and worsening of symptoms presented for CECT which revealed the following features: CT thorax with abdomen showed cystic dilatation of bronchi with thickened wall suggestive of bronchiectasis distributed predominantly in B/L upper lobes and right middle & lower lobes and dextrocardia with liver on the left side and the spleen on right side suggestive of situs inversus totalis. CT of Paranasal sinuses shows B/L maxillary sinusitis ;B/L middle and inferior turbinates showing polypoid changes with absent frontal sinuses

DISCUSSION

Kartagener syndrome is characterised by the clinical triad of situs inversus, chronic sinusitis and/or nasal polyposis and

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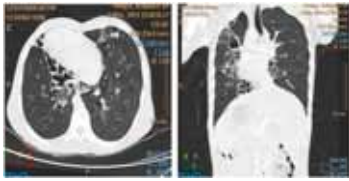


Fig 1&2 : Axial CT lung window showing cystic dilatation of bronchi with thickened wall suggestive of bronchiectasis distributed predominantly in B/L upper lobes and right middle & lower lobes

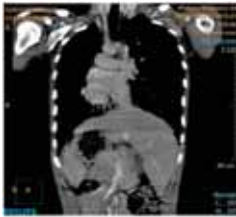


Fig 3: Abdominal CT coronal section showing dextrocardia with liver on the left side and the spleen on right side suggestive of situs inversus totalis and inferior turbinate showing polypoid changes with absent pneumatisation of frontal sinuses



Fig 4: Coronal CT of Paranasal sinuses shows B/L maxillary sinusitis; B/L middle

bronchiectasis. These features are attributed to abnormal ciliary motility.

Most important clinical manifestations include chronic upper and lower respiratory tract disease resulting from ineffective mucociliary clearance; the symptoms of chronic sinusitis, bronchitis, and bronchiectasis are more severe during the first decade of life but remit somewhat by the end of adolescence. Males are generally infertile because of immotile sperms, however some males have completely normal spermatozoa and cases of semi-sterility in females have been reported. Patients with Kartagener syndrome may also have anosmia.

Chest radiographs may illustrate bronchial wall thickening as an early manifestation of chronic infection, hyperinflation, atelectasis, bronchiectasis, and situs inversus that strongly suggests Kartagener syndrome.

CT is substantially more sensitive than chest radiography for showing bronchiectasis, which is characterized by lack of bronchial tapering, bronchi visible in the peripheral 1 cm of the lungs, and an increased bronchoarterial ratio producing the so-called signet-ring sign. According to appearance and severity, bronchiectasis can be classified as cylindrical, varicose, or cystic⁴. CT changes are milder than in cystic fibrosis⁵.

DIFFERENTIAL DIAGNOSIS

- 1) Malignancy
- 2) interstitial lung diseases: idiopathic pulmonary fibrosis and idiopathic interstitial pneumonia
- 3) conditions associated with bronchiectasis:
 - i) acquired (foreign body aspiration, tumor, lymphadenopathy, chronic obstructive pulmonary disease, and mucoid impaction)
 - ii) congenital bronchial obstruction (bronchomalacia, pulmonary sequestration yellow nail syndrome)
 - iii) recurrent infection (immunodeficiencies)
 - iv) cystic fibrosis
 - v) alpha-1 antitrypsin deficiency

CONCLUSION

CT scan of the chest, particularly high-resolution CT (HRCT) scanning, has gained importance in severity grading and monitoring of Kartagener's syndrome for clinical management and intervention studies. Consideration should be given to this imaging technique early in the presentation of Kartagener syndrome, when a chest radiograph may not be sensitive enough to identify disease processes or when another differential is being considered⁶.

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