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## **Heterotopic pharyngeal brain.**

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# Heterotopic Pharyngeal Brain

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Heterotopic brain tissue within the pharynx is an extremely rare condition of infancy and childhood. The most frequent presentation is respiratory distress. There are 11 cases in the literature to date in which clinical and pathologic aspects are emphasized, all with a paucity of radiologic features [1]. We are reporting a twelfth case, histologically proven, which includes computed tomographic (CT) demonstration in axial, coronal, and sagittal planes, conventional radiography, and polytomography.

## Case Report

A female infant was delivered at term of a para 4 gravida 5 woman. Respiratory distress and cyanosis were present at birth. After intubation, a large cystic soft-tissue mass was noted in the nasopharynx. No other abnormalities were detected on physical examination. Neurologic examination was normal. Real-time cranial sonography revealed no intracranial abnormalities.

A lateral radiograph of the neck (fig. 1A) obtained after intubation demonstrated a large nasopharyngeal soft-tissue mass. Subsequent polytomography and cranial CT (figs. 1B and 1C) confirmed a homogeneous soft-tissue mass confined within the nasopharynx. There was no demonstrable calcification within the mass. No bone erosion or dehiscence was demonstrated. Sagittally reconstructed CT also demonstrated the position of the mass in relation to skull base and confirmed the absence of bony findings. Examination of the nasopharynx under general anesthesia revealed a smooth mass attached to the posterosuperior aspect of the nasopharynx.

Tracheostomy was performed followed by surgical excision, approached via splitting of the soft palate. A polypoid mass was noted to be attached to the nasopharyngeal wall, anterior to the sphenoid sinus. No intracranial communication was found. Microscopic examination of the 1.5 × 1.6 × 0.6 cm mass revealed neuroglial tissue separated by vascular septa of varying size. Removal of the mass from precervical fascia was achieved without difficulty. The patient made an uneventful recovery.

## Discussion

Heterotopic pharyngeal brain tissue is an extremely rare entity with 11 cases reported to date [1]. Respiratory distress, as was present in our case, is the most frequent presentation. This is usually present from birth and may be

life-threatening. Dysphagia may be another presenting feature [2–4]. The entity may also be found incidentally [5]. Associated congenital anomalies have been noted in five of the 11 described cases, including micrognathia, pectus excavatum, cleft palate, and unilateral choanal stenosis [3–6].

The underlying embryology of this entity is not well understood. Numerous theories have been postulated. Derivation from the anterior cerebral vesicle has been proposed [5].

Displacement of neural tissue may occur after the formation of Rathke pouch if the craniopharyngeal canal persists or a persistent communication may exist between the cranial end of the foregut (Seessel pouch) and the base of the occipital portion of the fetal head [7]. It is also suggested that this entity may derive from an encephalocele that has lost its central connection or may follow extracranial separation of embryonic neuroglia [8].

Two important entities to be differentiated are nasal gliomas and encephaloceles, both of which may have intracranial communication. Nasal gliomas are rare neurogenic tumors, which are of developmental rather than congenital origin. They may represent parts of the brain substance isolated from the brain by sutural closure during development or may be of either blastomatous origin or neural origin in the nasal mucosa surrounding the olfactory bulb [9]. They are mostly extranasal in location, but may also be intranasal or both intra- and extranasal. Nasal gliomas may contain astrocytes and gemistocytes, but do not contain choroid plexus or ependyma [1, 10]. They are benign and seldom recur after surgical excision. The nasopharyngeal mass in our patient arose from the nasopharyngeal wall, anterior to the sphenoid sinus. An extranasal nasal glioma could therefore not be excluded in our case. Encephaloceles represent herniation of intracranial contents through skull defects and may be difficult to distinguish from nasal gliomas, especially those of the intranasal type [10]. They contain brain tissue consisting of mixed elements varying with the degree of herniation.

Numerous other lesions in the nasopharynx must also be considered in the differential diagnosis. Teratomas are rare

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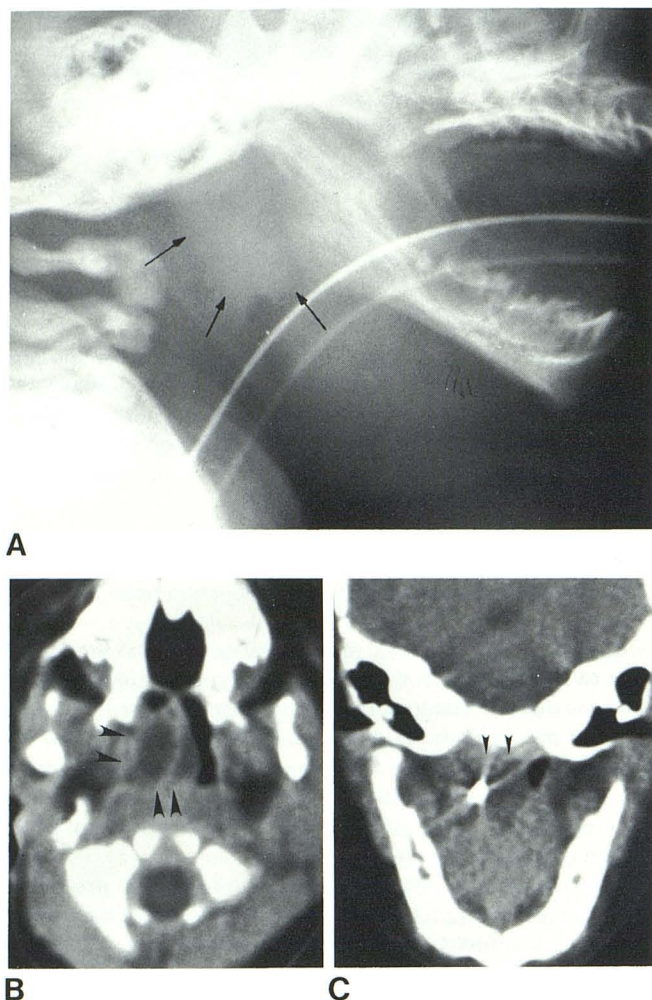


Fig. 1.—A, Lateral neck film. Large soft-tissue mass (arrows) in nasopharynx. B, Postcontrast axial CT scan shows nasopharyngeal mass (arrowheads) of mixed density mostly to right of midline. There are no distinct focal areas of contrast enhancement. C, Direct coronal CT scan reveals no evidence of bony defect adjacent to mass (arrowheads). A nasotracheal tube had been placed.

tumors of the nasopharynx, usually midline in location. There are four subtypes, of which dermoids are the most common [11]. The size and location of these tumors will determine the severity of symptoms. Respiratory distress may be present from birth. The presence of calcification will aid in the preoperative diagnosis. Neurofibroma, hemangioma, and lymphangioma may all present as nasopharyngeal masses. Malignant tumors of the nasopharynx are rare; the most common type is the sarcoma [12]. Rhabdomyosarcoma of the nasopharynx may initially appear as a benign rhabdomyoma [11]. Bone erosion or destruction in associ-

ation with the soft-tissue mass will lead to the diagnosis. Masses at the base of the tongue such as thyroglossal duct cyst or ectopic thyroid tissue can be excluded by virtue of their location.

Radiologic investigation of pharyngeal brain tissue should include conventional radiographs of the lateral neck and skull and CT of the head and neck. Radiographs of the lateral neck will usually demonstrate the soft-tissue mass. This was demonstrated well in our case (fig. 1A). A barium swallow may be indicated if dysphagia is a presenting symptom. CT of the head and neck will demonstrate the precise anatomic location of the mass and will also demonstrate the relative density of the mass. Calcification, hemorrhage, and fat will be readily appreciated. Administration of intravenous radiopaque contrast material will enable increased vascularity or tumor enhancement to be visualized. Attention to bone detail will demonstrate bone erosion or destruction. In our case no underlying bone abnormality was noted. At institutions where CT is not available, polytomography through the mass and skull base is recommended.

Surgical excision is the treatment of choice. Ten of the 11 previously described patients survived, with marked improvement of developmental status after relief of the respiratory problems in one patient [1].

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