Sinonasal Disease in Polyostotic Fibrous Dysplasia
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RESULTS

INTRODUCTION

Polyostotic fibrous dysplasia (PFD) and McCune-Albright Syndrome (MAS) are diseases marked by fibrous dysplasia of bone in multiple skeletal sites and are caused by the same somatic mutation of the GNAS gene.1 MAS has additional diagnostic characteristics of café-au-lait spots, precocious puberty and/or other hyper-functioning endocrinopathies (Figure 1). While craniofacial fibrous dysplasia is frequently present in these rare disorders,2 the significance of their sinonasal disease has not been well characterized and their optimal management has remained controversial.

The current investigation is a retrospective, cross-sectional and longitudinal analysis of a large cohort of patients with PFD and MAS aimed to further clarify the natural history, progression, clinical presentations, and effects of endocrinopathy with specific regards to their sinonasal disease.

METHODS AND MATERIALS

From 1998 to 2010, subjects diagnosed with PFD and MAS were enrolled into an IRB approved natural history protocol. Patients underwent baseline evaluation including history and physical exam, endocrine evaluation, and craniofacial computed tomography (CT) as indicated. The majority (88%) also underwent comprehensive otolaryngologic evaluation. Yearly follow-up was attempted. Six patients were excluded due to prior sinonasal surgery performed at an outside institution. Their medical records were reviewed and craniofacial CT scans were analyzed for the degree of sinonasal fibrous dysplasia (FD) which was graded based on a modified Lund-MacKay score (Table I). Subjects who had been followed for more than 4 years and had undergone serial CT imaging were included for longitudinal analysis. Statistical analysis was performed using Student’s t-test.

RESULTS

•Demographics and clinical manifestations are summarized in Table I.
•FD scored frequently involved the sinuses and/or nasal cavity (92%)
•Chronic nasal congestion and headache/facial pain were the most common symptoms and present in approximately 1/3 (Figure 4)
•Only chronic nasal congestion & hyposmia significantly correlated with the severity of sinonasal bony disease. (Figure 4)
•Growth hormone (GH) excess and hyperthyroidism were both associated with significantly more severe bony disease, while precocious puberty was not. (Figure 5)
•Radiologic disease progression was more common in younger subjects (Table III) and was both rare and minimal after adolescence (Figure 6).
•Endocrinopathy and use of bisphosphonates did not demonstrate any significant effect on progression of disease.
•No major complications of sinus disease from FD were identified.

CONCLUSIONS

•Sinonasal fibrous dysplasia is common in PFD and MAS patients. Involvement of the sinuses and/or nasal cavity was common (92%) while sinonasal symptoms were present only in a minority. Patients with GH excess and hyperthyroidism were found to have a significant correlation with an increased severity of their sinonasal bony disease. This is not surprising given these hormones critical role in bone growth and development.3,4 Progression of disease after puberty was rare and minor: endocrinopathy and bisphosphonate use had no significant effect. It is important to note the limitations of the grading system used which may not be sensitive enough to detect subtle changes of FD involvement.

•Optimal management of these disorders is controversial with some advocating for radical excision and others for more conservative intervention.2 The present study demonstrates that FD associated paranasal sinus symptoms, complications, and progression are infrequent and typically minor, thus supporting a less aggressive approach. While the potential for major complications exists, the authors recommend conservative management while reserving surgical treatment for significant symptoms or compression of vital structures.

REFERENCES