Neuroimaging Highlight

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Pregnancy-induced Cystic Degeneration of Fibrous Dysplasia

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A 24-year-old pregnant woman in her third trimester presented with a recurrent visual field cut from sphenoidal fibrous dysplasia. Her disease had been stable for three years with improved vision after previous craniotomy for bilateral optic nerve decompression for a bitemporal homonymous hemianopsia (Figure 1), until the recurrent right visual field defect developed during her pregnancy (dense right superior temporal visual field defect). Computed tomogram (CT) obtained at routine follow-up one-year earlier showed dramatic cystic degeneration in her fibrous dysplasia (Figure 2). She underwent subsequent bifrontal craniotomy for repeat optic canal decompression shortly after she had delivered. The recurrent visual field defect resolved after surgery.

Acute cystic degeneration (ACD) of craniofacial fibrous dysplasia (CFD) is a rare phenomenon where an acute change of a previously stable CFD lesion causes new symptoms such as sudden dysmorphism, pain, or visual deficits because of acute lesion expansion[1,2]. Acute cystic degeneration has been documented in several case reports and a small series, but neuroimaging showing the progressive bony changes immediately before and after ACD is rarely presented[1]. The largest study on ACD in CFD consisted of nine patients[1], and seven of the nine patients carried a prior diagnosis of CFD at the time of ACD onset, which occurred at a mean of 2.8 years (1–7 years) after the initial CFD diagnosis. The patients ranged in age from 6 to 40 years, but the vast majority were between 10 and 22 years old[1]. This same study identified sudden mass enlargement (78%) and pain (67%) as the most common symptoms in ACD of CFD[1]; however, four other patients in their series presented with abnormal visual symptoms, including two patients with resultant optic nerve compression and blindness. The propensity for causing visual loss is important to note as another large series of CFD patients demonstrated ACD was the most common cause of visual loss[3].

Although several case reports have documented initial CFD presentation occurring during late pregnancy[4-6], ACD in CFD is exceptionally rare in pregnancy and has not been documented with neuroimaging. We report the first known case of ACD in stable CFD, documented by recent CT scan, occurring during a patient's pregnancy (Figure 2). Because of the paucity of literature on this subject, no significant predisposing factors contributing to ACD of CFD have been identified. Furthermore, no significant correlation between onset of ACD and the relation of a prior surgery to treat CFD has been demonstrated; however, in addition to our case, there has been another report of ACD in CFD occurring after a prior surgery[1]. The jaw is an area where

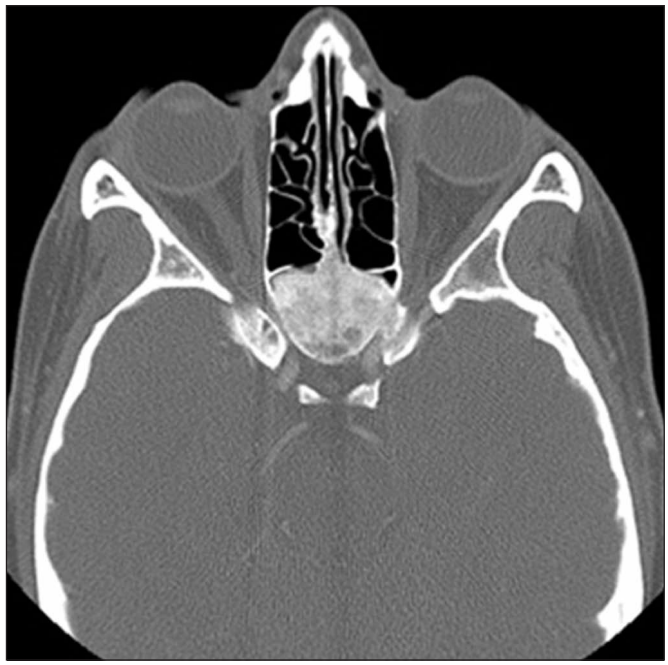

Figure 1: *Axial CT of the head without contrast enhancement demonstrating the extent of the patient's CFD and the right optic canal narrowing (arrow) after prior craniotomy and before her pregnancy and subsequent ACD.*

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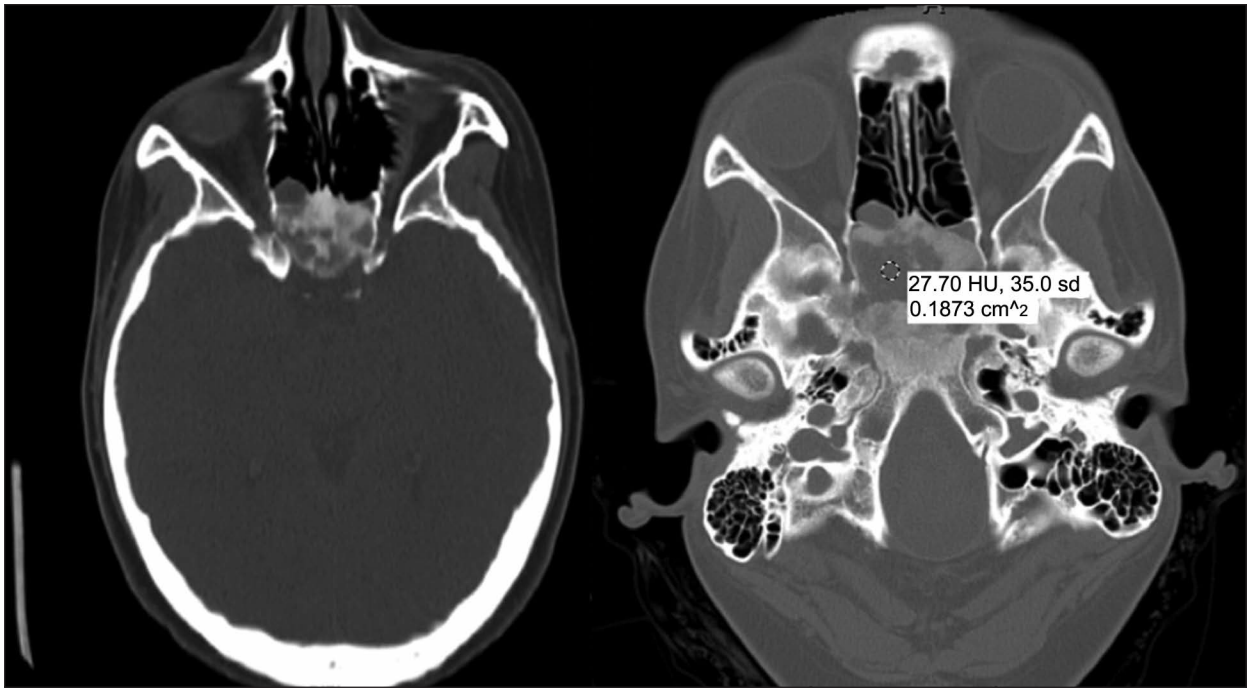

Figure 2: *A: Axial CT scan without contrast enhancement demonstrating acute cystic degeneration that occurred at the time of presentation during the patient's pregnancy demonstrating worsened optic canal narrowing that had become symptomatic. B: Axial CT image without contrast enhancement showing the Houndsfield Units of the area of ACD, which confirm that the hypodense region is fluid and not fibrous bone.*

several authors have reported recurrences of bone cysts after the removal of fibro-osseous lesions, but the correlation to ACD in CFD is unclear[7]. Acute cystic degeneration in non-craniofacial fibrous dysplasia has been demonstrated in many additional sites throughout the body, including the ribs and tibia.

Interestingly, high concentrations of estrogen receptors have been found in the osteogenic cells removed from the abnormal bone of fibrous dysplasia patients, and this may suggest a mechanism to explain initial CFD presentations occurring in pregnancy[4]. This case raises the question of whether pregnancy may also be related to ACD in CFD and shows the profound bony transformation that can occur. Surgical treatment is recommended when CFD causes visual impairment or cosmetic deformity.

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References

1. Diah E, Morris DE, Lo LJ, Chen YR. Cyst degeneration in craniofacial fibrous dysplasia: clinical presentation and management. J Neurosurg. 2007 Sep;107(3):504-8.
2. Lisle DA, Monsour PA, Maskiell CD. Imaging of craniofacial fibrous dysplasia. J Med Imaging Radiat Oncol. 2008 Aug;52 (4):325-32.
3. Dumont AS, Boulos PT, Jane JA, Jr., Ellegala DB, Newman SA, Jane JA, Sr. Cranioorbital fibrous dysplasia: with emphasis on visual impairment and current surgical management. Neurosurg Focus. 2001;10(5):E6.
4. Stevens-Simon C, Stewart J, Nakashima II, White M. Exacerbation of fibrous dysplasia associated with an adolescent pregnancy. J Adolesc Health. 1991 Jul;12(5):403-5.
5. Nelson RB. Pregnancy associated with polyostotic fibrous dysplasia. Med Ann Dist Columbia. 1948 Mar;17(3):157-9.
6. Daly BD, Chow CC, Cockram CS. Unusual manifestations of craniofacial fibrous dysplasia: clinical, endocrinological and computed tomographic features. Postgrad Med J. 1994 Jan;70 (819):10-6.
7. Ferretti C, Coleman H, Dent M, Altini M. Cystic degeneration in fibrous dysplasia of the jaws: a case report. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 1999 Sep;88(3):337-42.