Nasal Chondroma Presenting as Hypertelorism

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ABSTRACT
Purpose. To report a rare case of nasal chondroma presenting as hypertelorism.
Case Report. We report a case of a 16-year-old boy with a large calcified mass arising from the posterior nasal cavity presenting as hypertelorism. Surgical excision was done, and the histopathological examination revealed a chondroma. The hypertelorism resolved postoperatively.
Conclusions. Nasal chondroma may also present innocuously as hypertelorism as in this case.

Hypertelorism is defined as an abnormally increased distance between two organs or parts. It is classified into ocular, orbital, and interorbital hypertelorism. Ocular hypertelorism is the increased space between the eyes as determined clinically by interpupillary measurement. Orbital hypertelorism signifies an increased distance between both medial sides and lateral sides of the orbits, whereas interorbital hypertelorism denotes increased distance only between the inner orbital walls. Ocular and orbital hypertelorism usually coexist. In a retrospective analysis of 90 patients with hypertelorism, the most common cause is a frontonasal malformation followed by craniofrontonasal dysplasia. Paramedian craniofacial clefts and sincipital encephalocele are the other but infrequent causes of hypertelorism. A miscellaneous category includes mostly chromosomal and syndromic disorders. It is possible, however, for the eyes to be displaced laterally by encroachment into the orbital space by tumor, neural tissue, or ethmoid air cells. We report a rare case of nasal chondroma presenting as hypertelorism.

CASE REPORT
A 16-year-old boy with no known premorbid was referred to our clinic with a complaint of increasing and noticeable distance between the two eyes for the past 4 years (Fig. 3A). It was first noticed by his parents and it has seemingly become less pleasing aesthetically. However, there was no diplopia or other symptoms. The visual acuity was 6/6 for both eyes with an interpupillary distance of 125 mm. The extraocular movements were full with no strabismus detected. There were no other significant ophthalmological or systemic findings. Computed tomography (CT) scan was ordered to identify the cause and extent of the hypertelorism. A contrast-assisted CT of the brain revealed a large calcified mass arising from the nasal cavity with minimal contrast enhancement (Fig. 1).

Resection of the mass was carried out under general anesthesia by the neurosurgical team (Fig. 2). An irregular mass of cartilaginous tissue measuring 6 cm and weighing 214 g was removed. Histopathological examination reported a lobulated tumor composed of stellate cells within lacunae, surrounded by abundant cartilaginous matrix. There were focal cellular areas with occasional binucleated forms. Some of the tumor lobules are rimmed by thinned-out bony trabeculae and foci of ossification. There were no cystic changes or abnormal mitotic activity seen, which was compatible with the diagnosis of a nasal chondroma.

Postoperatively, there was an immediate resolution in the hypertelorism with an interpupillary distance of 115 mm (Fig. 3B). Inner canthal distance also decreased by 6 mm from 46 to 40 mm. Inner canthal distance is a race- and age-dependent variable. For the Chinese race, as in the case of our patient, it has been reported as 37.2 mm on average in male subjects. Unfortunately, the patient developed alternating exotropia, which was corrected surgically 6 months later. The binocular stereoscopic vision was normal eventually. There has been no recurrence over the past 1 year.
DISCUSSION

Chondrogenic tumors include chondroma and chondrosarcoma. Chondromas are benign encapsulated cartilaginous tumors with lobular growing patterns. Based on location, chondromas can be divided into enchondromas or ecchondromas. Enchondromas are tumor growths within the bone whereas ecchondromas grow outward from the bone. It is crucial to differentiate between a chondroma and a chondrosarcoma in the histopathology examination, although it may be difficult. In a chondroma, chondrocytes represent normal cells and produce the cartilaginous matrix. However, the presence of hypercellularity, binucleation, or myxoid change may resemble a low-grade chondrosarcoma. There was no radiological evidence of invasion in our case. Moreover, histopathologically, our patient did not show any mitotic activity.

Chondromas are common in the long bones of the body but are rare occurrences in the head and neck region. These tumor regions are most often malignant. However, should it occur in the head and neck region, the most common site is in the midface. Murthy et al. reported the sites of predilection in the head and neck region, which include the ethmoidal sinus (50%), maxilla (18%), nasal septum (17%), hard palate, nasopharynx (including the sphenoidal sinus, 6% each), and alar cartilage (3%). It was postulated that these tumors probably arise from the remnants of the embryonal cartilaginous skeleton that escape from resorption during endochondral ossification.

Chondromas of the nasal region are very rare. It was first described by Morgan in 1842. Fu and Perzin reported a nasal chondroma series of seven patients, from the ages of 10 to 46 years, with an average of 26 years. There is no sex predilection. A series of 391 nose and paranasal sinus tumors reported by Ringertz found only two cases in which one arose from the posterior edge of the nasal septum and the other one, from the ethmoids and middle turbinate. It is indeed a very rare tumor. The most common symptoms of a nasal chondroma include nasal obstruction, headache, epistaxis, and, occasionally, an external nasal deformity. We believe that this is the first case of nasal chondroma presenting as hypertelorism without other suggestive symptoms. It can be postulated that this sole presentation may be attributed to the lateral growth of the mass, with little anterior and inferior projection into the nasal cavity, accounting for the paucity of nasal symptoms.

Diagnosis of orbital hypertelorism requires radiologic studies to confirm the presence of a wide separation of the orbits. Of the various methods, the preferable one is probably the interorbital measurement from posteroanterior skull radiographs. Values of 2 SDs or greater above the mean compared with Tessier data are...
diagnostic of orbital hypertelorism. As with other growth parameters, the interorbital measurement is age related, with about 16 mm at birth to 27 mm in adult life. The upper limit of normal in the adult is about 30 mm. Alternatively, several quantitative methods such as inner canthal measurement, interpupillary measurement, canthal index, and circumference interorbital index have been used in an attempt to establish the position of the orbits in relation to each other.

Radiographic evidence with high-resolution CT and magnetic resonance imaging are used as complementary investigations to precisely delineate the extent of the tumor and to define the involvement of the cribiform plate, anterior cranial fossa, and retromaxillary space. Surgical options for a nasal chondroma are varied and complex, with options including local excision, intranasal ethmoidectomy, or wide excision (usually via a lateral rhinotomy or maxillectomy approach). Most of the known cases have been successfully treated with a wide excision approach and no recurrence were noted thereafter.

In conclusion, despite its slowly progressive and aggressive local extension, nasal chondroma may also present innocuously simply as hypertelorism, as in our case.

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