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Nasal Chondrosarcoma in an Infant: Radiologic and Histologic Correlation

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AJNR Am J Neuroradiol 2006, 27 (7) 1394-1395 http://www.ajnr.org/content/27/7/1394 lent way for medical students, residents, and junior faculty to gain writing and publishing experience.² Just as Dr. Rich received a lesson preparing his manuscript, his letter to the editor is also a good lesson for me.

In their article published in 2002, Rich et al reported a new case of Hyrtl's fissure in a 5-year-old child with bacterial meningitis. Many CT scans and approximately 3 pages of historical inquiry were presented. The case we reported in our article was of a young girl who had been previously admitted to our department in August 1999 for a clear otorrhea without a history of meningitis. We presented CT and MR imaging figures.

Even though it was published and reported after the Rich et al article, it seems correct to consider our imaging case as the first chronologically documented and diagnosed case with CT and MR imaging that can be found in a search of PubMed. The Rich case was diagnosed in 2002, and the cases discussed in the Phelps book cannot be found in PubMed at all. Moreover, it is not clear why the keyword "Hyrtl" leads to a list in which our article and the Gacek article do not appear in PubMed. Finally, it is not unanimously acknowledged that Google can be considered a scientific tool for medical publication.

One of the originalities of the case we reported is MR imaging. In such rare and potentially hazardous pathology, CT scan associated with MR imaging is not an excessive imaging in the diagnosis process. It is still a challenge for perilabyrinthine fistulas to be diagnosed before the onset of bacterial meningitis.

The historical aspect of Rich et al is of great interest because they have followed with precision every track allowed them to reach the truth in the Hyrtl's fissure mystery. This was not our objective, and we trust the references of the renowned authors' articles we have read. As Rich et al specified it in their article, failure to find the truth does not prove it does not exist. The real origin of the first description of Hyrtl's fissure is still unknown, and it is possible that Joseph Hyrtl himself was the first to do it. I agree with this author that tympanomeningeal fissure should still be named Hyrtl's fissure.

The case we have presented is a real case of Hyrtl's fissure, a rare anomaly that should be known by physicians. The literature must lead to a better understanding of diagnosis pathway of new pathology, and the Gacek article, the Rich article, our article, and Dr. Rich's letter to the editor contribute to that understanding, which is the most important point.

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Nasal Chondrosarcoma in an Infant: Radiologic and Histologic Correlation

We report an unusual case of "nasal chondrosarcoma in an infant." An 11-month-old girl presenting with swelling near the inner canthus of her left eye she had experienced since 2 months of age underwent axial CT to assess the suspected orbital mass (Fig 1). CT showed a large heterogeneously enhancing low-attenuation mass centered in the left ethmoid sinuses with extension into left orbit and left maxillary sinuses (Fig 2). The tumor also eroded the cribriform plate and

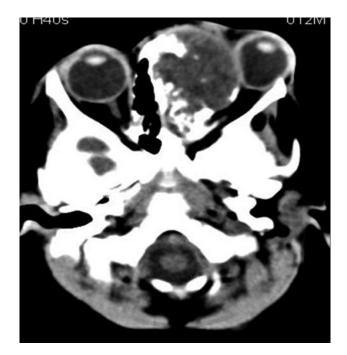


Fig 1. Plain axial CT scan shows a large expansile low-attenuation mass originating in left nasal cavity with extension into left orbit. There are subtle chondroid matrix mineralizations within the mass.

floor of anterior cranial fossa with intracranial extension (Fig 2). Only subtle calcific foci were noted within the mass (Fig 1). Biopsy of the mass showed histologic findings consistent with myxoid chondrosar-coma. Her parents, unfortunately, refused treatment.

Chondrosarcomas are malignant cartilagenous tumors that constitute approximately 10%–20% of all primary malignant osseous neoplasms, of which only about 10% arise in the head and neck region.¹ The highest incidence of craniofacial chondrosarcoma occurs in the 4th decade of life.

In the pediatric population, primary chondrosarcoma of head and neck is rare and usually occurs in the maxillary sinus or mandible. It is also typically low grade.² In rare cases, it may arise from the nasal cavity and nasal septum. There are only a few studies in the literature. In a study done by Gadwal et al, 14 such cases between 3 and 18 years of age were reported, only 2 of which originated from the nasal cavity.² The case discussed here is of nasal chondrosarcoma and is unusual

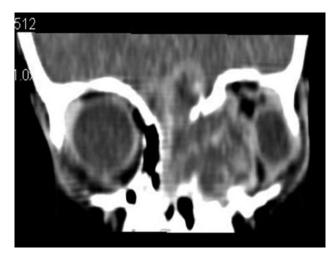


Fig 2. Postcontrast reformatted coronal CT shows septal and peripheral enhancement of the mass with extension into left maxillary sinus and anterior cranial fossa.

because it was seen in infancy and showed myxoid degeneration, which is more common in higher grades of chondrosarcoma.

Chondrosarcomas are classified pathologically as conventional, mesenchymal, clear cell, and dedifferentiated types. Myxoid degeneration is commonly found, particularly in higher-grade conventional chondrosarcomas, but it is not a well-established entity. The myxoid variety has a more aggressive clinical course than the low-grade conventional chondrosarcoma.

Imaging diagnosis principally depends on CT, which shows characteristic chondroid type of calcification. The typical appearance of mineralized chondroid matrix is a ring-and-arc pattern of calcification that may coalesce to form a flocculent, flecklike pattern. This characteristic chondroid calcification is the most useful and dominant feature of a cartilagenous lesion.³ Higher-grade chondrosarcomas and myxoid variety, because of their aggressive clinical behavior, however, frequently show an aggressive type of bone lysis and less extensive calcifications.⁴ Because of high water content, it also shows low attenuation on CT and high signal intensity on T2-weighted images. The enhancement is commonly mild and septal to peripheral.⁴ MR imaging can depict the exact extent of the tumor and has a significant role in monitoring recurrence. CT is optimal to detect the matrix mineralization, particularly when it is subtle or when the lesion is located in anatomically complex areas.³ Complete surgical resection is the treatment of choice. Even with extensive local invasion, the long-term prognosis is excellent for pediatric patients with primary chondrosarcoma of the head and neck.²

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