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OSTEOSARCOMA OF THE MAXILLA

F.M. Vanhoenacker[1,2], M. Camerlinck[1,2], D. De Vuyst[1], K. Chapelle[3]

Key-word: Bone neoplasms

Background: A 25-year-old male patient presented with tooth pain and progressive swelling of his left cheek. Dental examination was unremarkable.

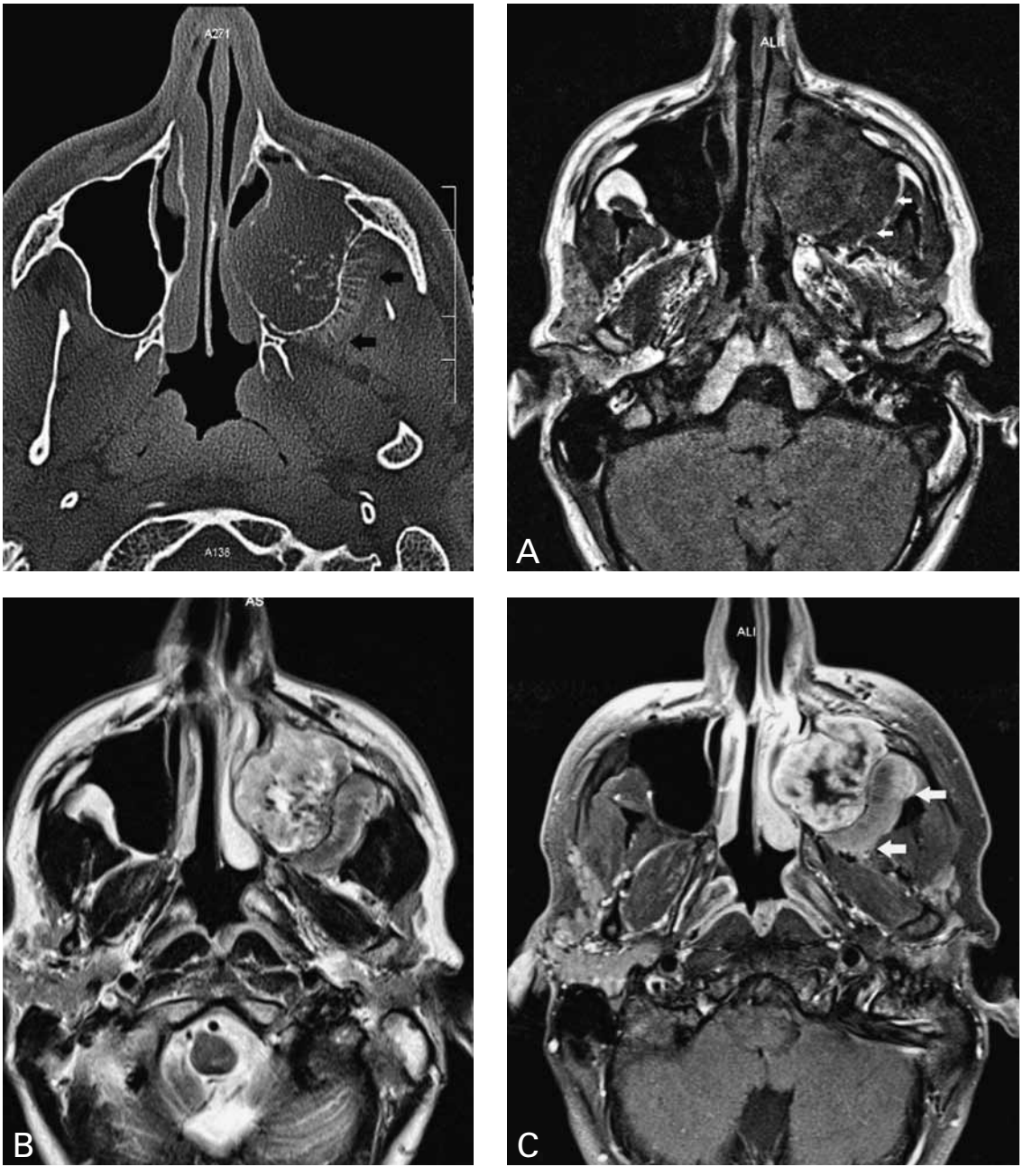

Fig. 1 | 2A / 2B | 2C

Departments of 1. Radiology, AZ Sint-Maarten, Duffel-Mechelen, Duffel, Belgium and 2. Antwerp University Hospital, Edegem, Belgium, 3. Department of Maxillofacial Surgery, AZ Sint-Maarten, Duffel-Mechelen, Duffel, Belgium

Work-up

CT scan of the paranasal sinuses (Fig. 1) shows opacification of the left maxillary sinus, with intralesional mineralized areas and spiculated ("hair-on-end") type of periosteal reaction (black arrows). MRI of the paranasal sinuses (Fig. 2) shows on A, axial T1-weighted image, a lesion slightly heterogeneous and predominantly isointense to muscle. Note soft tissue extension beyond the posterolateral wall of the left maxillary sinus into the infratemporal fossa (white arrows). On B, axial T2-weighted image, the lesion is heterogeneous appearance with areas of low, intermediate and high signal. Note also soft tissue extension within the infratemporal fossa. On C, axial fat-suppressed contrast enhanced T1-weighted image, the lesion shows marked and heterogeneous enhancement with peripheral papillary projections. The soft tissue component of the lesion is enhancing vividly (white arrows).

Radiological diagnosis

CT and MRI findings are consistent with an *osteosarcoma of the maxilla* (gnathic osteosarcoma). Histology after surgical biopsy confirmed a high grade osteosarcoma.

Discussion

Osteosarcoma (OS) is the most common primary malignant tumor of bone in adolescents and young adults, accounting for approximately 40-60% of all primary malignant bone tumors. The tumor may also be the result of malignant transformation of benign lesions such as Paget's disease, fibrous dysplasia or occurs after previous irradiation. OS of the jaw are designated as gnathic osteosarcomas. They constitute 6-13,3% of all skeletal OS. Gnathic OS is more prevalent between 20-36 years, whereas extragnathic OS is predominantly seen between the ages of 11 and 20 years. Gnathic OS are most commonly located in the body of the mandible, posterior alveolar ridge or maxillary antrum. The main symptoms are swelling and pain, but paresthesias, loosening of teeth, bleeding and nasal obstruction have been reported. Histologically, the main subtype is chondroblastic, followed by the osteoblastic subtype. Radiographically, gnathic OS is similar to conventional osteosarcoma, with presence of osteoid matrix in 60-80% of cases, aggressive periosteal reaction (such as a spiculated periosteal reaction) and soft tissue extension. Besides areas of sclerosis and mineralization, there are often associated areas of lytic bone destruction. Opacification of the maxillary sinuses is also a frequent finding in maxillary lesions. CT is the preferred imaging modality to demonstrate areas of mineralized osteoid, periosteal reaction and cortical breakthrough. MR imaging is more appropriate to assess both the intraosseous and extraosseous components of the lesion. Lesions are of low to intermediate signal intensity on T1-weighted MR images and are of heterogeneous signal intensity with T2-weighting. Mineralized areas are of low signal, whereas other components are of high signal. Foci of central hemorrhage are of high signal on all pulse sequences. Fat suppressed T1-weighted Gd-enhanced MR images are particularly helpful for delineating local tumor extension. Treatment of gnathic OS is difficult and includes a combination of surgical excision, radiation therapy and chemotherapy. Distant metastases, especially to the lungs, are less frequent than in other osteosarcomas. Current multidisciplinary treatment within a multi-center setting may result in long-term survival in over two-thirds of patients. Unfortunately, local recurrence is common, particularly in maxillary lesions and is often uncontrollable. Involvement of extragnathic sites and failure to achieve radical surgical resection are strong negative prognostic factors, leading to patient death.

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