A rare tumor of nasal bone in a child: osteoblastoma

Sumru Kavurt[1], Funda Çorapcıoğlu[1], Sema Bay[1], Emre Üstündağ[2], Gür Akansel[3], Bahar Müezzinoğlu[4]

[1]Division of Pediatric Oncology, Department of Pediatrics, and Departments of [2]Otorhinolaryngology, [3]Radiology, and [4]Pathology, Kocaeli University Faculty of Medicine, Kocaeli, Turkey

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Benign osteoblastoma is an uncommon bone tumor, representing 1% of all benign tumors and 3% of all primary bone tumors. Intranasal or paranasal osteoblastoma is particularly rare. Osteoblastoma occurs more frequently in males, at a peak age between 10 and 20 years. We report an osteoblastoma of the nasal septum in an 11-year-old boy who presented with swelling of the nasal septum. The diagnosis and management of this unusual lesion, as well as the histopathology and imaging characteristics, are reviewed. Magnetic resonance imaging (MRI) revealed a solid midline nasal mass originating from the septum with intense contrast enhancement. There was no evidence of osteoid or chondroid matrix mineralization on computed tomography (CT). Biopsy was performed. Histologically, the tumor contained abundant plump osteoblast-like cells, with eosinophilic cytoplasm and large nuclei that were frequently juxtaposed to the newly formed osteoid. Conservative excision was performed. The literature concerning occurrence of osteoblastoma in this unusual location is reviewed.

Key words: osteoblastoma, nasal septum, children.

Tumors of the nasal cavity are a rare but serious problem in the pediatric population. Nonspecific symptoms such as nasal congestion and rhinorrhea may persist for months before a nasal mass is suspected[1]. The differential diagnosis of nasal mass goes into a wide range of lesions varying from inflammatory proliferations and infective lesions to benign and malignant neoplastic conditions[2].

Osteoblastoma originating from the nasal cavity is very rare in childhood. It generally occurs in adolescents and young adults and arises within the medullary cavity, on the surfaces of the long bones or posterior of the spine[3-5]. Radiologically, osteoblastomas tend to be expansile and to remodel adjacent bone. Computed tomography (CT) is usually the examination of choice offering detailed information about the origin, extent and nature of the tumor. Osteoblastoma is usually a well-defined, expansile lytic bone lesion with sclerotic rim and a central portion with granular appearance due to punctate mineralization of the osteoid[6]. CT scanning and magnetic resonance imaging (MRI) are diagnostic but not pathognomonic. As a result, biopsy prior to resection is important in determining the diagnosis. Histologically, the most pathognomonic features are the presence of osteoblasts, small trabeculae of woven bone and rich vascular fibrous stroma[7].

To our knowledge, the nasal septum is a frequent location of osteoblastoma, but it is rare in childhood. We report a new case of benign osteoblastoma of the nasal septum with atypical CT findings. The pediatric oncologist must be aware of possible nasal septum involvement with this entity, and it must be kept in mind when a patient suffers from pain and swelling in the nasal septum.

Case Report

An 11-year-old boy was referred to Kocaeli University Pediatric Oncology Department because of swelling on the nasal dorsum. His medical history revealed nasal congestion and rhinorrhea for nearly two months. He

had been diagnosed as sinusitis and received antibiotics and decongestive treatment. However, his complaints were not relieved with symptomatic treatment, and during this period, a swelling developed on the nasal septum. MRI revealed a solid midline nasal mass originating from the septum (Figs. 1a, 1b) with intense contrast enhancement (Fig. 1c). There was no evidence of osteoid or chondroid matrix mineralization or calcified mass on CT (Fig. 2). A biopsy specimen of the tumor was obtained intranasally and the pathological diagnosis was an osteoblastic tumor suggestive of osteoblastoma. Local excision and curettage were performed under general anesthesia, and the bone mass was totally excised from the surrounding smooth tissue. Macroscopically, the excised material had multiple fragments of gritty, gray-tan tissue measuring 3x2x2 cm. Histologically, the tumor contained abundant plump osteoblast-like cells, with eosinophilic cytoplasm and large nuclei that were frequently juxtaposed to the newly formed osteoid. The fibroblastic stroma was intermingled with inflammatory cells, osteoclast-like multinucleated cells and abundant capillary vessels (Fig. 3). No cells displaying overt dysplastic features or abnormal mitotic activity were detected, and there was no cartilaginous differentiation. There was also no evidence of entrapped or infiltrated residual bone trabeculae, and the mass was diagnosed as osteoblastoma.

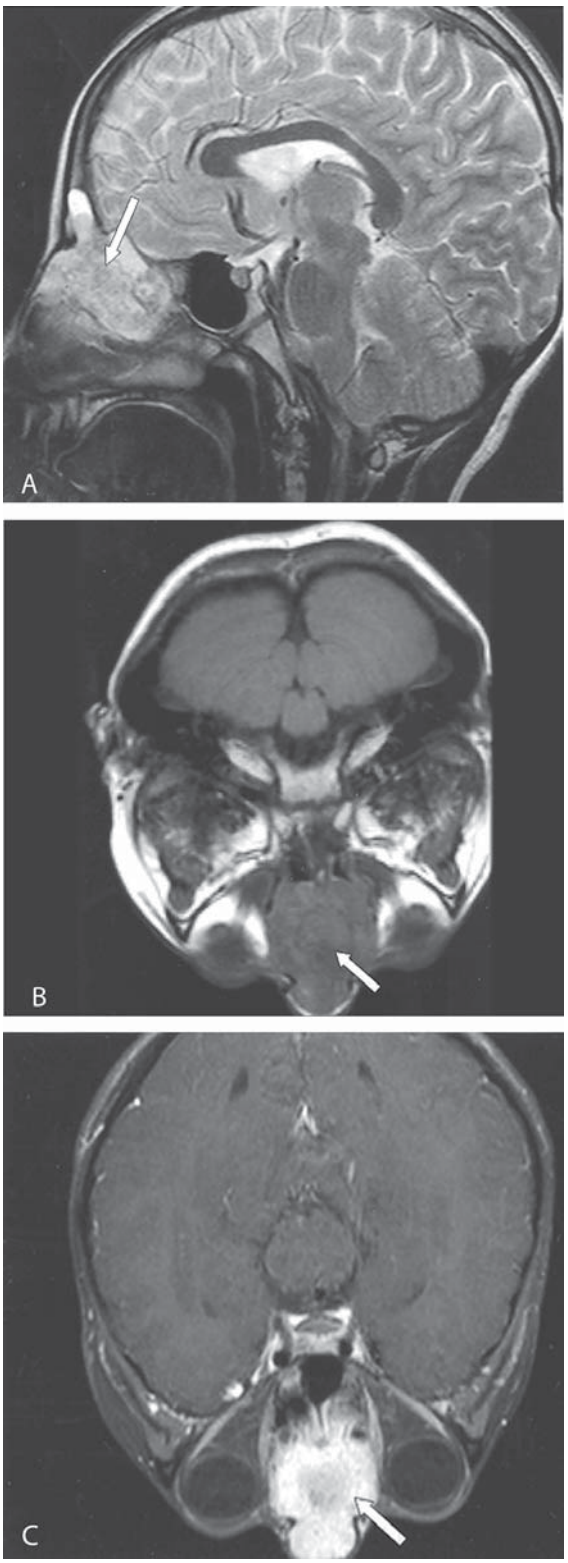

Fig. 1a-c. Sagittal T2-weighted **(1a)** and axial T1-weighted **(1b)** MR images show a solid mass originating from the nasal septum (arrows). Axial T1-weighted **(1c)** image after intravenous gadolinium shows bright, homogeneous enhancement throughout the lesion (arrow).

Discussion

Tumors of the nasal cavity in children are particularly challenging regarding diagnosis because of the nonspecific symptoms, which are often initially misdiagnosed as sinus infection and treated empirically. Treatment failure or the onset of new symptoms eventually prompts further diagnostic work-up, revealing the true underlying pathology[1]. The differential diagnosis of nasal mass includes a wide range of lesions varying from inflammatory proliferations and infective lesions to benign and malignant neoplastic conditions[2]. Benign lesions are generally congenital, such as the rare developmental midline nasal masses in children[8]. The most common malignant lesions arising in the nasal cavity in children are rhabdomyosarcoma, esthesioneuroblastoma, lymphoma, and malignant soft tissue and bone

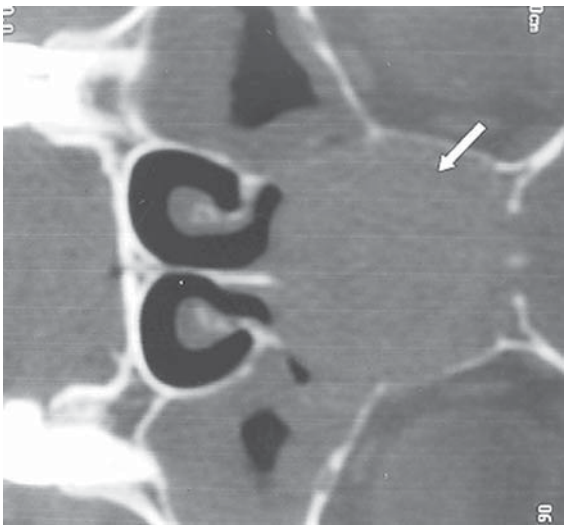

Fig. 2. Coronal CT image of the mass reveals the lack of mineralization of osteoid or chondroid type (arrow).

tumors[1,9]. We report here a patient suffering from nasal congestion and rhinorrhea for nearly two months; when swelling developed further, a diagnostic work-up was performed. Osteoblastoma is a rare, benign, bone-forming tumor that accounts for approximately 1% of all bone neoplasms and 3% of all benign primary bone tumors[10,11]. Intranasal or paranasal osteoblastoma is particularly rare. Osteoblastoma arising from the paranasal sinus and/or nasal cavity was reported in 21 cases in the literature[12]. Twelve of these were reported in children. Osteoblastoma occurs more frequently in males, at a peak age between 10 and 20 years[10,11]. The youngest patient was a three-year-old girl who had nasal bulging[13].

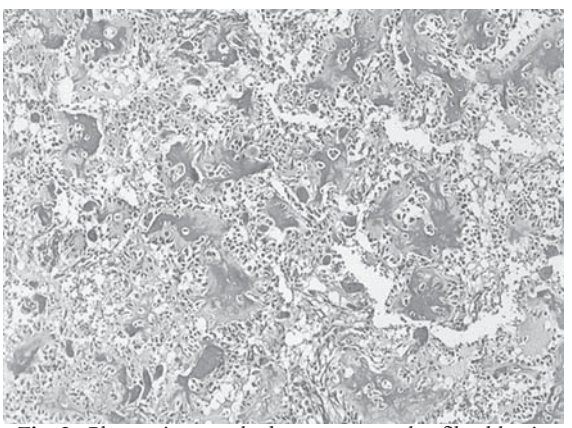

Fig 3. Photomicrograph demonstrates the fibroblastic stroma rich in vascular component having neoplastic osteoblastic cells forming irregular osteoid (hematoxylin-eosin, x200).

Clinically, the most common presenting complaint for patients with osteoblastoma is palpable mass and pain. Other common manifestations are gait disturbances, warmth and tenderness. Aggressive forms of osteoblastoma tend to present with more pain, likely due to localized areas of destruction[4]. The clinical presentation of benign osteoblastoma commonly features swelling and pain[4,13]. Our patient also suffered from swelling in the nasal septum.

The lesion remains fairly well-circumscribed without causing bone destruction[12,14]. One of the difficulties regarding diagnosis of osteoblastoma is that the radiographic features are not necessarily distinctive, creating problems in distinguishing them from osteosarcoma, osteoid osteoma, aneurysmal bone cyst, chondroblastoma, and chondrosarcoma[4]. Although plain radiographs are the basis of the diagnosis, the features can be nonspecific[10]. CT scan is a useful adjunct to the diagnosis and provides the most specific information about the lesion. It defines the tumor location and size and helps differentiate it from other benign and malignant lesions[10,11]. CT findings of osteoblastoma are very characteristic and consist of a well-circumscribed calcified mass with a thin cortical margin, and MRI findings include a decreased signal on both T1- and T2-weighted images[4]. Our case is highly atypical because of the lack of even a negligible amount of calcium detectable on CT. MRI is helpful in evaluating the extent of soft tissue involvement, defining the relationship to the surrounding neurovascular structures[15,16].

The biological behavior of benign osteoblastoma shows wide variations. Although considered benign, some lesions show aggressive behavior and rare malignant transformation into histologically typical osteosarcoma. It has been reported that one-fourth of benign osteoblastomas display radiographic features of a malignant neoplasm[5]. Furthermore, our patient's radiological findings were not compatible with a benign lesion.

On histological examination, the osteoblastoma appeared to be composed of small irregular bony trabeculae and osteoid containing abundant blood vessels. The trabecular bone is usually surrounded by numerous osteoblasts, and some giant osteoclastic cells are present

in the fibrous stroma[4,5,15]. The rich vascularity of the fibrous stroma accounted for severe bleeding in some surgical procedures[7].

Treatment of osteoblastoma with local conservative excision and curettage should be adequate. The procedure of choice depends on the size of the lesion, the severity of presenting signs and symptoms, the proximity of the tumor to adjacent structures, and prior procedures[3,5]. The use of adjuvant therapies for pediatric tumors of the nasal cavity depends on many factors, including the histopathologic tumor type, the initial extent of disease, and the status of postoperative margins[17]. Benign osteoblastoma is not usually life-threatening. Conservative surgical excision is the prime treatment for osteoblastoma[4,5]. In the current case, local excision and curettage were performed under general anesthesia.

In summary, osteoblastoma is rarely seen in the nasal cavity; nevertheless, it should be considered in the differential diagnosis in patients suffering from nasal congestion, rhinorrhea, pain, and swelling in the nasal cavity that cannot be explained by more common causes. To our knowledge, this is the first case of osteoblastoma with atypical radiological findings. While the reason for this remains unclear, it must be kept in mind, and histopathological examination should be done.

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