This 17-year-old African-American female presented to the Obstetrics Service for delivery of a full-term infant. She had full pre-natal care and a medical history significant only for sickle cell trait. At the time of presentation, the patient complained of headache and was found to be hypertensive with a blood pressure of 165/80. A caesarian section was performed under general anesthesia, secondary to fetal intolerance of labor. Twelve hours after the caesarian section, the patient began to complain of decreased vision, only able to see gray shadows. At this time, she was noted to have a low-grade fever of 100.5 and hypertension of 170/90, which was subsequently controlled. A full lab panel was performed, which was normal. All bacterial and viral cultures of her CSF and blood were negative. Over the next 36 hours, the patient’s condition declined to a comatose state and, at 48 hours, the patient expired.

Diagnosis: Posterior Reversible Encephalopathy Syndrome with Resultant Brainstem Infarction

Discussion:

The differential diagnosis for the findings in this case include brainstem infarct, posterior reversible encephalopathy syndrome (PRES), central pontine myelinolysis (CPM), and rhombencephalitis. Any of these entities can present with abnormal increased T2-signal in the brainstem. This patient had a low-grade fever of 100.5 and hypertension of 170/90, which was subsequently controlled. A full lab panel was performed, which was normal. All bacterial and viral cultures of her CSF and blood were negative. Over the next 36 hours, the patient’s condition declined to a comatose state and, at 48 hours, the patient expired.

PRES is a potentially fatal condition, which, if detected early and treated, can be completely reversible. Early detection and diagnosis by the radiologist are imperative in helping the clinicians guide management by correcting the underlying cause. This case is unusual with the essentially isolated brainstem involvement, but illustrates the need to seek out relevant clinical information when these abnormalities are seen in the posterior circulation.

References:

47-year-old male with a recent history of minor trauma to the head underwent a head CT for evaluation of subsequent dizziness and nausea. The patient, otherwise, is neurologically intact without localizing signs or symptoms.

Findings:
Pre-contrast T1-weighted axial and sagittal images (Figures 2a, 2b, 2c) show a predominantly hypointense well-delineated mass posterior to the clivus. There is mass effect on the adjacent pons. Foci of hyperintensity are noted within the mass on T1-weighted images (Figures 2a and 2c, arrow). T2-weighted imaging (Figure 2e) shows diffuse hyperintensity. No significant enhancement is seen on post-contrast T1-weighted imaging (Figure 2d).

CT of the skull base with thin collimation reveals numerous calcifications within the mass which demonstrate a ring- and-arc configuration (Figures 2f and 2g). Axial and sagittal images reveal that there is some continuity of the calcific foci with the posterior cortex of the clivus. Careful comparison with the

Discussion:
Represented on the given images is an unusual location of an intracranial chondroma. They have been reported arising from the dura mater, or choroid plexus, and these may be secondary to embryological remnants. They are usually midline in location. The mass image above is of midline, a more typical location for chordoid tumors. In addition, chordomas are usually locally invasive, have a relatively homogeneous hyperintense appearance on T2-weighted images, and show clear enhancement on post-contrast imaging. None of these features are present in this patient.

The main differential considerations, therefore, become chondroma and chondrosarcoma. Chondrosarcoma may have a CT and MR appearance quite similar to chordoma. Globular calcifications and ring-and-arc gadolinium enhancement may be seen in both of these chordoid tumors. With chondrosarcoma, however, there is usually marked enhancement. Enhancing fibrovascular septations have also been described in low-grade chondrosarcomas. In the current case, no significant enhancement is seen and, therefore, the diagnosis of chondroma is favored.

Chondromas are benign tumors that arise from cartilage. These may occur anywhere in the body, but an intracranial origin is a rare occurrence. Indeed, it accounts for less than 1% of all intracranial neoplasms. Among intracranial chordomas, most arise from the skull base. Most investigators believe they develop from embryonic cartilage rests along the embryonic skull. Either

References:
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Figure 2a. Axial SE (450/7) Figure 2b. Axial SE (450/7) Figure 2c. Mid sagittal SE (666/14) Figure 2d. Axial SE (416/8) Figure 2e. Axial SE (6000/98) Figure 2f. Mid sagittal SE (416/8)
Neuroradiology Case of the Day

Case 3:

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History:

An endoscopic transphenoidal biopsy of the tumor was obtained, histologically revealing fibrosarcoma. The patient underwent surgical resection through a skull base approach requiring removal of the soft tissue fibrosarcoma and the adjacent bony expansile mass of a fibrous dysplasia.

Discussion:

Fibrous dysplasia is a development disease of bone in which normal bone is replaced by abnormal fibrous tissue. This disease affects mainly the younger population (75% under the age of 30 years). The bony involvement may be monostotic or polyostotic. The craniofacial bones are the most common site of involvement. The most common clinical presentation of patients with cranial fibrous dysplasia is atypical facial pain or headache, followed by sinus congestion or infection that is often initially interpreted as sinusitis. Proposis, diplopia, and visual changes are the manifestations of orbital involvement. Some patients may also present with hearing loss or facial numbness. Treatment is aimed at correcting or preventing functional problems and achieving normal facial aesthetics. Total excision of the involved bone is the most successful form of treatment, but it produces the greatest functional and cosmetic deficits. A conservative therapeutic approach involving a modest reduction in the bulk of the lesions may be sufficient to relieve the signs and symptoms. The mere presence of cranial fibrous dysplasia is not an indication for treatment.

Fibrous dysplasia is a rare tumor, representing less than 5% of primary bone sarcomas. This tumor originates from mesenchymal cell and can occur as a soft tissue mass or as a bone tumor. It is rare to see fibrosarcoma in the sinonasal cavity. Primary and secondary forms of fibrosarcoma exist in bone. Primary fibrosarcoma arises from the bone. If the tumor arises from a pre-existing lesion or after radiation to an area of bone, it is called secondary fibrosarcoma. It has been reported that fibrosarcomas can arise from fibrous dysplasia, chronic osteomyelitis, bone infarcts, Paget disease, and in previously irradiated areas of bone. Complete surgical excision with wide margins is the treatment of choice. Prognosis is frequently very poor.

Accurate clinical history, such as pain and swelling, should alert the physician promptly and raise the suspicion of malignant transformation of fibrous dysplasia. The most reliable radiographic finding is the extension of the lesion through the bony cortex to the surrounding soft tissues. If the sarcomatous degeneration is suspected, computed tomography and especially MRI are the best noninvasive means for detecting the malignant transformation. Differential diagnoses of a mass primarily located within the sphenoid sinus vary. In this particular case, the identification of the associated bony abnormality was critical in the direction of the differential diagnosis. The identification of the thickened bone on MR with the associated soft tissue enhancing mass led to the performance of the computed tomographic scans allowing the diagnosis of fibrous dysplasia to be definitely made and in the direction of a malignant transformation to a fibrosarcoma. Otherwise, the differential diagnosis was unclear and included a host of other differential diagnostic considerations, including mucocle, primary sphenoid sinus squamous cell carcinoma, chondrosarcoma, chordoma, pituitary macroadenoma, and meningioma.

References:

The differential diagnosis for intramedullary spinal cord lesions includes astrocytoma, ependymoma, hemangioblastoma, and metastases. Of these tumors, astrocytomas and ependymomas make up 90%. Ependymomas and astrocytomas can be very difficult to differentiate on MR imaging alone. Ependymomas are the most common tumors in adults, comprising 60% of cord tumors. As in this case, they can show cord enlargement, edema, and often have cystic changes. An intensely enhancing nodule tends to be in the central part of the cord, as the tumor is believed to arise from ependymal cells lining the central canal. The enhancing nodule in this case is peripherally located in the posterior cord. Ependymomas often show hypointense rims on T2-weighted imaging, representing hemosiderin in areas of previous hemorrhage, a finding not seen in this patient. Astrocytomas do not commonly occur in the cervicothoracic spine. In distinction to ependymoma, they can be eccentric in location although they tend to involve the full diameter of the cord. The enhancing nodule seen on the above post-contrast T1 images is eccentric, but does not involve a majority of the cord diameter. Uneven enhancement may be seen with astrocytoma, compared with the intense focal enhancement demonstrated in this case. Intramedullary metastases are uncommon lesions, but are becoming more common as patients with cancer live longer. Spinal metastases are common in the cervical cord, have associated edema, enlarge the cord, and enhance. In this previously healthy young patient, there is no history of a primary tumor.

The clinical history and contrast-enhanced MRI of the spine, therefore, offer some clues which argue against ependymoma, astrocytoma, and metastases, although these remain viable diagnostic considerations. Hemangioblastoma remains a possibility as well. Differentiation among these four diagnoses is, thus, quite difficult. A dynamic phase MR angiogram is quite useful in further evaluation of this mass. The hypervascularity of this lesion is demonstrated by prompt enhancement on the early arterial phase images. A feeding vessel is identified arising from the left vertebral artery, as well as a draining vein which is seen on early phase images and persists into late venous phase. While cystic formation, cord expansion, edema, and enhancement are nonspecific findings regarding the differential entities listed above, the presence of feeding arteries, draining veins, and arteriovenous shunting is highly suggestive of hemangioblastoma.

Hemangioblastoma is a rare benign tumor of the CNS. They make up 5-10% of spinal cord neoplasms and are the third-most common intramedullary spinal cord tumor after ependymoma and astrocytoma. Hemangioblastomas involving the spinal cord occur either as solitary lesions or as part of the von Hippel-Lindau complex. This entity is inherited in autosomal dominant fashion, with patients showing manifestations such as retinal hemangioblastomas, renal cysts, pancreatic and liver cysts, islet cell tumors, and endolympathic sac tumors. The hallmark of the syndrome is multiple CNS hemangioblastomas. 10-30% of patients with hemangioblastomas in the spinal cord will have VHL syndrome. In this particular case, the patient had no other sequela of the von Hippel-Lindau complex. Most spinal cord hemangioblastomas occur in the thoracic spine (50%) or the cervical spine (40%). Imaging studies reveal hypervascularity, and syrinx or tumor cyst is present in 50-80% of cases. Cysts can be of variable signal intensity, depending on protein content. Most appear as an enhancing tumor nodule within a cyst or syrinx. A unique feature of these tumors is extensive and unusual spinal cord enlargement unrelated to cyst or syrinx formation. It is hypothesized that this may be secondary to venous congestion and edema. On MRI, one may find diffuse expansion of the cord. Isointensity is often seen on T1 images, but T2 imaging reveals increased signal intensity with prominent flow voids. A prominent cyst is usually seen. Post-contrast T1 images demonstrate bright enhancement of the tumor nodule. Spinal angiography shows a hypervascular mass with feeding arteries and draining veins, as well as AV shunting. Less frequently, serpiginous flow voids are seen with MRI, representing feeding arteries and draining veins noted on spinal angiography. Such draining veins are considered characteristic.

References:
9-year-old female presented with long-standing seizures refractory to treatment.

**Diagnosis:** Dysembryoplastic Neuroepithelial Tumor (DNET)

**Findings:**
Sagittal and axial T1-weighted images demonstrate a well-defined hypointense mass involving the cortex of the right temporal lobe (Figures 5a-5b). The mass has a gyriiform shape involving the superficial and lateral cortex of the entire temporal lobe. On the coronal T2-weighted images, the mass is hyperintense (Figures 5c-5d). The mass shows no enhancement on contrast studies (Figures 5e-5f). Minimal edematous change is seen.

The patient underwent resection of the mass, and histological examination revealed findings consistent with DNET. The patient did well postoperatively with resolution of the seizures.

**Discussion:**
Dysembryoplastic neuroepithelial tumor (DNET) is a benign intracranial neoplasm commonly affecting the supratentorial structures. Typical clinical manifestations are long-standing, intractable, partial-complex seizures affecting children. Mean age of onset of symptoms is 9 (range 1-19 years). Recognition of this tumor by MR studies provides the best chance at adequate treatment requiring complete resection of the mass.

Imaging studies reveal a mass which is well-demarcated with mixed cystic to solid components and which is cortically based. The tumor involves the temporal lobe in 50% to 62% of cases and the frontal lobe in 31% of cases. Involvement of adjacent subcortical white matter can also be seen. DNETs are hypointense on T1-weighted images and hyperintense on T2-weighted images. Contrast enhancement is seen in less than 30% of cases, and mass effect is variable. On CT, the mass is typically hypodense without contrast enhancement.

Histopathologically, DNET is a benign, predominantly intracortical lesion composed mainly of oligodendrocyte-like cells with admixtures of mature ganglion cells and astrocytes that are located in a myxoid or dense neurofibrillary matrix.

Differential diagnoses for intracranial processes manifested as medically refractory seizures in children include ganglioglioma, pilocytic astrocytoma, focal cortical dysplasia, pachygyria, and oligodendroglioma. Gangliogliomas are hypo- to isointense on T1-weighted images and hyperintense on T2-weighted images. Enhancement following gadolinium administration is highly variable, ranging from nonenhancing to ringlike to diffuse. A hypodense mass with frequent tumoral calcification is a common finding on CT. Even though gangliogliomas share many imaging features with DNET, their ringlike enhancement on postcontrast T1-weighted images and lack of a gyriiform appearance at the cortical margin distinguish them from DNET. MRI signals of pilocytic astrocytomas are similar to DNET except their propensity to markedly enhance with gadolinium. In addition, the appearance of cystic or cyst with a mural tumor nodule on imaging studies of pilocytic astrocytomas helps differentiate them from DNET. Focal pachygyria appears as a thick cerebral cortex with few, broad gyri and shallow sulci. However, the signal intensity is exact isointense on both the T1- and T2-weighted images to grey matter. Oligodendrogliomas typically have large nodular, clumpy calcifications (80%) and commonly enhance. In addition, oligodendrogliomas are usually located in the white matter, but may gradually extend to the periphery of the cortex.

References: