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Parapharyngeal angiofibroma.

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Discussion

Primitive neuroectodermal tumor is a term used for morphologically similar tumors arising in both the central and peripheral nervous systems [3, 4]. PNETs occur preponderantly in children, and spread along cerebrospinal pathways is common. The most common PNET of the CNS occurs in the cerebellum (medulloblastoma) [4]. Extraneural metastases of medulloblastomas are well recognized [5]. Other than those arising in the cerebellum, primary PNETs in the CNS are infrequent and primary spinal intradural PNET is rare [1]. One such tumor with lung metastases has been reported [6]. In that case, metastatic spread was confined to the lungs; whether these were parenchymal metastases was not specified. In children with intracranial PNET, metastases to lung, lymph nodes, and liver have been reported [7].

Given the morphologic similarity of cerebellar PNET (medulloblastoma) and primary spinal PNET, it is not surprising that primary spinal PNET should give rise to similar radiologic metastatic manifestations. Sclerotic bone metastases, paravertebral soft-tissue mass, extrapleural mass, and pleural effusion have been reported with cerebellar PNET [8–12].

The prognosis for patients with metastatic CNS tumors is poor [2]. Although the tumor described in this report is rare, radiographic findings similar to this case are not unusual, and metastatic CNS tumors therefore should be included in the differential diagnosis.

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REFERENCES

1. Kepes JJ, Belton K, Roessman U, Ketcherside WJ. Primitive neuroectodermal tumors of the cauda equina with no detectable primary intracranial neoplasm: three case studies. *Clin Neuropathol* **1985**;4(1):1–11
2. Hoffman HJ, Duffner PK. Extraneural metastases of central nervous system tumors. *Cancer* **1985**;56:1778–1782
3. Rorke LB. The cerebellar medulloblastoma and its relationship to primitive neuroectodermal tumors. *J Neuropathol Exp Neurol* **1983**;42(1):1–15
4. Becker LE, Hinton D. Primitive neuroectodermal tumors of the central nervous system. *Hum Pathol* **1983**;14:538–550
5. Kleinman GM, Hochberg FH, Richardson EP. Systemic metastases from medulloblastoma: report of two cases and review of the literature. *Cancer* **1981**;48:2296–2309
6. Smith DR, Hardman JM, Earle KM. Metastasizing neuroectodermal tumors of the central nervous system. *J Neurosurg* **1969**;31:50–58
7. Duffner PK, Cohen ME. Extraneural metastases in childhood brain tumors. *Ann Neurol* **1981**;10:261–265
8. Parkinson D, Ross RT, Shields CB. Metastatic medulloblastoma. *Can J Neurol Sci* **1974**;1:253–254
9. Stolzenberg J, Fischer JJ, Kligerman MM. Extradural metastasis in medulloblastoma 10 years after treatment. Report of a case. *AJR* **1970**;108:71–74
10. Brutschin P, Culver GJ. Extracranial metastases from medulloblastoma. *Radiology* **1973**;107:359–362
11. Debnam JW, Staple TW. Osseous metastases from cerebellar medulloblastoma. *Radiology* **1973**;107:363–365
12. Placios E, Shannon M, Fine M. Unusual metastases from a medulloblastoma: case report. *Neuroradiology* **1979**;17(4):219–222

Parapharyngeal Angiofibroma

Juvenile angiofibromas are uncommon tumors that usually occur in adolescent males and account for approximately 0.5% of all neoplasms of the head and neck [1]. These tumors virtually always arise from the nasopharynx in the region of the sphenopalatine foramen and pterygopalatine fossa [1–6]. This report presents a unique case of an angiofibroma that involved the parapharyngeal space, without involvement of the sphenopalatine foramen or nasopharynx.

Case Report

A 25-year-old man presented with a parapharyngeal mass of 6-months duration. He had noticed an alteration in the quality of his voice, but he denied having pain, hemorrhage, or respiratory difficulty. Physical examination revealed a submucosal mass protruding into the left side of the oropharynx with displacement of the soft palate.

A CT scan showed an intensely contrast-enhancing mass in the left parapharyngeal space, with remodeling and anterior displacement of the left pterygoid plates (Figs. 1A and 1B). The pterygomaxillary fossa was normal. Angiography showed that the mass was hypervascular, with the primary blood supply from the internal maxillary artery (Fig. 1C).

After selective embolization of the internal maxillary artery, the mass was removed surgically. Pathologic examination revealed moderately large, stellate stromal cells distributed evenly within fibrous tissue and moderate-sized vascular channels, typical of a nasopharyngeal-type angiofibroma (Fig. 1D).

Discussion

Juvenile angiofibromas arise from the superolateral aspect of the nasopharynx near the sphenopalatine foramen. These lesions are usually histologically benign but tend to be locally invasive. Characteristically, expansion of the pterygomaxillary fossa occurs secondary to tumor growth [1–3].

Bryan et al. [2] have divided nasopharyngeal angiofibromas into three types according to which anatomic compartment the tumor occupies. Type 1 lesions extend medially from the sphenopalatine foramen and pterygomaxillary fossa to lie within the nasal cavity; type 2 lesions extend laterally into the infratemporal fossa; and type 3 tumors extend intracranially. Previous reports [1–7] have stressed that essentially all of these tumors have a component within the sphenopalatine foramen.

The present case is unusual because the location of the lesion in the parapharyngeal space was exclusive of involvement of the pterygomaxillary fossa. Also, the pterygoid plates were displaced anteriorly, unlike the usual posterior bowing seen with typical nasopharyngeal angiofibromas, which expand the pterygomaxillary fossa.

This case adds another lesion to the differential diagnosis of parapharyngeal masses. The more common masses include deep parotid tumors, schwannomas, glomus tumors, lymph nodes, metastases, and hemangiomas [1]. Although atypical in location, our case showed the characteristic CT and angiographic findings of a nasopharyngeal angiofibroma. In the parapharyngeal space, this appearance would be difficult to distinguish from a glomus tumor, hypervascular metastasis, or even a vascular tumor such as a hemangiopericytoma. However, the occurrence of a radiographically similar lesion in an adolescent male should raise the possibility of a juvenile nasopharyngeal angiofibroma.

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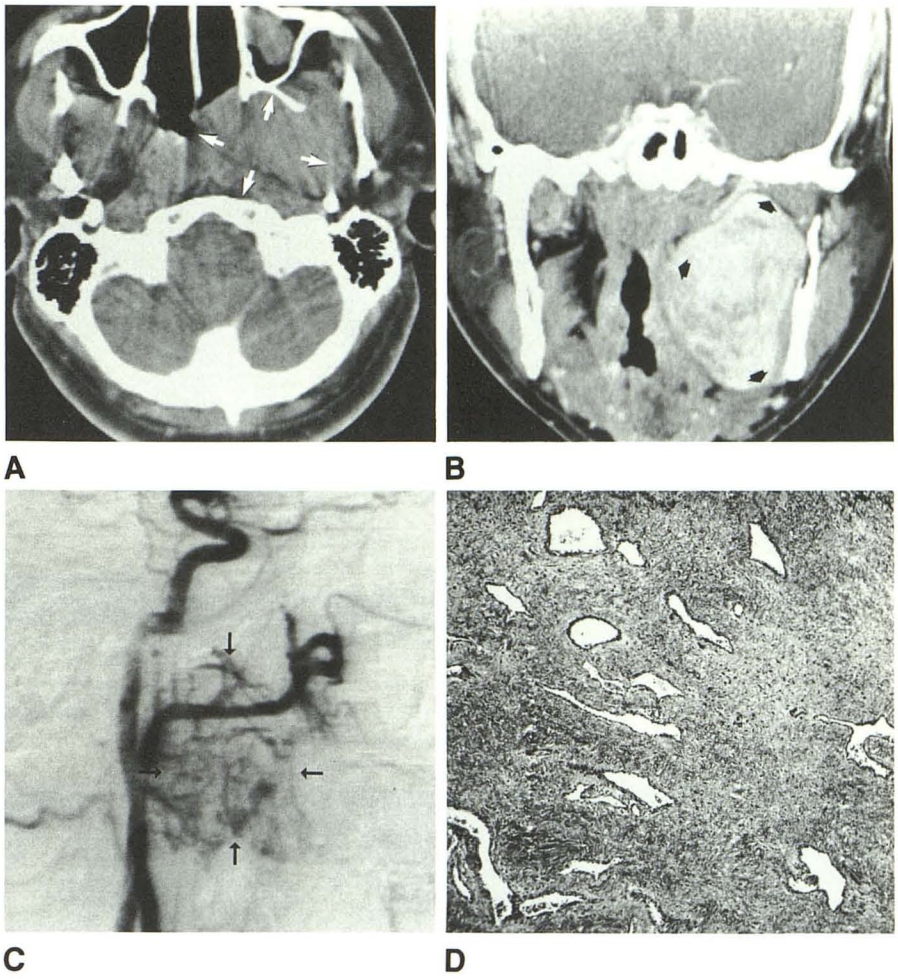

Fig. 1.—25-year-old man with parapharyngeal angiofibroma.

***A*, Unenhanced axial CT scan shows a mass (*arrows*) in parapharyngeal space, with anterior displacement and splaying of pterygoid plates.**

***B*, Enhanced coronal CT scan shows an enhancing left-sided parapharyngeal mass (*arrows*) displacing pharynx toward right.**

***C*, Angiogram of left common carotid artery shows a hypervascular mass (*arrows*) supplied by internal maxillary artery.**

***C*, Histologic section shows elongated vascular spaces and wavy collagen bundles. (H and E ×60)**

REFERENCES

1. Som PM. The paranasal sinuses and the parapharyngeal space. In: Bergeron RT, Osborn AG, Som PM, eds. *Head and neck imaging*. St. Louis: Mosby, **1984**:128–250
2. Bryan RN, Sessions RB, Horowitz BL. Radiographic management of juvenile angiofibromas. *AJNR* **1981**;2:157–166
3. Chandler JR, Moskowitz L, Goulding R, Quencer RM. Nasopharyngeal angiofibromas: staging and management. *Ann Otol Rhinol Laryngol* **1984**;93:322–329
4. Neel HB, Whicker JH, Devine KD, Weiland LH. Juvenile angiofibroma: review of 120 cases. *Am J Surg* **1973**;126:547–556
5. Heffner DK. Problems in pediatric otorhinolaryngic pathology. II. *Int J Pediatr Otorhinolaryngol* **1983**;5:125–138
6. Witt TR, Shah JP, Sternberg SS. Juvenile nasopharyngeal angiofibroma: a 30 year clinical review. *Am J Surg* **1983**;146:521–525
7. Ali S, Jones WI. Clinical records: extranasopharyngeal angiofibromas. *J Laryngol Otol* **1982**;96:539–565

Sonographic Demonstration of Cerebral Sinus Thrombosis

Thrombosis of the cerebral venous sinuses is a frequent complication in the sick newborn; it is detected in 3.7% of cerebral angiograms in infants and children [1]. Several conditions predispose infants to sinus thrombosis, including dehydration, infection, congenital heart disease, and blood dyscrasias [2, 3]. Many infants with these conditions are in the critical care facility of the hospital, where real-time sonography is the most convenient and least traumatic imaging method of screening for intracranial disease [4]. We present two cases of superior sagittal sinus thrombosis detected by sonography.

Case Reports

Case 1

A newborn boy (42 weeks gestation) was evaluated for hypotonia with marked respiratory distress after delivery with meconium-stained amnionic fluid. The infant was treated with a course of ampicillin and gentamicin for sepsis. A sonographic examination showed a focal echogenic, distended, and clearly marginated structure in the midline adjacent to the inner table of the skull that was thought to be thrombosis of the torcula (Fig. 1A). CT (Fig. 1B) and MR (Fig. 1C) confirmed the thrombus of the torcula and showed involvement of the transverse sinus as well.

Case 2

A baby girl (36 weeks gestation) was delivered by cesarean section because of fetal distress. She became apneic, mottled, and hypotensive at 6 hr of age. Blood cultures were positive for gram-negative β-hemolytic streptococci. A sonogram showed a distended superior sagittal sinus and torcula with faint but definite internal echos, thought to be thrombus (Fig. 2A). A CT scan (Fig. 2B) confirmed the thrombus and showed additional involvement of the straight sinus.