Case 1: Periorbital swelling

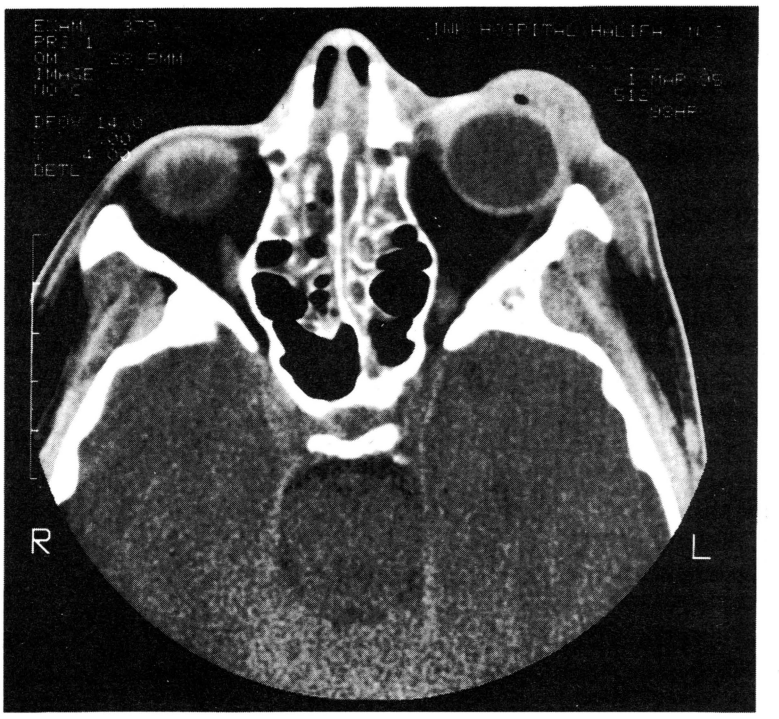
A 13 year old female was admitted with a one day history of left eye swelling. She had an upper respiratory tract infection for the week prior to admission, and developed left eye pain worse with eye movement two days prior to admission. She had been seen in the Emergency Room on the day prior, and was discharged home with a prescription for Ceclor. One month prior to admission she had streptococcal pharyngitis. There was no history of trauma.

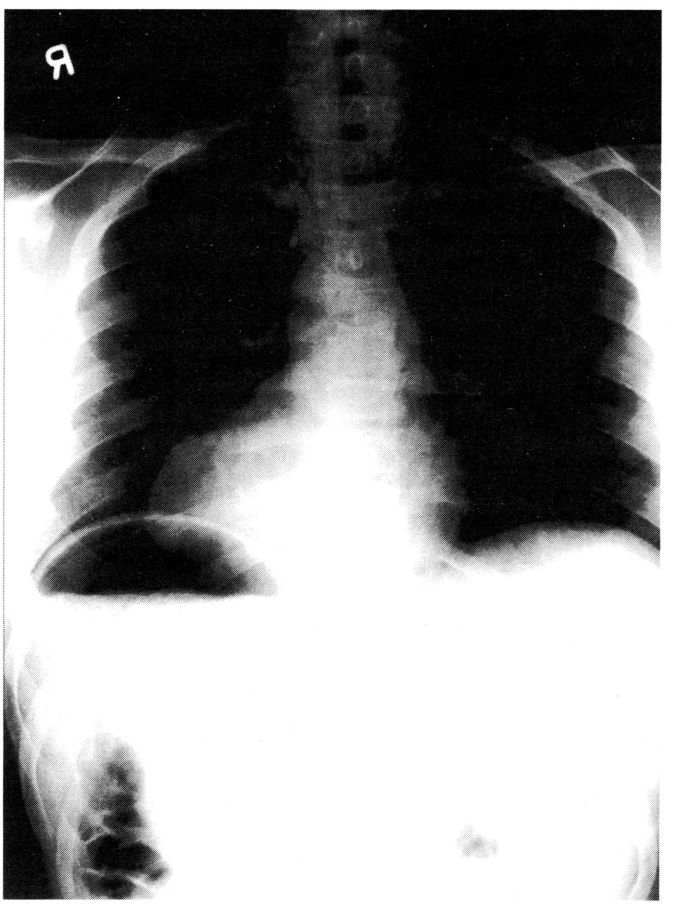
On admission, her oral temperature was 38°C, and the left eyelids were swollen and erythematous. The cornea was clear although there was mild conjunctival edema. Fundi were normal. There were bilateral cervical lymph nodes palpable, but head and neck exam was otherwise normal.

Computed tomogram (CT) of the region is shown at right.

What is the diagnosis? What is appropriate management of this problem?

(Answer on page 51)





Case 2: An abnormal chest x-ray

A 15 year old male was referred to the Otolaryngology Department at the IWK Children's Hospital for assessment of frequent upper respiratory tract infections. He had multiple episodes of sinusitis and pneumonia over the previous 3 years, some requiring intravenous antibiotics. The referring physician had noted an abnormality on the chest radiograph. He had been well earlier in his childhood, and his family history was negative.

The patient was taken to the operating room for bronchoscopy and biopsy, with no unusual findings noted. Routine postoperative chest radiograph is shown.

What does the chest x-ray at left show? Was bronchoscopy and biopsy appropriate for this child? Can this occur spontaneously?

(Answer on page 52)

Case 1.

Answer: Pansinusitis with left periorbital cellulitis

Sinusitis complicates 0.5-5% of upper respiratory tract infections (URTIs) in children (1). The clinical picture includes either nasal discharge or cough (or both) for at least 10 days without improvement, or a severe URTI with high fever and purulent nasal discharge (1). Supportive diagnostic studies include radiographs showing sinus opacification, or needle aspirate of the sinus contents yielding bacteria (1). CT scans are more accurate in showing sinus disease than plain films (2).

The same respiratory pathogens that cause otitis media cause sinusitis (i.e.: *Hemophilus influenza*, *Streptococcus pneumoniae* and *Moraxella catarrhalis*; 1-3). Thus the first line antibiotic choices are similar, including Amoxicillin and Trimethoprim-Sulfamethaoxazole (2). Treatment should continue until the child is free of symptoms for one week, resulting in typical treatment periods of 2 to 3 weeks in acute sinusitis (1,2). Adjunctive therapy includes saline nasal irrigation or topical decongestants (1). Antihistamines are not recommended since they may cause inspissation of secretions (1).

Recurrent or chronic sinusitis in children occurs when there is obstruction to normal mucus flow. This can be secondary to anatomic abnormalities such as septal deviation, bullous middle turbinates or polyps, foreign bodies in the nose, as well as immunodeficiency, allergic rhinitis, or coexisting respiratory tract disease (such as asthma or cystic fibrosis; 3).

Complications of sinusitis include osteitis or osteomyelitis, mucoceles, intracranial extension, for example purulent meningitis and abscess (epidural, subdural, and brain) and orbital inflammation (4). Sinus infections are the most common cause of the latter (4). The orbit forms the floor of the frontal sinus, the roof of the maxillary sinus, and shares its medial wall (lamina papyracea)with the ethmoid sinus (5). Spread occurs via dehiscences and foramina in the lamina papyracea or less commonly, through the valveless ophthalmic venous system (6). Orbital complications are most common in children and may have serious sequelae such as blindness from central retinal artery occlusion, optic neuritis, nerve compression, keratitis and corneal ulceration, or panophthalmitis (6,7).

Orbital complications of acute sinusitis are classified into 5 groups (Chandler's classification) having significance for management and indicating progression of disease (6). These are defined by clinical or surgical findings (6), or CT appearance (7,8). Group I, inflammatory edema, is manifested by nontender lid swelling and Group II, orbital cellulitis, involves edema of the entire orbital contents. Both require intravenous antibiotics (6). These disorders are typically cared for by primary care physicians with the monitoring of an ophthalmologist, whose expertise is required for a minimum standard of care of periorbital inflammation secondary to sinusitis (9). Groups III and IV are both abscesses; the first is a circumscribed subperiosteal collection which does not interfere with ocular mobility although it may cause proptosis. The latter is a discrete abscess within the orbital tissues causing ophthalmoplegia and more marked proptosis.

Abscesses require drainage as well as intravenous anti-biotics (5,9). Subperiosteal abscesses, most common on the medial wall, can be drained by an external ethmoidectomy or by opening the ethmoid air cells in the middle meatus and accessing the lamina papyracea endoscopically (9). Cultures may be obtained during this procedure.

Cavernous sinus thrombosis, Group V of Chandler's classification, represents the most severe stage of this entity. It results from extension along the veins to the cavernous sinus, followed by clinical signs in the opposite eye (6). Meningismus, multiple cranial nerve palsies and a moribund clinical appearance support the diagnosis (8).

Management of periorbital cellulitis includes CT scanning if visual acuity is less than 20/40, the patient has proptosis and decreased ocular mobility, or the etiology of the cellulitis is uncertain (8). Intravenous antibiotics and if necessary abscess drainage is coupled with serial ophthalmological and CT assessments (8).

The patient described in Case 1 received intravenous Cefuroxime but showed no improvement after 48 hours, so she underwent endoscopic sinus surgery which revealed pus in the left frontal sinus. The lamina papyracea was decompressed and orbital pus was evacuated. This grew Group A streptococcal species sensitive to Clindamycin. She received this intravenously for a week then was discharged home with a prescription for oral antibiotics. Ophthalmological assessment showed that her acuity was 6/6 with some diplopia on upgaze which gradually resolved.

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Case 2.

Answer: Situs inversus with pneumomediastinum.

The referring physician noticed that the chest x-ray showed situs inversus, as the heart, stomach bubble, and liver are reversed to their normal location. This post-biopsy chest x-ray shows pneumomediastinum as well as situs inversus. A thin dark line representing air is clearest at the right heart border. This patient also had clinical signs of pneumomediastinum including subcutaneous emphysema at the neck and Hamman's sign (a series of precordial crackles synchronous with the heartbeat; 1). This is a known complication of bronchial biopsy, and was treated conservatively with complete resolution in this case.

The bronchial biopsy was performed in order to obtain a sample of respiratory mucosa for electron microscopy to analyse ciliary morphology. This is appropriate investigation for recurrent/chronic respiratory disease, following investigation to rule out allergic disease, cystic fibrosis, and hypogammaglobulinemia as underlying entities (2). The putative diagnosis was Kartagener's syndrome, a combination of situs inversus and primary ciliary dyskinesia, which manifests as chronic sinusitis, purulent rhinitis, otitis media, bronchitis, recurrent pneumonia, bronchiectasis, nasal polyps, and male sterility (2). Two other syndromes are relevant in this clinical scenario; the first is primary ciliary dyskinesia syndrome, which combines abnormal cilia with normal visceral status, and Young syndrome where the clinical picture consists of sinusitis, bronchiectasis, and obstructive azospermia but ciliary ultrastructure is normal (2).

Ciliary beat has a crucial role in the clearance of secretions from sinuses and the lower respiratory tract. The most common abnormality associated with this clinical scenario is absence of dynein arms resulting in ineffective motility although other defects are also possible (2).

Pneumomediastinum can occur secondary to asthma, inhalational drug use, labour, coughing, rapid reduction in atmospheric pressure, or other activities associated with Valsalva maneuvers where terminal alveoli are thought to burst allowing air to track along the pulmonary vasculature into the mediastinum (1,3). Thirty percent of those presenting to the Emergency Room with this condition have no antecedent event, resulting in a diagnosis of spontaneous pneumomediastinum (1). This is most common in young males but has been reported in children as well (3). The most common symptom is stabbing retrosternal pain radiating to the back or shoulders, which is lessened by shallow respiration or leaning forward (3). Dyspnea, dysphagia or a feeling of fullness in the throat may also be present (3,4). Signs include subcutaneous emphysema, Hamman's sign, and decreased cardiac dullness on percussion (3). Diagnosis is made by chest x-ray (1,3,4). Management in the past has consisted of hospitalization for bedrest and analgesia, along with serial chest x-rays (3). A recent review of 17 cases suggests that patients without underlying disease and stable vital signs may be managed with outpatient rest, analgesics, and avoidance of Valsalva producing activities (4). These authors found that serial chest x-rays did not change management in any of their cases, and advise patients to return if symptoms change at which time a chest x-ray is repeated (4).

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Permission to present these cases was given by KD Clarke, Head of the Department of Otolaryngology, Izaak Walton Killam Children's Hospital. Members of the Nova Scotia medical community are invited to submit cases for this diagnostic challenge section.