DESCRIPTION

A 13-year-old girl presented at our hospital with a 3-month history of nasal obstruction, headache, hearing loss and severe weight loss. She also had nasal obstruction with occasional bleeding and a persistent ‘nasal twang’. These symptoms had previously been referred to as chronic sinusitis. Oral inspection showed a bulging soft palate (figure 1) and nasal examination by anterior rhinoscopy detected a soft mass totally obstructing choanae. No cervical lymphadenopathy was palpable. Blood examination showed leukocytosis, elevated C-reactive protein and erythrocyte sedimentation rate, and high titres of Epstein-Barr virus (EBV) antibodies. Cranial MRI revealed a 7×3×8 cm nasopharyngeal mass, deeply vascularized and partially eroding the skull base (figure 2). Differential diagnosis included lymphoma, rhabdomyosarcoma, juvenile angiofibroma, germ-cell tumor and epithelial carcinoma. Histopathologic examination was consistent with undifferentiated carcinoma, a subtype of nasopharyngeal carcinoma (NPC), and the girl was immediately referred to the closest oncology department for appropriate treatment. The annual incidence of NPC in the UK is 0.3 per million at age 0–14 years, and 1–2 per million at age 15–19 years. Incidence is higher in the Chinese and Tunisian populations. Although NPC is a rare disease in children, representing less than 1% of childhood cancers, it does constitute 20–50% of pediatric malignancies of the nasopharynx. Aetiological factors include EBV, genetic susceptibility and consumption of food with possible carcinogens (volatile nitrosamines). NPC is classified pathologically into three subtypes: squamous cell carcinoma, typically found in the older adult population; non-keratinizing carcinoma and undifferentiated carcinoma (also known as lymphoepithelioma), which is nearly the only subtype found in children.
More than 80% of the children present with locoregional advanced disease, while clinically overt metastases are found in a small proportion of patients. Surgical approach is not indicated for the delicate anatomical position. The standard of care for locoregional disease includes radiotherapy (RT). Doses of 50–72 Gy are recommended for patients older than 10 years, and a 5–10% reduction in dose is indicated for younger children. Concomitant administration of cisplatin-based chemotherapy and RT has improved the outcome over the past four decades. However, many survivors had long-term treatment-related morbidities. Use of adjuvant interferon-β has also shown to be promising.

Figure 2  MRI scan. Axial T1-weighted sequence, showing a large mass growing in the nasal cavity and eroding occipital, temporal and sphenoid bones.

COMPETING INTERESTS None.

PATIENT CONSENT Obtained.

REFERENCES