(SPS2_207) Dynamic contrast enhanced MRI of the temporomandibular joint in juvenile idiopathic arthritis

Start Time: 9:57 AM

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Abstract Details
Purpose:
We have shown that contrast enhanced MRI is useful in the evaluation of inflammatory arthritis: we first showed that, using a method of ratios of enhancement of the synovia to the longus capitus muscle, that contrast enhanced MRI can be used to establish normative values for synovial enhancement in asymptomatic joints. We then showed that this same ratio method can distinguish normal temporomandibular joints from inflamed joints in the setting of juvenile idiopathic arthritis. These prior studies, however, used measurements of enhancement at single time points following gadolinium administration, that leaves this approach open to error that may be introduced by variation in contrast administration techniques, such that a dynamic contrast enhanced study of TMJ synovial enhancement is needed. To date, dynamic contrast enhanced MRI (DCE-MRI) has been applied only for the evaluation of appendicular extremity joints, and thus far has demonstrated improved correlations with histopathology and treatment effects. To our knowledge, DCE-MRI has not been used to evaluate temporomandibular joint (TMJ) synovitis in the setting of juvenile idiopathic arthritis (JIA). Our purpose is to determine the dynamic enhancement curves of both normal and inflamed TMJ synovia that may provide a basis for the assessment of response of the inflamed TMJ to therapeutic interventions such as intraarticular steroid injections.

Methods & Materials: This study is a prospective IRB approved study performed on a 3T GE scanner utilizing dynamic contrast enhanced MRI through the TMJs in patients clinically characterized JIA and in normal controls. MRI scanning parameters include a single pre-contrast coronal T1 weighted sequence followed by a total of 10 consecutive post-contrast T1 weighted sequences each with a scan time of 1-minute for a total duration of 10 minutes. A region of interest (ROI) was placed in the synovium of the left and right TMJ with a reference ROI placed in the longus capitus muscle belly. Dynamic enhancement characteristics of the inflamed TMJs were determined and compared to clinical exam findings of synovitis.

Results: The synovia in the JIA patients demonstrated an initial peak enhancement at 5 minutes after contrast administration followed by a second peak at 10 minutes and showed twice the intensity of enhancement compared to normal controls. In contrast, the normal synovia demonstrated peak
enhancement at approximately 3-4 minutes after contrast administration and slowly decreased in enhancement thereafter.

Discussion: This study demonstrates proof of concept and the utility of dynamic post-contrast enhanced images of the TMJs in patients with synovitis. This method demonstrates peak enhancement of inflamed synovia and maximal difference between inflamed and non-inflamed synovia at 5 minutes post-injection, and may be used to evaluate treatment-related response in patients with JIA.
Purpose

Zygomatic complex fractures (ZCF) have been well described. A subset of patients, however, with ZCFs go on to develop trismus and limitation of jaw opening. The purpose of our study is to review retrospectively the CT imaging findings in patients with ZCF fractures and trismus and to correlate the imaging and clinical findings in this group compared to a control group of patients with ZCF fractures and no trismus.

Results

The clinical and CT imaging findings of 30 patients, 22 male and 8 female, (median age, 37 years old; age range, 14-85 years old), with zygomatic complex fractures, were retrospectively reviewed. Patients with mandibular fractures or h/o prior facial surgery were excluded. Approval from the institutional review board was obtained for chart and scan review and informed consent was waived for this HIPAA compliant study. History and physical exam records and CT scans were available in all 30 patients. The records were reviewed for the subjective complaint of trismus defined as pain on opening of the mouth from repose, the maximal jaw opening (maximal incisal opening or MIO), and for the mechanism of injury. The CTs were reviewed for fractures in the maxillae, zygoma, sphenoid, and temporal bones, for retroposition of the fractured zygomatic body, and for the distance between the coronoid process of the mandible and the fractured zygomatic body, zygomatic arch, and the posterolateral wall of the maxilla. The results of the trismus and nontrismus groups were then compared by standard T-test and a p-value was calculated.

Results

16 of the 30 patients reported trismus following ZCF. The MIO in the trismus group was reduced at 20-30 mm. The mechanisms of injury included assault or sports-related, fall, and MVA and did not differ significantly between the two groups. The zygomae and maxillae were fractured in all 30 patients; the sphenoid bone in 18 (12 trismus, 6 non trismus), and the temporal bone in 20 patients (13 trismus and 7 non trismus). There were statistically significant differences between the trismus and nontrismus groups in the relative retroposition of the fractured zygomatic body compared to the unfractured side, and the relation of the coronoid process of the mandible to the fractured zygomatic body.
CONCLUSION
In our study group, patients with ZCF fractures who develop trismus had higher rates of fracture of the sphenoid and temporal bones and there were statistically significant differences between the two groups in the retroposition of the fractured zygomatic body and in the distance between the coronoid process of the mandible and the fractured zygomatic body. These findings may play a role in surgical follow up and management of patients with ZCFs.
Abstract Details

Purpose: Slit lamp ophthalmologic examination and ocular B-scan sonography examinations of the globe are frequently constrained by technical limitations in the setting of traumatic orbital injury. The latter may put the globe at risk by inducing pressure onto a ruptured globe. The main purpose of this study was to evaluate the diagnostic performance of CT in acute anterior segment ocular injuries as an adjunctive diagnostic modality. Methods: Following IRB approval, we retrospectively identified 85 patients who presented to our ED (from January 2014 to April 2016) with recent direct trauma to the anterior segment of the eye. De-identified multiplanar thin-slice CT images were reviewed by two subspecialty board-certified neuroradiologists for presence of anterior segment rupture, hyphema, as well as lens, ciliary body, and lacrimal gland injury. The CT findings were compared to slit lamp, B-scan sonography, and/or operative data as the criterion standard. Results: The neuroradiologists' CT evaluation demonstrated high sensitivity (92.3%, CI: 74.9-99.1%) and specificity (96.6%, CI: 88.3-99.6%) in diagnosing anterior segment rupture. Detection of lens dislocation and hyphema showed a
sensitivity/specificity of 86.7%/91% and 75%/83.6%, respectively. Although the experienced neuroradiologists were able to evaluate involvement of the lacrimal apparatus on CT with high specificity (97.5%), their sensitivity was unexpectedly low (50%). However, a shallow anterior chamber was detectable with a sensitivity/specificity of 93.8%/88.4% respectively. This critically important sign when confirmed to be true, predicted anterior globe rupture in 19 out of 26 patients (OR = 38, P < 0.0001). Conclusion: Subtle ocular findings on CT can provide valuable and accurate information to the ophthalmologist concerning acute trauma to the ocular anterior segment.
Abstract Details

Purpose: To evaluate the rarely occurring tumor, orbital paraganglioma, with dynamic contrast-enhanced magnetic resonance imaging (DCE MRI) using a time-signal intensity curve (TIC) and positive enhancement integral (PEI) images.

Description: Paragangliomas are tumors of the paraganglia that arise from neural crest progenitor cells, which are distributed all over the body. Paragangliomas may be adrenal or extra-adrenal. Extra-adrenal paragangliomas in the head and neck are not common and may be located at the common carotid artery bifurcation, the jugular foramen, along the vagus nerve, and within the middle ear.

The orbit is an extremely unusual site for paragangliomas, and the existence of normal paraganglia in the orbit is not well documented in humans. Some authors suggested that orbital paragangliomas may arise from sustentacular cells or ciliary paraganglia. This tumor is hypervascular and infiltrative in nature, often making surgery for it difficult. More common orbital tumors that may mimic paragangliomas in imaging are meningioma, cavernous hemangioma, and schwannoma.

DCE MRI is a noninvasive imaging technique that can be used to derive quantitative and semiquantitative parameters that reflect the microcirculatory structure and function in imaged tissues. Researchers have investigated this technique in a wide range of oncologic applications, including for head and neck tumors. However, given the rare occurrence of paraganglioma in
the orbit, diagnosis of it remains challenging.

We present herein a case of pathologically proven orbital paraganglioma. The patient presented with a longstanding complaint of left orbital proptosis that progressed over the previous 1 year. MRI revealed a 33 x 16-mm ovoid mass in the left superior lateral orbit with associated bone remodeling of the lateral orbital wall. The mass was mildly heterogeneous in signal on T2-weighted images, with prominent vascular flow voids within the tumor. The mass extended into the periorbital soft tissues and superior eyelid as well as along the subcutaneous soft tissues overlying the zygomatic arch.

DCE MRI demonstrated a hypervascular mass with early, rapid enhancement after gadolinium administration very similar to arterial vascular enhancement. A TIC demonstrated a rapid initial upslope and rapid washout pattern. The semiquantitative parameter based on the TIC revealed high peak enhancement, a high maximum signal enhancement ratio, and a short time to maximum enhancement. These findings were more distinctive for orbital paraganglioma than for other more common hypervascular tumors, such as meningiomas. Using postprocessed PEI images generated from the area under the TIC was very helpful in delineating the tumor margin, as it infiltrated the periorbital soft tissue, eyelid, and subcutaneous soft tissue overlying the zygomatic arch. Postprocessing of DCE MRI was simple and practical in clinical setting.

Summary: Orbital paraganglioma has distinctive DCE MRI characteristics. Using DCE MRI as an adjunct to conventional MRI to assist diagnosis and delineation of tumor margin for orbital paraganglioma is promising. Simple assessment of the TIC, semiquantitative parameters, and postprocessed PEI images should be considered in evaluation of orbital masses found on MRI scans.
(O&V_11) Compressive Optic Neuropathy from the Normal and Abnormal Internal Carotid Artery

Start Time: 10:25 AM

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Abstract Details
The causes of optic neuropathy are myriad, and include demyelinating, inflammatory, ischemic, traumatic, and compressive etiologies. Only 20% of cases will identify a compressive etiology upon the optic nerve as the source of the optic neuropathy. Mass lesions along the course of the optic nerve, including but not limited to meningiomas, hemangiomas, hemangiomas, lymphangiomas, lymphoma, and extraocular muscle enlargement from thyroid eye disease, all have been known to compress the optic nerve, leading to damage to the optic nerve, disc pallor, and subsequent loss of vision.

The internal carotid artery (ICA) is another potential mass lesion that has been rarely described in causing a compressive neuropathy. Neurovascular conflict from the 5th, 6th, and 7th cranial nerves have been well described, but a vascular conflict to the 2nd cranial nerve is less often seen and not often thought of by the radiologist as the source for optic neuropathy.

Aneurysms from the ICA, whether they be fusiform or saccular, can compress the optic nerve. Additionally, a normal appearing nonaneurysmal ICA can compress the optic nerve, leading to optic neuropathy.

In our presentation, we will show several examples (at least 5 cases) of abnormal and normal appearing ICAs that are believed to be the cause of the optic neuropathy. Each of the cases clearly show optic atrophy, the normal or abnormal ICA compressing the optic nerve, and have concordant ophthalmologic findings that suggest the ICA to be the source optic neuropathy. The clinical outcomes of some of these cases will be examined, if and when possible.
Optic nerve magnetic resonance imaging characteristics in OPA1 related and WFS1 related optic neuropathy

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Abstract Details
Purpose: Dominant optic atrophy (DOA) and Wolfram syndrome (WFS) are inherited optic neuropathies caused by mutations in the OPA1 and WFS1 genes, respectively, and are characterized by slowly progressive, bilateral visual loss. Few studies have examined the MRI features of the optic nerves in these patients. Our purpose is to determine the MRI characteristics of OPA1 and WFS1 related optic neuropathy and to correlate clinical with MRI findings.

Methods: Using an updated retrospective database of 111 patients with bilateral optic atrophy referred for genetic testing, we screened for OPA1- and WFS1-positive patients who had an MRI as part of their work-up that was available for analysis. The signal and caliber of the optic nerves were measured on coronal STIR sequences. The signal was recorded both as a raw score and as a ratio normalized to the signal within CSF, vitreous, corpus callous, and longus capitus. A sample of 6 subjects without ocular or central nervous system disease served as the control group. Clinical features of disease severity among OPA1 patients were analyzed as a function of normalized T2-weighted signal and optic atrophy.

Results: MRIs were available for 8 patients with OPA1 mutations and 2 patients with WFS1 mutations. Raw T2-weighted signals in the optic nerves were not different between control subjects and OPA1 or WFS1 patients. Normalized T2-weighted signal ratios, however, in the optic nerves of OPA1- and WFS1-positive patients were significantly increased (~2-fold on average) above that of control subjects. Optic nerve size was also reduced in OPA1 patients. Among OPA1 patients, normalized T2-weighted signal, but not optic nerve size, significantly correlated with clinical measures of disease severity including visual acuity impairment, cup-to-disc ratio and scotoma density. This method improved the sensitivity of MRI for optic neuropathy for the OPA1 patients: the subjective prior interpretation of the MRIs identified abnormal T2-weighted signal in only 3/8 of the OPA1 patients, and atrophy in only 5/8 of the OPA1 patients.

Conclusion: We have established a clinically feasible method for measuring T2-weighted signal in the optic nerves that corresponds with clinical severity in genetic OPA1 and WFS1 optic neuropathies and that may increase the sensitivity of MRI for the detection of optic neuropathy.
White Matter, a Good Reference for Signal Intensity Evaluation in MRI for the Diagnosis of Uveal Melanoma

Abstract Details

White Matter, a Good Reference for Signal Intensity Evaluation in MRI for the Diagnosis of Uveal Melanoma

Purpose: To determine the accuracy of Magnetic Resonance Imaging (MRI) in the diagnosis of uveal melanoma using normal white matter as a reference tissue for the signal intensity evaluation on T1w images and vitreous body on T2w compared with the conventional method of using the vitreous body as a reference on both T1w and T2w.

Materials and Methods: This retrospective study was approved by the institutional review board. The medical records and MRIs of 36 patients who underwent MRI, between August 2006 and December 2015, due to clinically suspicious ocular masses were blindly reviewed by two neuroradiologists. Seventeen patients had histopathologically proven diagnoses (11 melanomas, 3 metastases, 2 retinoblastomas, 1 medulloepithelioma) and 19 patients had clinically presumed diagnoses (1 melanoma, 5 metastases, 13 benign lesions such as retinal/choroidal detachment, hemorrhage). For all clinically presumed benign lesions a 2-year follow-up was required in order to confirm their benignity.

By using white matter as a reference for the signal intensity evaluation on T1w images and the
vitreous body as a reference on T2w images, uveal melanomas were suggested by hyper-intense signal intensity on T1w and hypo-signal on T2w with homogeneous enhancement. The accuracy of the diagnosis of uveal melanoma using white matter as a reference on T1w was compared with the conventional method of using the vitreous body as a reference on both T1w and T2w images. 

Results:
The diagnosis of uveal melanoma using white matter as a reference gave a sensitivity of 91.67% (95%CI=82.64-100.7), specificity of 100.0% (95%CI=100.0-100.0), PPV=100.0% (95% CI=100.0-100.0), and NPV=96.0% (95%CI=89.6-102.4).
By using the vitreous body as a reference, sensitivity as high as 100.0% (95%CI=100.0-100.0) was obtained, but with a low specificity of 58.33% (95%CI=42.23-74.44), PPV=54.55% (95% CI=33.28-70.81), and NPV=100.0% (95%CI=100.0-100.0).
Inter-observer agreement was almost perfect between both radiologists, Kappa= 0.835 (95%CI=0.707-0.874, P value < 0.001).

Conclusions:
The presence of hyper-intense signal intensity on T1w compared with normal white matter, hypo-signal on T2w compared with the vitreous body and homogeneous enhancement appear to be a highly accurate method for the diagnosis of uveal melanoma.

Keywords: Uveal melanoma, Magnetic Resonance Imaging (MRI), white matter, reference
Figure 1: MR imaging appearance of melanoma in a 67-year-old woman with blurred vision in the left eye for 10 days. (a) Axial T1-weighted spin-echo MR image (TR/TE=400/10) shows the ocular mass (*) with hyper-intense signal compared with normal white matter (arrow). (b) Axial T1-weighted spin-echo MR image (TR/TE=5000/90) shows hypo-intense mass (1) compared with the vitreous body (arrow). (c) Contrast-enhanced T1-weighted image (TR/TE=400/12) shows homogeneous enhancement (arrow).

Figure 2: MR imaging of a 36-year-old man with known adenocarcinoma of the lung who developed blurred vision in his left eye. (a) Axial T1-weighted spin-echo MR image (TR/TE=471/12) shows hypo-intense broad based mass (b) Axial T2-weighted spin-echo MR image (TR/TE=3000/90) shows hypo-intense mass (c) Axial contrast-enhanced T1-weighted spin-echo MR image (TR/TE=551/12) shows enhancement of intraocular mass and adjacent retrobulbar fat as evidence of extraocular invasion (arrow). Small metastasis with surrounding edema in right temporal lobe is noted (arrowhead).

Figure 3: MR images of melanoma in a 61-year-old woman with blurred vision in the left eye for 1 year. (a) Axial T1-weighted spin-echo MR image (TR/TE=400/9) revealed iso to slightly hypo-intense signal intraocular mass (arrow). (b) Fat suppressed T1-weighted spin-echo image (TR/TE=400/12). (c) Axial T2-weighted gradient-echo MR image (TR/TE=611/11, FA=18°) shows evidence of remote internal hemorrhage seen as markedly hypo-intense signal. (d) Post contrast.
Abstract Details
Minimizing ionizing radiation dose in pediatric patients is fundamental to the practice of pediatric radiology. For the evaluation of craniosynostosis, the current most widely accepted imaging examination is computed tomography of the head, an examination which involves ionizing radiation. An alternative screening exam is ultrasound examination of the cranial sutures.

A retrospective review of all cranial suture ultrasound examinations at our facility, over the course of a three-year period, was performed. Results from these studies were compared to Head CT examinations and/or clinical follow-up in order to evaluate the accuracy of cranial suture ultrasound as a screening tool to rule in or rule out craniosynostosis.

Of the 60 studies that were performed, 53 were deemed to be adequate for inclusion (with criteria
being adequate characterization and documentation of all 6 major cranial sutures as well as access to clinic follow-up information or additional imaging). 46 of these examinations did not reveal findings consistent with craniosynostosis. In each of these 46 instances, follow up physical exam findings and/or CT imaging confirmed that there was no abnormal premature suture closure. In all 7 cases where ultrasound findings did demonstrate synostosis, there was correlation with comparison CT exam or operative reports that confirmed premature suture closure.

With these results, we feel that screening ultrasound offers a reliable alternative for initial evaluation of possible craniosynotosis.
(SPS2_202) Pediatric Craniocervical Metrics Revisited: Establishing Landmark Basion-Cartilaginous Dens Interval in Infants Using CT Sagittal Soft Tissue Measurements

Start Time: 10:53 AM

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Abstract Details
Purpose:
Craniocervical Junction (CCJ) distraction injuries are potentially devastating yet remain a diagnostic challenge, especially in the very young. The bony basion dens interval (BDI) is one of the key measurements for assessing atlanto-occipital distraction injuries on X-ray and bone targeted CT, the principle being that when the destabilized head is distracted from the spine, the interspace between the basion and odontoid tip may widen. However, the BDI does not reflect the true distance between the basion and dens tip when there is still an immature cartilaginous dens cap. In fact, the bony interspace may appear spuriously wide, particularly when the upper dens is entirely cartilaginous; additionally, the normal BDI range changes as the os terminale appears and is used for the measurement. In effort to address these limitations and offer a direct measurement of the true interspace applicable to the very young, we introduce the novel concept of measuring the pediatric “Basion Cartilaginous Dens Interval” (BCDI) using sagittal soft tissue CT reformat and will establish the upper limit normal BCDI among infants up to 24 months of age.
Materials & Methods: Midline sagittal soft tissue targeted reconstructions of the normal craniocervical junction (CCJ) were retrospectively analyzed in a total of 86 infants up to 24 months of age who underwent GE 64 multidetector head CT, first excluding patients with CCJ structural deformity, spinal injuries or motion degradation. There were 50 female and 36 male patients in the cohort. The shortest distance between the sagittal midline cartilaginous tip of the dens as viewed on soft tissue windows and the midline bony basion tip as demarcated on bone windows, were independently measured on iSite PACS by 2 separate readers. The inter-reader reliability was measured by both the Pearson correlation coefficient and the interclass correlation (ICC). Mean, median, standard deviation and range were calculated. The normal maximum value was defined as two standard deviations (SD) above the mean.

Results: Of the two readers, the Pearson correlation was 0.91 (95% CI: 0.87 – 0.94) and the ICC was 0.90 (95% CI: 0.85 – 0.93). The combined BCDI measurements for the 2 readers ranged from 0.4mm to 4.9mm with median of 2.15mm, mean of 2.36mm and SD of 1.02mm. The upper limit normal of study population is calculated to be 4.4mm.

Conclusion: The cartilaginous dens cap is visible on midsagittal soft tissue reformatted MDCT, allowing us to move beyond the sole reliance on bony landmarks when assessing basion dens interval in the setting of potential CCJ distraction injuries in the very young. We introduce the BCDI as a more direct measurement of the BDI in the immature spine, establishing the upper limit normal BCDI of 4.4mm for the 0-2 year age group. Analysis of BCDI in the immature spine may serve to complement traditional bony BDI and other craniometric relations presently used in assessing high risk babies with potential CCJ dissociative injuries.
The Opticocarotid Recess: A Critical But Frequently Missed Route of Intracranial Spread of Sinus Disease

Start Time: 11:01 AM

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Abstract Details
Purpose: We define the endoscopic and radiologic anatomy of the opticocarotid recesses through an illustrative case series identifying the vital anatomic landscape and careful preoperative assessment prior to surgical intervention.

Materials/Methods: A retrospective review of teaching file cases at a tertiary academic center was performed to identify intracranial opticocarotid spread and complications of sphenoid sinus disease.

Results: Five cases of intracranial spread of disease were identified from the medial and lateral opticocarotid recesses.

Conclusions: The medial and lateral opticocarotid recesses are frequently missed sites of sphenoid sinus disease leading to intracranial spread, with possible significant morbidity and mortality. This important anatomic region has not been previously described in the imaging literature, and it is vital that the head and neck imager be aware of this potential pitfall.

Figure Legend. Figure 1. Coronal bone algorithm CT demonstrates the opticocarotid recess (yellow arrow) interposed between the optic nerve medially and the carotid artery laterally.
References:


Fibrosarcoma masquerading as Gorham disease of the calvarium

Abstract Details
PURPOSE: Gorham disease, also known as Gorham-Stout disease or vanishing bone disease, is a rare condition characterized by progressive osteolysis due to replacement by uncontrolled proliferation of hemangiomatous or lymphangiomatous tissue. While it can occur in any bone, involvement of the skull or skull base is unusual. The radiographic differential diagnosis of Gorham disease includes a variety of benign and malignant processes, and tissue diagnosis is required for definitive diagnosis.

CASE REPORT: A 33 year-old woman developed left-sided head pain. Initial imaging demonstrated lytic bone destruction of the calvarium, and biopsy at that time was reportedly consistent with Gorham disease. Her symptoms and imaging slowly progressed over several years, despite treatment with a variety of medical therapies. She ultimately underwent left suboccipital craniotomy, and pathology revealed low-grade fibrosarcoma without evidence of vascular morphology. This was followed by radiation therapy for unresectable residual disease.

IMAGING FINDINGS: CT demonstrated regional lytic bone destruction with replacement by hyperattenuating soft tissue involving the left temporal and parietal bones, and left greater than right occipital bone with erosion of both inner and outer tables. At MRI this lesion demonstrated hyperintensity on T2-weighted images and homogenously enhancing infiltration of the calvarium on post-gadolinium T1-weighted images with subjacent dural thickening/enhancement. On MRV, the adjacent left transverse-sigmoid sinus junction was narrowed by mass effect or infiltration.

SUMMARY: Spindle cell sarcomas such as low-grade fibrosarcomas may convincingly mimic the radiographic features of Gorham disease. It is important to obtain adequate tissue when Gorham disease is being considered to ensure a definitive pathological diagnosis prior to treatment, in order to maximize the opportunity to offer curative therapy of alternative diagnoses.