Case Report / Olgu Sunumu

Nasal chondromesenchymal hamartoma: a rare nasal benign tumor

Nazal kondromezenkimal hamartom: Nadir bir nazal benign tümörü

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ABSTRACT

Nasal chondromesenchymal hamartoma (NCMH) is a rare nasal benign tumor, which arises from the nasal cavity or paranasal sinuses. In this article, we present a five-year-old male patient with rhabdomyosarcoma unity in remission that emerged with nasal obstruction. Synchronous diagnosis of pediatric tumors such as pleuropulmonary blastoma in the literature is a remarkable finding. We found a mass within the left nasal cavity originating from superior portion of nasal septum, extending to the olfactory cleft and resected all tumor via endoscopic surgical approach. Histopathological diagnosis revealed that NCMH contained cartilaginous and mesenchymal components. In conclusion, NCMH is a rare surgically treated benign tumor that can be synchronously diagnosed with pleuropulmonary blastoma and should be kept in mind for differential diagnosis of unilateral pediatric nasal mass.

Keywords: Chondromesenchymal hamartoma; tumor; nasal benign tumor.

ÖZ

Nazal kondromezenkimal hamartom (NKMH) nazal kavite ya da paranazal sinüslerden kaynaklanan nadir bir benign burun tümörüdür. Bu yazıda burun tıkanıklığıyla ortaya çıkan ve remisyonda rabdomyosarkom birlikteliği gösteren beş yaşında erkek hasta sunuldu. Plevropulmoner blastom gibi pediatrik tümörlerin eşzamanlı tanısı literatürde dikkate değer bir bulgudur. Hastanın endoskopik muayenesinde sol nazal kavitede septum superior kısmından kaynaklanan ve olfaktör klefte kadar uzanan bir kitle saptandı ve endoskopik cerrahi yaklaşımla tümör tamamen rezeke edildi. Histopatolojik tanıda NKMH'nin kıkırdak ve mezenkimal komponentler içerdiği görüldü. Sonuçta, NKMH pediatrik tek taraflı nazal kitle durumunda ayırıcı tanıda akla gelmesi gereken nadir, plöropulmoner blastom birlikteliği eşzamanlı tanı konabilen ve cerrahi olarak tedavi edilen benign karakterli bir tümördür.

Anahtar Sözcükler: Kondromezenkimal hamartom; tümör; nazal benign tümör.

Nasal chondromesenchymal hamartoma (NCMH) is an extremely rare benign tumor of the nasal cavity and paranasal sinuses.[1] McDermott et al.[2] first suggested the term nasal chondromesenchymal hamartoma as a distinct pathologic entity first in 1998. This tumor has predominantly mesenchymal and cartilaginous components, and presents as polypoid lesions in the nasal cavity. Few cases in infantile, early pediatric, and adolescent populations have been reported in the literature. Patients usually present with symptoms such as nasal obstruction,

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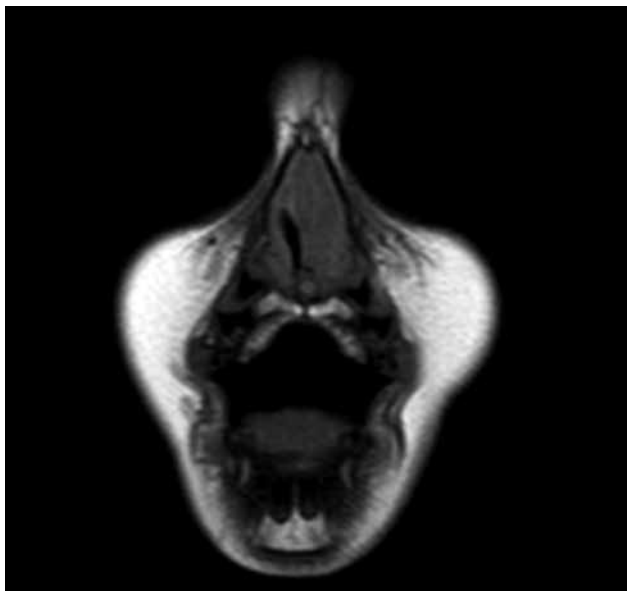

Figure 1. *The mass obliterating the nasal vestibule with a hypointense signal void on T_1 image.*

rhinorrhea and loss of smell. Complete resection is thought to be enough for treatment.

CASE REPORT

A five-year-old boy who presented with nasal obstruction and had recurrent sinusitis over the last year was referred to our clinic. An anterior rhinoscopic examination revealed a pink-bluish polypoid mass within the left nasal cavity originating from the superior portion of the nasal septum, extending to the olfactory cleft. There were no other pathologic findings on detailed physical examination or laboratory tests. The patient also had a history of chemotherapy because of rhabdomyosarcoma in the neck when he was aged three months. We learned that he underwent chemotherapy 10 times and had no anomalies or other malignancy. The lesion was heterogeneous with low signal intensity on T_1 magnetic resonance imaging (MRI) and was heterogenously hyperintense on T_2 weighted MRI. In addition to this, we identified that the tumor arose from the nasal septal cartilage on axial section of T_2 sequences (Figure 1, 2). We decided to perform an endoscopic surgical resection under general anesthesia. Intraoperatively, we determined that the lesion had occluded the left nasal vestibule and was attached to the anterior skull base. A total resection was performed. On histopathologic analysis, the mass consisted of irregular cartilage islands with mesenchymal spindle cells (Figure 3). The patient had no complications and was discharged from hospital on the second postoperative day. There were no further symptoms in the four months of follow-up.

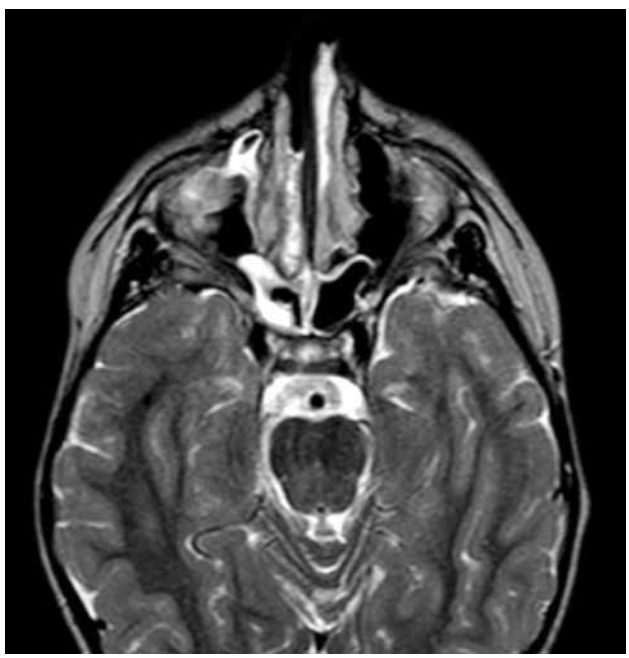

Figure 2. *On T_2 axial section, the mass is identified as originating from the nasal septum.*

DISCUSSION

Nasal chondromesenchymal hamartoma (NCMH) is an extremely rare, benign pediatric tumor. It has been referred to with

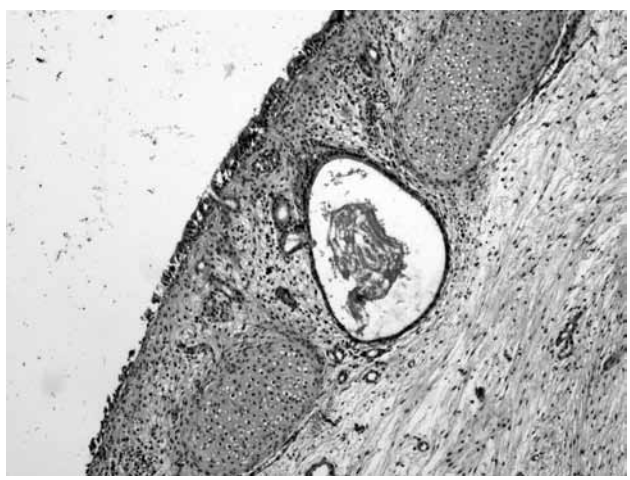

Figure 3. *The mass consists of irregular cartilage islands with mesenchymal spindle cells (5517 H-E x 100).*

such different nomenclatures as chondroid hamartoma, mesenchymoma, and nasal hamartoma. Nasal chondromesenchymal hamartomas are composed of cartilaginous and mesenchymal elements. Hamartomas are rarely seen in the head and neck region. Although NCMHs can have developmental or congenital etiologic origin, the exact pathologic process remains unclear. It has been predominantly linked to a genetic predisposition with a chronic inflammatory course or some kind of endocrinologic disturbance.[3-5] In the literature, NCMHs are invariably diagnosed in infants or in early childhood.[1,2,6-15]

Patients with NCMHs commonly present with an intranasal mass that causes symptoms such as nasal obstruction, rhinorrhea and epistaxis. The symptoms and clinical presentation of the patient are mostly determined by the size and location of tumor. Proptosis, enophthalmos or impairment of eye movement can be the presenting symptoms or findings in cases with orbital involvement.[2,6,9-14,16-19] Intracranial extension of the tumor can result in neurologic manifestations.[2,17] Symptoms or signs such as difficulties in respiration or nourishment, epistaxis, rhinorrhea, middle ear effusion can be encountered due to tumor size and site.[2,7,8,12,20] Our patient only had nasal stuffiness and epistaxis. He had no neurologic and ophthalmologic symptoms because there was no orbital involvement or intracranial invasion.

Radiologic diagnosis is often difficult and misdiagnosis is likely. There are too many differential diagnoses that should be taken into account. Hemangioma, angiofibroma, antrochoanal polyp, nasoethmoidal encephalocele, nasal glioma, inverted papilloma, giant cell reparative granuloma, ossifying fibroma, chondro-osseous respiratory adenomatoid hamartoma, and aneurismal bone cysts along with other rare benign pediatric tumors constitute the broad list of differential diagnose.[8,20-22] Exclusion of nasoethmoidal encephalocele and nasal glioma in differential diagnosis is considerably important so as to avoid neurologic complications of surgical therapy. On imaging sections of nasoethmoidal encephalocele, a bone defect is almost always seen with no destruction of the anterior cranial fossa, cystic lesions show a thin peripheral halo with soft central tissue.[21] Nasal glioma is associated with the brain and shows the same signal density as brain on MRI.[8,21] Furthermore, another pattern in the differential diagnosis is antrochoanal polyp. Antrochoanal polyps have low signal intensity on T_1 sequences and high signal intensity on T_2 sequences, peripheral enhancement in post-contrast series as distinct from NCMHs.[23] Paranasal CT and MRI are two complementary radiologic modalities that should be undertaken before surgery. They can reveal the extent of tumor, site of origin, relationship with critical structures and help surgical planning. In most reported cases heterogeneous soft-tissue masses with predominantly solid and cystic components can be identified on CT scans.[10,18,21] The tumors are reported to be heterogeneous with low signal intensity on T_1-weighted images and have high signal intensity on T_2-weighted images with significantly heterogeneous contrast enhancement.[10,18,21] Our patient had similar radiologic findings.

Involvement of paranasal sinuses is mostly seen in the ethmoidal sinus, but may include the maxillary and sphenoid sinuses. Cases limited to the nasal cavity are more infrequent.[4,9] Histopathologic diagnosis is mandatory.

As mentioned earlier, our patient had a history of rhabdomyosarcoma of the head and neck, which was localized in the right submandibular region. Schultz et al.[4] reported a patient with a combination of nasal chondromesenchimal hamartoma and botryoid embryonal rhabdomyosarcoma of the cervix. Unlike the former case, our patient had a different site of rhabdomyosarcoma (head and neck) and had no malignancy such as a Sertoli-Leydig cell tumor, thyroid carcinoma or pleuropulmonary blastoma.

Malignant transformation of tumor is significantly unusual. To date, we found only one reported case of NCMH becoming malignant.[24] Recurrence is not common but possible if the tumor has not been completely removed; only three cases of recurrence have been reported in the literature.[1,2,8] Thus, total endoscopic resection should be the main treatment in cases of NCMH. No adjuvant radiotherapy or chemotherapy is necessary if the tumor is completely resected.

In conclusion, NCMH is a rare surgically-treatable benign tumor that can be synchronously diagnosed with pleuropulmonary blastoma and should be kept in mind as a possible differential diagnosis for unilateral pediatric nasal masses.

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