ABSTRACT

A 10 year-old male presented to our clinic with a two-year history of progressive right nasal airway obstruction. He reported occasional headaches, but denied other complaints such as epistaxis, vision changes, diplopia, numbness, or pain. On examination, he was found to have an obstructive mass in the right nasal cavity. A CT (figure 1) and MRI (figure 2) showed a well-defined 53 mm by 67 mm expansile lesion arising from the right nasal cavity with partially calcified internal matrix. The patient was taken to the operating room to undergo complete excision of the mass, which was accomplished via image-guided endonasal endoscopic approach. No infiltration or permissive pattern of invasion was present. The nuclei were small, dark and pyknotic and without atypia or mitotic activity. The microscopic appearance was diagnostic of enchondroma (figure 3A, B). On follow-up the patient has been stable with no new complaints. Endoscopic examination revealed a normal nasal cavity.

BACKGROUND

Enchondromas are benign intramedullary tumors composed of nodules of hyaline cartilage. They represent a distinct histological entity from chondromas, which occur in the soft tissues. Unlike chondromas, which are more frequently encountered sinonasal tumors, this is the first reported case in the literature of a solitary sinonasal enchondroma. Local recurrences are uncommon after gross total resection based on review of patients with multiple enchondromas. More data is needed to conclude whether solitary enchondromas can transform to malignant tumors, as has been reported in patients with enchondromatosis.

PATHOLOGY

Sections of the tumor reveal variably sized nodules of hyaline cartilage occupying and distending the medullary space (figure 3A). No invasive or permissive patterns were observed. At the periphery of nodules, formation of bone in the manner of enchondral ossification was present. Thinning of the cortex is present and tumor is seen adjacent to the submucosal space focally (Figure 3B). Cellularity is low to moderate. Chondrocytes display moderate amounts of clear cytoplasm and small dark pyknotic nuclei. Mitotic activity is not present, nor are areas of necrosis.

DISCUSSION

Enchondromas are typically incidental findings diagnosed only after radiographic examination for other, unrelated symptoms1,2. They represent a distinct histological entity from osteochondromas, and from soft tissue chondromas. In a large published series the risk for local recurrence after resection for conventional enchondroma was 5%3,4. Following surgical removal, which may require radical surgery, the prognosis may be good5. Recurrence is seen only in those in whom tumors were inadequately excised6. Malignant transformation in the absence of multiple enchondromatosis is quite rare6. In fact, this is a rare event, with only a few cases reported in the literature of solitary benign enchondromas dedifferentiating into higher grade osteosarcoma7. There is not enough literature of enchondromas of the nasal cavity and paranasal sinuses, and as such it is difficult to conclude about the recurrence or potential malignant transformation of these lesions. We propose that the best treatment approach for these solitary enchondromas of the sinonasal cavities is to obtain complete resection of the lesion with clear margins if possible in a similar manner as the treatment laid out by Chiu et al in management of inverted papillomas8.

BACKGROUND

Enchondromas are benign intramedullary tumors composed of nodules of hyaline cartilage. Solitary enchondromas are most often seen in the age group of 20-40 years with no sex or racial predilection1,2. Thirty-five percent of enchondromas arise in the hand1,2. There are many ways of classifying benign chondrogenic tumors, and the widely accepted classification is the one proposed by Lichtenstein1.

Enchondromatosis is a rare, heterogeneous skeletal disorder in which patients have multiple enchondromas2.

REFERENCES