DIAGNOSTIC CHALLENGE

Case 1: Unilateral rhinorrhea

A 3 year old male presented with a longstanding history of right nasal discharge. He had no known allergies and was otherwise healthy. There was no abnormality appreciable when looking in his nose. Plain views of the sinuses were normal. This CT scan shows a congenital abnormality accounting for his symptoms. What is the diagnosis? How is this problem managed? What other congenital anomalies are frequently associated with this?

(Answer on page 60)





Case 2: Draining Buttock Wound

A 73 year old female complained of a 3 month history of a "hole" in her right hip. It was not painful but discharged copious amounts of serous fluid. One week prior she had retrieved a 3 cm long piece of hard matter from the wound. Her past medical history included osteoarthritis, a lower gastrointestinal hemorrhage 2 weeks prior and congestive heart failure. On examination she had a 0.5×0.5 cm sinus tract in the upper outer quadrant of the right buttock with a hard mobile mass palpable just superior to it. This shows the plain film of the region. What is the abnormality and what can cause it? Which systemic diseases should be ruled out?

(Answer on page 60)

DIAGNOSTIC CHALLENGE - ANSWERS

Case 1.

Answer: Right choanal atresia

Choanal atresia is the total obstruction of the posterior nasal choanae, the opening between the nose and nasopharynx(1). It occurs in 1 of 5000 live births, is twice as common in females and twice as likely to be unilateral as bilateral(1). A recent CT study suggests that 30% are purely bony obstructions and 70% are mixed bony and membranous(2). The bone in the atretic region may be 1 to 12 mm thick and is bounded by the underside of the body of the sphenoid superiorly, the medial pterygoid lamina laterally, the horizontal portion of the palatal bone inferiorly, and the vomer medially(3).

Embryologic theories explaining choanal atresia include errors in neural crest cell migration or persistence of the buccopharyngeal membrane(3).

Bilateral choanal atresia is a medical emergency since newborns are initially obligate nasal breathers. These children typically present with cyclical cyanosis worsened by feeding and improved by crying(1). Respiratory distress is relieved by crying because the child then breathes through its mouth. Confirmation of the diagnosis is achieved by failure to pass a number 8 French catheter past 32 mm from the anterior nares(1). Initial management consists of an oral airway and oral-gastric tube feedings(1). The baby learns to mouthbreathe within hours to days(1). Presentation of unilateral choanal atresia is more subtle, as a 1 or 2 year old child with unilateral rhinorrhea or sinusitis(1). Axial CT scans best show the defect(2).

Sixty percent of children with choanal atresia have a recognized syndrome or association of other anomalies(4). Most of these have at least one anomaly described by the CHARGE association, which includes: Colobomata of retina/iris, Heart defects, Atresia of the posterior choanae, Retarded growth and development, Genital abnormalities in males, External ear malformations, and deafness.

Repair of choanal atresia takes place once the child's medical status has been optimized. Approaches are transnasal or transpalatal; the latter may interfere with palatal growth and cause malocclusion and so is recommended for older children(5). Transnasal repair utilizes an endoscope or microscope and removal of the atretic plate with a rotating burr, curette or laser(5). Stenting with a silastic tube for up to 2 months is done to maintain patency(1,2,5). At the IWK Children's Hospital these children are most commonly treated with a transnasal approach using a Holmium:YAG laser to create a posterior choana, without post-operative stenting. This is in an attempt to decrease the incidence of restenosis, a common problem requiring dilatation or revision surgery(5).

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Case 2.

Answer: Heterotopic calcification

Calcification in abnormal areas such as soft tissue is commonly seen on plain radiographs, especially in the buttock region(1,2,3). Numerous mechanisms are thought to be related. Calcium abnormalities such as primary and secondary hyperparathyroidism, hypervitaminosis D, sarcoidosis and other granulomatous disorders, lymphomas, idiopathic hypercalcemia, Williams syndrome, and Vitamin D metabolic disorders may result in heterotopic calcification(4). Connective tissue disorders such as systemic lupus erythematosis, scleroderma, CREST syndrome, and inflammatory myositis are also associated, as is trauma, including burns and intramuscular injections(1,2,3,4). In the latter, it is unclear whether the mechanism is related to the trauma of injection, or a characteristic of the medication injected(4). In any case, a granuloma develops which becomes calcified with time(1). Medications implicated include vaccinations, antibiotics, insulin, Vitamins E and D, papaverine, calcium gluconate, and ferric chloride(1,4). Typically these form ring-like calcifications in common injection sites such as the buttock or deltoid region(2,4).

Haramati and colleagues reviewed 338 consecutive pelvic CTs to describe calcifications in buttock soft tissues(1). Twenty percent had these, 93% were in fat with the remainder in muscle. They also measured subcutaneous fat thickness in the upper outer quadrant of the buttock on these scans and found it to average 5 cm. This is thicker than the length of a long hypodermic needle (3.8 cm) used for intramuscular injections in this area, suggesting that these medications are actually injected into fat. Injecting medications into fat as opposed to muscle results in altered pharmacokinetics, fat necrosis, granuloma formation, and thus calcification.

This patient underwent an autoimmune workup which was negative. The calcification was excised and the area debrided under local anaesthesia. With careful wound care, it gradually healed by secondary intention.

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