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CASE REPORT

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Chondromesenchymal Hamartoma of the Anterior Cells of the Ethmoidal Labyrinth

ABSTRACT

Nasal chondromesenchymal hamartoma is a benign tumor of the sinonasal tract that arises in young and older children. Here, we report a rare clinical case of nasal chondromesenchymal hamartoma in a 15-year-old patient. After performing the necessary diagnostic tests, surgical treatment was recommended for the patient. In the course of surgery, a mass in the area of the anterior cells of the labyrinth was removed. According to the histological examination of the operative material, the mass had the structure of a chondromesenchymal hamartoma.

Keywords: Chondromesenchymal hamartoma, nasal tumor, mutation of DICER1 gene, differential diagnosis

INTRODUCTION

Nasal chondromesenchymal hamartoma (NCMH) is a rare benign tumor that arises more frequently in young and older children.[1,2] The etiology of hamartoma is still not fully established.[3] One of the probable causes of its development is a mutation of DICER1 gene, which is also known as a causative agent of pleuropulmonary blastoma, Sertoli Leydig cell tumor, and cystic nephroma.[4] Only approximately 57 clinical cases of NCMH have been reported in foreign literature.[5]

Most often, the neoplasm is localized in the area of the nasal septum, nasal vestibule, and ethmoidal cells and less often in the maxillary and sphenoid sinuses.[6]

The main clinical symptoms are nasal breathing difficulties, recurrent nasal bleeding, and nasal discharge.[3,6,7] Intracranial dissemination of the nasal hamartoma leads to headache; intraorbital dissemination causes visual disturbances, ptosis, and ophthalmoplegia.[3]

Diagnostics of nasal hamartoma include methods such as the anterior rhinoscopy, endoscopic examination of the nasal cavity, computer tomography (CT) of the nasal cavity and paranasal sinuses, and histological examination of the operative material.

Treatment tactics for nasal hamartoma consist of the endoscopic removal of the mass. The expediency of radiation therapy has not been revealed thus far.

CASE PRESENTATION

Patient L, 15 years old, came to the clinic with complaints of nasal breathing difficulties and recurrent nasal discharge.

The above-mentioned complaints had been bothering the patient for 2 years; conservative therapy with topical corticosteroids was carried out; however, the patient did not report any significant improvement.

An endoscopic examination showed a right-sided polypoid mass in the middle nasal passage that had an extension into the nasopharynx.

A CT scan of the nasal cavity and paranasal sinuses revealed a round-shaped haziness of the cells on the right side of the ethmoidal labyrinth with a heterogeneous content (focal ossifications and calcifications) spreading into the nasopharynx. No bone-destructive changes were detected. The remaining paranasal sinuses were pneumatized (Figure 1). The patient had no pulmonary lesions at diagnosis and during follow-up.

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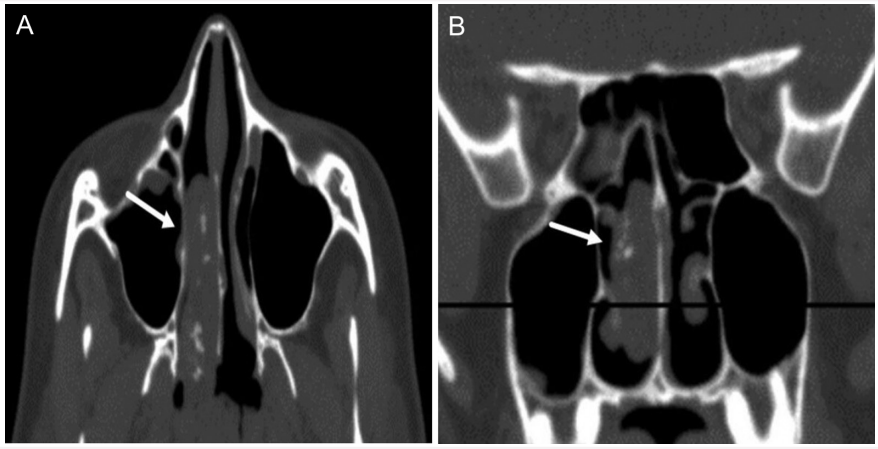

Figure 1. A, B. Computed tomography of the nose and paranasal sinuses in axial (A) and coronal (B) projections. The arrow indicates the mass.

Surgical treatment was recommended for the patient. Using the endotracheal anesthesia, the right half of the nose was examined with a microscope, a mass in the middle and common nasal passages was observed, and the middle nasal concha was lateralized by the mass.

Using Blakesley nasal forceps, the anterior cells of the ethmoidal labyrinth were opened, and the mass was removed. The material was sent for histological examination (Figure 2).

The postoperative period passed without complications. According to the histological examination, large fragments of the loose fibrous connective tissue covered with the atrophic cylindrical epithelium were observed microscopically. In different fields of view, there were multiple cystic cavities lined by the atrophic epithelium and filled with basophilic mucoid masses. In separate fields of view, cylindrical epithelium with mucus formation was detected. There were foci of hyaline cartilage located close to the cysts and separately; besides, there were foci of bone tissue. The formation had the structure of a hamartoma.

One year after the surgery, the patient was invited for an examination. The patient reported no adverse symptoms. The endoscopic examination of the nasal cavity showed that the common nasal passage was freely passable, but a scar was observed in the area of the middle nasal concha in the place where the hamartoma had been located (Figure 3). Written informed consent

MAIN POINTS

- The main clinical symptoms are nasal breathing difficulties, recurrent nasal bleeding, and nasal discharge.
- Nasal hamartoma tends to be destructive to the surrounding structures.
- The distinctive features of a hamartoma are the absence of a capsule, the presence of ossification, and calcification zones on computed tomography.
- The method of treatment is the surgical removal of the tumor.

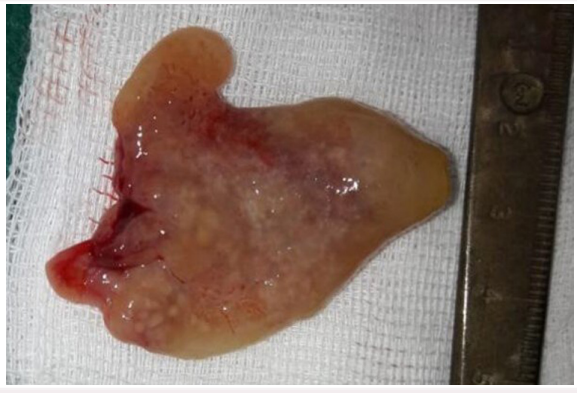

Figure 2. Macroscopic image of the chondromesenchymal hamartoma.

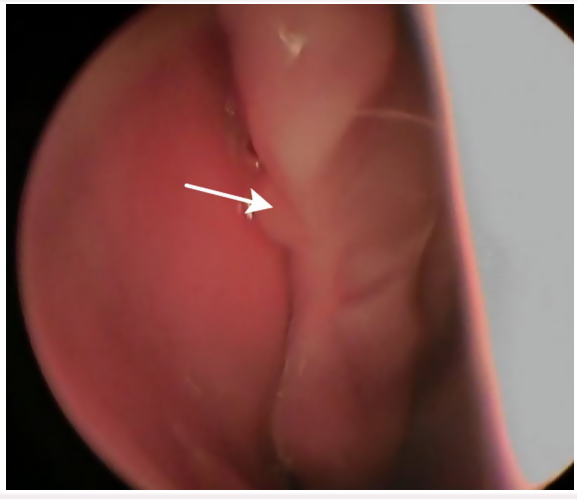

Figure 3. Endoscopic examination of the nasal cavity 1 year after surgery. The arrow indicates the scar in the area of the middle nasal concha.

was obtained from the patient to report his condition as a case report.

DISCUSSION

Chondromesenchymal nasal hamartoma was first described by McDermott et al[8] in 1998 as analogous to mesenchymal hamartoma of the thorax. In 4 out of 7 cases analyzed, the authors observed erosion of the surrounding bone tissue, including the cribriform plate, which was accompanied by neurological symptomatology.[8]

In addition to the chondromesenchymal nasal hamartoma, other types of hamartoma are reported in foreign literature: adenomatoid, lipomatous, and chondroid, depending on the nature of the mesenchymal component.[9]

In most cases, the nasal hamartoma is localized in the nasal septum, cells of the ethmoidal labyrinth spreading into the nasopharynx. Peric et al[10] described 2 clinical cases in adult men with an extremely rare location of the mass in the posterior part of the nasal septum and near the pharyngeal orifice of the auditory tube.

There is a possibility of malignant transformation of the mass in adults. In 2013, Li et al[11] reported that a 40-year-old female had a tumor recurrence 3 years after the resection of a hamartoma localized in the left nasal cavity spreading to the maxillary and the rectal sinuses. Histological examination of the operative material revealed cellular atypia and foci of necrosis.[11]

Hamartoma of the nose is a benign mass; however, it tends to be destructive to the surrounding structures. Differential diagnosis should be performed both with benign tumors such as hemangioma, inverted papilloma, angiofibroma, and nasal glioma and with malignant neoplasms such as esthesioneuroblastoma, rhabdomyosarcoma, and chondrosarcoma.[1,12,13]

The distinctive features of the chondromesenchymal hamartoma of the nose are the absence of a capsule, the presence of ossification, and calcification zones that are observable on a CT scan. In addition, the mass is poorly vascularized, with no hypervascular enhancement shown on magnetic resonance imaging, whereas inverted papilloma is observable on a CT scan of the nose and paranasal sinuses as a homogeneous haziness with an area of ossification, which indicates the location of the primary tumor invagination. Nasal hemangioma is a vascular neoplasm presented on a CT scan as a soft tissue structure, usually without destruction of the surrounding bone tissue. Angiofibroma has infiltrating growth and an indistinct surface, and it frequently originates from the roof of the nasopharynx or the pterygoid fossa. Nasal glioma is usually located subcutaneously in the bridge or the back of the nose and presents as a cosmetic defect.[14] In the nasal cavity, it appears as a polypoidal mass preferentially localized in the middle nasal cavity. A CT scan of the nasal glioma may reveal the nasal bone divergence and nasal septal deformity.[14]

Esthesioneuroblastoma is a homogeneous mass originating from the horizontal lamina of the ethmoid bone. Malignant neoplasms are more aggressive to the surrounding bone structures and are characterized by rapid growth.

Thus, a careful differential diagnosis is necessary, but only a histological examination of the surgical material can establish a definitive diagnosis.

This clinical case is of great interest due to the rare occurrence of the disease, as well as the difficulty of differential diagnosis of nasal cavity masses.

Informed Consent: Written informed consent was obtained from the patient to report his condition as a case report.

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