Nasal Chondromesenchymal Hamartoma masquerading as malignant paediatric tumour

ABSTRACT

Nasal Chondromesenchymal Hamartoma (NCMH) is a rare, benign tumour of sinonasal tract usually presents in infants. Even though it is benign in origin, its aggressive clinical behaviour and florid radiological finding always mislead the initial diagnosis, mimicking a malignant sinonasal tumour. We report a case of NCMH in a 2-year-old boy who was referred to by an ophthalmologist for progressive facial distortion, left eye proptosis, persistent left rhinorrhoea and epiphora. Biopsy from the nasal cavity which appeared to be lobulated mass on endoscopy examination confirmed it was Nasal Chondromesenchymal Hamartoma (NCMH). Surgical excision of the tumour was performed. The symptoms of NCMH depend on the tumour location, thus the clinical presentation can be varied. The only way to establish the diagnosis of NCMH is by histopathology and immunohistochemistry of the tumour. Complete surgical treatment is advocated once the diagnosis is confirmed. This benign tumour has an established relation with the DICER1 mutations, hence other DICER1 tumours need to be ruled out at the time of diagnosis.

Keyword: Paediatric nasal mass; Hamartoma; Chondromesenchymal hamartoma; DICER 1 mutation