Kartagener's syndrome a rare cause of massive polyposis and recurrent sinusitis

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ABSTRACT

Kartagener's syndrome is a rare, autosomal recessive genetic ciliary disorder comprising the triad of situs inversus, chronic sinusitis, and bronchiectasis. The basic problem lies in the defective movement of cilia, leading to recurrent chest infections, ear/nose/throat symptoms, and infertility. We reported a case of young girl presented with recurrent sinusitis with massive polyposis which was not respond to usual treatment. Nasoendoscopic examination revealed very thick nasal secretion with massive nasal polyp which involve all paranasal sinuses confirmed by CT scan. Chest x-ray was performed due to high suspicious of Kartagener's syndrome clearly show dextrocardia. She underwent Functional Endoscopic Sinus Surgery to open all the sinuses and remove the polyp. Post operatively, regular follow up was needed to clear the thick secretion retained in the paranasal sinuses even though it was opened widely during the operation. She also developed recurrent polyposis despite of compliant steroid nasal spray administration. The correct diagnosis of this disorder in early life is important in the overall prognosis of the syndrome, as many of the complications can be prevented if appropriate management is done. It may exhibit variable and atypical clinical presentations and severity due to its multisystem involvement and reverse positioning of internal organs. Although there is no specific treatment for this clinical entity, failure to diagnose this may subject the patient to unnecessary repeated admissions, investigations and inappropriate treatment.

Rare angiosarcoma of inferior turbinate; a case report and literature review

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ABSTRACT

Angiosarcoma is a rare soft-tissue sarcoma. It is an aggressive, malignant endothelial cell tumour of vascular or lymphatic in origin. Angiosarcoma accounts for 2% of all sarcomas and over half of it occurred in head and neck region. Its treatment is challenging with a poor prognosis. We presented a case of angiosarcoma of inferior turbinate occurring in a 73-year-old man who presented with left neck swelling for two weeks with a friable mass arising from posterior end of left inferior turbinate. FNAC of the left neck swelling showed atypical cell with a suspicious of malignancy and Computed tomography scan of neck and paranasal sinuses showed an enhancing lobulated mass at the posterior aspect of left inferior turbinate. Histopathological examination of the mass revealed a vascular tumour in favour of angiosarcoma (FNCLCC Grade 3).