SELF ASSESSMENT ANSWERS

Abdominal pain and vomiting in an elderly diabetic woman

Q1: What abnormalities are seen on the plain film (fig 1; see p 709) and computed tomogram (fig 2; see p 709) of the abdomen?

Plain film shows air in the biliary tract and gall bladder, dilated loops of small intestine, and multiple air-fluid levels in small bowel loops. The computed tomogram shows large gall stone impacted in jejunum and multiple air-fluid levels in small intestine.

Q2: What is the diagnosis?

Gall stone ileus.

Q3: What other variants of this disease are known to occur?

The complications of cholelithiasis are shown in table 1. Gall stones can get impacted at various sites, the commonest being the ileocaecal junction. The other described sites of gall stone impaction with in the bowel are the jejunum, stomach, colon, duodenum, and pylorus (Bouveret's syndrome).

GALL STONE ILEUS

Gall stone ileus is a mechanical bowel obstruction caused by passage of gall stones from the biliary system through a biliary-enteric fistula with impaction within lumen of the bowel.

It is an uncommon complication of biliary stone disease, accounting for only 2% of all cases of intestinal obstruction. Gall stone ileus is, however, more common in the elderly and accounts for approximately 25% of all cases of intestinal obstruction in patients over 65 years of age.[1]

The gall stone that causes ileus is usually more than 2.5 cm in diameter. Multiple stones are present in 3%–15% of cases.[1 2]

Gall stones usually enter the intestinal lumen through a cholecystoenteric fistula, and 68% of these are between the gall bladder and the duodenum. A history of prior biliary tract disease is present in almost 50%–60% of patients with gall stone ileus.[2]

The most frequent site of stone impaction is the ileum (>60% of cases), as it is the narrowest part of the bowel. Other sites of obstruction are the jejunum (16%), stomach (14%), colon (4%), and duodenum (3%). Gastric outlet obstruction, or Bouveret's syndrome, occurs when the gall stone lodges in the duodenal bulb (1%).[1]

Table 1 Complications of cholelithiasis

Complication	Percent
Biliary colic	70–80
Acute cholecystitis	10
Emphysematous cholecystitis	<1
Mirizzi's syndrome	<1
Hydrops of the gall bladder	<1
Small bowel obstruction (gall stone ileus)	1
Perforation of gall bladder	<1
Acute biliary pancreatitis	12
Acute suppurative/obstructive cholangitis	–

Clinical features

Usually abdominal pain is a prominent symptom, and associated illnesses such as diabetes and cardiovascular disease are common. It causes signs of small bowel obstruction like nausea, vomiting, abdominal distension, and absence of bowel sounds in cases of complete obstruction. Signs of intestinal obstruction are seen mostly if the gall stone impacts at the ileocaecal junction. However, the characteristic features of intestinal obstruction are found in only 50% to 70% of patients.[2 3] This may be because as the gall stone ''tumbles'' through the gastrointestinal tract, it impacts and disimpacts, producing intermittent mechanical obstruction.

Investigations

The diagnosis of gall stone ileus is often difficult to make.

The classic radiographic signs of gall stone ileus on abdominal plain film are pneumobilia, mechanical small bowel obstruction, and the presence of a new stone or changed position of a previously identified stone, known as Rigler's triad.[1]

Ultrasound reveals diseased gall bladder, whether there is gas in it or in the bile ducts or both, and fluid filled bowels that can be followed to the stone in the intestine. The presence of stones in the gall bladder will modify the planned operative procedure in the treatment of gall stone ileus.

The characteristic features of Rigler's triad are easily identified on computed tomography even if abdominal plain film is subtle. Even if abdominal radiography reveals the characteristic signs of small bowel obstruction, computed tomography is useful for excluding complications (for example, strangulation). When unexplained bowel obstruction is present, particularly in the elderly, the early use of computed tomography is strongly recommended.

Endoscopy has been the main diagnostic procedure for Bouveret's syndrome. The diagnosis was made endoscopically in more than 90% of the cases.[4]

Treatment

Gall stone ileus is a serious geriatric surgical emergency. It has a high morbidity (15%–18%) and mortality (17%).

The management of gall stone ileus is controversial. The choice is between performing simple enterolithotomy or a single stage procedure involving enterolithotomy, cholecystectomy, and fistula closure. Current reports favour enterolithotomy only, with definitive biliary surgery performed later if symptoms persist. Advocates of the combined procedure contend that it prevents recurrent gall stone ileus, cholangitis, and gall bladder carcinoma complications that occur in nearly one third of patients who undergo enterolithotomy only. Simple enterolithotomy carries a mortality of 11.7% compared with 16.9% for one stage procedure. The most common source of operative morbidity is wound infection, occurring in 30%–40% of cases.[5 6]

In duodenal stone impaction extracorporeal shock wave lithotripsy is successful in fragmenting the stone. Endoscopic stone removal is especially indicated in poor risk patients. A dislodged impacted stone can migrate distally and cause small bowel mechanical obstruction that might require urgent enterolithotomy.[4]

The recurrence rate of gall stone ileus is less than 2%.[2]

Final diagnosis

Gall stone ileus.

References

1 **Foss HL**, Summers JD. Intestinal obstruction from gall stones. *Ann Surg* 1942;**115**:721–35.
2 **Reisner RM**, Cohen JR. Gallstone ileus: a review of 1001 reported cases. *Am Surg* 1994;**60**:441–6.
3 **Clavien PA**, Richon J, Burgan S, *et al*. Gallstone ileus. *Br J Surg* 1990;**77**:737–42.
4 **Williams IM**, Hughes ODM, Hicks E, *et al*. Gall stone ileus following multiple endoscopic retrograde cholangiopancreatographies. *J R Coll Surg Edinb* 1997;**42**:423–4.
5 **Deitz DM**, Standage BA, Pinson PW, *et al*. Improving the outcome in gallstone ileus. *Am J Surg* 1986;**151**:572–6.
6 **Rodriguez-Sanjuan JC**, Casado F, Fernandez MJ, *et al*. Cholecystectomy and fistula closure versus enterolithotomy alone in gallstone ileus. *Br J Surg* 1997;**84**:634–7.

Proptosis in an asthmatic patient

Q1: What is the diagnosis of this respiratory condition?

The diagnosis in this case is allergic bronchopulmonary aspergillosis (ABPA). ABPA occurs in 6%–20% of individuals with asthma and approximately 10% of individuals with cystic fibrosis.[1] It is characterised by repeated episodes of exacerbations interspersed with periods of remissions. Diagnostic criteria include (1) asthma, (2) fleeting pulmonary opacities on chest radiographs, (3) positive skin tests for *Aspergillus fumigatus*, (4) peripheral blood eosinophilia, (5) precipitating antibodies to *A fumigatus*, (6) raised serum IgE levels, and (7) central bronchiectasis.[2] Other minor criteria include expectoration of golden brown sputum plugs, positive sputum culture for aspergillus species and late (Arthus-type) skin reactivity to *A fumigatus*.

If the disease is not diagnosed early and treated adequately, lung damage continues to progress silently leading to fibrosis in the upper lobes.[3] The upper lobe fibrosis of ABPA can mimic pulmonary tuberculosis and patients are often unnecessarily treated with antituberculosis therapy as in our case.

Q2: Which respiratory diseases have coexistent sinus and orbital involvement and what was the cause in this patient?

Allergic and granulomatous diseases commonly involve the lung, sinuses, and the orbit (box 1).[4] The diagnosis in this condition was allergic fungal sinusitis with orbital aspergillosis in a patient with allergic bronchopulmonary aspergillosis.

Q3: What is the significance of C-ANCA in chronic lower respiratory tract infections?

Positive C-ANCA is characteristic of Wegener's granulomatosis. However, in a study done by Ohno *et al* raised myeloperoxidase ANCA and bactericidal/permeability increasing protein ANCA levels were found in patients with chronic lower respiratory

Box 1: Allergic and granulomatous diseases

- *Allergic group:*
  - ABPA.
- *Granulomatous group:*
  - Classical Wegener's granulomatosis.
  - Necrotising sarcoid granulomas.
  - Behçets disease.
  - Midline granulomas.
  - Polyarteritis nodosa.
  - Churg-Strauss syndrome.
  - Relapsing polychondritis.
  - Benign lymphocytic angitis.
  - Nasopharyngeal lymphomas with pulmonary Hodgkin's disease.

infection.[5] Proteinase-3 C-ANCA was also positive in a single case each of the 21 diffuse panbronchiolitis and 16 bronchiectasis patients included in the study. The raised C-ANCA levels found in our patient can thus be explained by the fact that he had underlying chronic respiratory tract infection.

Discussion

Allergic fungal sinusitis is a saprophytic fungal growth involving one or more paranasal sinuses. Allergic fungal sinusitis appears to be more prevalent in the temperate regions and around areas of high relative humidity with equal preponderance in both sexes.[6] It can manifest itself either with signs and symptoms of nasal obstruction and allergic rhinitis or purulent rhinorrhoea, headache, and epistaxis. Orbital aspergillosis occurs as an extension of allergic fungal sinusitis into the adjacent spaces, which may have a dramatic clinical presentation such as diplopia or visual loss due to compression of the ophthalmic nerve. The gross facial disfigurement and orbital abnormalities consisting of proptosis and telecanthus are more often seen in children than in adults. The diagnosis of allergic fungal sinusitis is principally based on pathological findings in the specimens obtained from the paranasal sinuses. These findings are similar to those seen in the mucoid impaction of ABPA.[6]

Concomitant occurrence of ABPA and allergic fungal sinusitis is well recognised with substantial data available on the same.[7] It has been postulated that the hallmark of this saprophytic colonisation is an identical pathophysiology occurring in ABPA, allergic fungal sinusitis, and orbital aspergillosis. There are several case reports of coexisting ABPA with allergic fungal sinusitis[7] and allergic fungal sinusitis with orbital aspergillosis.[6] The presence of ABPA and allergic fungal sinusitis in the same patient is often overlooked when either is being treated by two different specialties.

The management of allergic fungal sinusitis with orbital involvement warrants an immediate surgical removal of the fungal growth including resection of any recurring disease. Medical management includes the use of corticosteroids as in ABPA, while topical corticosteroids may be effective in controlling local inflammation. It is also recommended that systemic antifungals be used to prevent progression to invasive forms of fungal sinusitis. Intravenous amphotericin-B has been found to be more beneficial than the less toxic agents of the azoles group in view of the poor in vitro activity of these agents.[6]

In conclusion, patients with ABPA should be evaluated for the presence of coexistent allergic fungal sinusitis and orbital aspergillosis to avoid further complications.

Final diagnosis

Allergic fungal sinusitis with orbital aspergillosis in a patient with allergic bronchopulmonary aspergillosis.

References

1 **Slavin R**, Stanevyak D, Lonigro A, *et al.* Allergic bronchopulmonary aspergillosis: a North American rarity. *Am J Med* 1969;**47**:306–8.
2 **Rosenberg M**, Patterson R, Mintzer R, *et al.* Clinical and immunological criteria for the diagnosis of allergic bronchopulmonary aspergillosis. *Ann Intern Med* 1977;**86**:405–14.
3 **Shah A**, Panjabi C. Allergic bronchopulmonary aspergillosis: a review of a disease with worldwide distribution. *J Asthma* 2002;**39**:273–89.
4 **Seaton A**, Seaton D, Leitch AG. Pulmonary angiitis and granulomatosis. *Crofton and Douglas's respiratory diseases*. 4th Ed. New Delhi: Oxford University Press, 1989:748–60.
5 **Ohno S**, Saku N, Sugiyma Y. ANCA (antineutrophil cytoplasmic autoantibody) in chronic lower respiratory tract infection. *Chest* 2001;**120**(suppl):4.
6 **McClay John E**, Marple Bradley. *Allergic fungal sinusitis*. Accessed 28 December 2001 (www.emedicine.com/ent /topic 510.htm); volume 2, number 12.
7 **Shah A**, Panchal N, Agarwal AK. Concomitant allergic bronchopulmonary aspergillosis and allergic aspergillus sinusitis a review of an uncommon association. *Clin Exp Allergy* 2001;**31**:1896.

Rheumatoid arthritis and neck pain

Q1: What are the features seen in the lateral radiographs of the cervical spine (p 711)?

Lateral radiographs of the cervical spine show increased atlas-dens interval anteriorly and reduced posterior atlas-dens interval in flexion views but normal intervals in the extension views. The spinal canal shows corresponding reverse changes. In addition, degenerative changes are seen at the C5–C6 intervertebral disc space.

Box 1: Groups of C1–C2 involvement in rheumatoid arthritis based on the radiological features

- Group 1 (46%): severe joint space narrowing and subchondral sclerosis with lateral mass collapse.
- Group 2 (44%): joint space narrowing and subchondral sclerosis occurs without lateral mass collapse.
- Group 3 (10%): lateral subluxation without joint space narrowing or subchondral sclerosis.

Q2: What abnormalities are seen on the MRI scan (p 711)?

Sagittal T2 weighted MRI scan of the cervical spine shows narrowing of the bony spinal canal at the arch of C1 vertebra. This in turn gives rise to generalised narrowing and distortion of the spinal cord at the craniomedullary junction. There is some signal change within the cord itself at this level.

Q3: What is the likely diagnosis?

Anterior atlantoaxial subluxation involving supra-axial spine.

Q4: How can this condition be treated?

Atlantoaxial subluxation, when symptomatic or is severe, can be treated by posterior cervical fusion with or without stabilisation at this level. Arthrodesis with autologous corticocancellous bone graft is augmented by various stabilisation techniques.

Discussion

Rheumatoid arthritis is a chronic, inflammatory disorder characterised by symmetric polyarthritis involving multiple joints. The cervical spine can be involved in 17%–86% of patients with rheumatoid arthritis. It can involve the supra-axial or subaxial spine with the craniovertebral junction being most often affected.[1] Cervical instability is the most serious and potentially lethal manifestation of rheumatoid arthritis. Also, a patient with rheumatoid arthritis and an unstable cervical spine will present a major anaesthetic problem if not stabilised. Atlantoaxial subluxation can be anterior, posterior, or lateral with the anterior type being most common.[2] It is usually the result of the destruction of joints, ligaments, and bone caused by erosive synovitis involving atlantoaxial, atlanto-odontoid, and atlanto-occipital joints.[3] The inflammatory destruction via synovitis of the transverse ligament leads to anterior subluxation of the atlas on the axis. Erosion of the odontoid process frequently coincides with this process. Protrusion of the odontoid process posteriorly into the spinal canal can result in clinical symptoms or signs such as suboccipital pain or neuralgia and myelopathy. Further progression of the disease involves loss of alar and capsular ligament integrity and leads to further erosion of the dens.[1] If the inflammation persists in the atlantoaxial joints, their cartilage and bone structures will be eroded, the joint spaces will narrow down, and the atlas falls down around the axis (atlantoaxial impaction). When the atlantoaxial joint surfaces are

Box 2: Ranawat scale used for neurological assessment in rheumatoid arthritis

- Grade 1: pain, no neurological deficit.
- Grade 2: subjective weakness, hyperreflexia, dysaesthsia.
- Grade 3a: objective weaknesss, long tract signs, ambulatory.
- Grade 3b: objective weaknesss, long tract signs, non-ambulatory.

eroded they become rough and the instability decreases (box 1).[2 4]

Anterior atlantoaxial subluxation implies a widening of the joint space between the anterior arch of the atlas and the odontoid process. Radiographic diagnostic criteria have been developed as descriptors of existence and advancement of atlantoaxial subluxation. Two of the most commonly used criteria are anterior and posterior atlantoaxial intervals (AADI and PADI respectively). The determination of these intervals involves constructing a line that connects the centroids of the anterior and posterior rings of the atlas on a lateral plain radiograph at maximal flexion. The AADI is the distance along this line that measures the difference between the posterior surface of the anterior arch of the atlas and the anterior surface of the dens. The PADI, which is complementary to AADI, is the distance between posterior surface of dens and anterior surface of posterior arch of atlas. The normal AADI is 1 to 2 mm in adults (sometimes referred to as atlantodens interval, ADI).[3] In children, the ADI may be as much as 4.5 mm and can show an increase of 0.5 mm in flexion. Atlantoaxial subluxation is defined as the AADI >3 mm or PADI $\leqslant 14$ mm.[1]

Atlantoaxial subluxation of 9 mm reduces the area of the spinal canal (space available for the cord) to 60%. Theoretically, full rotation of 47° would further reduce the spinal canal to 21% which must cause compression of the cord (as cord normally occupies 27 to 30% of the spinal canal). The rheumatoid pannus also contributes to medullary compression in cases of atlantoaxial subluxation.[3]

The preoperative PADI value is a more reliable indicator of the development and severity of paralysis. Moreover, PADI is a better predictor than AADI of whether postoperative neurological recovery is expected. This is because PADI correlates closely with the space available for the spinal cord.[1 5]

Patient controlled flexion and extension views are evaluated to determine the AADI and PADI. Instability is present with a 3 mm of AADI difference in flexion and extension views, although radiographic instability in rheumatoid arthritis is common and is not an indication for surgery. Cervical spine surgery is seldom indicated solely by radiographic findings. These patients should be examined by with MRI to get more information about possible spinal cord compression and also to visualise other soft tissues, such as pannus, before a final decision on surgical treatment.[2]

The assessment of cervical disease in patients with rheumatoid arthritis can be difficult due to coexisting systemic illnesses, neurological abnormalities, and rheumatoid polyarthropathy.[5] The Ranawat scale is often used to grade rheumatoid myelopathy because its coarse grading structure accepts the major musculoskeletal disability in rheumatoid arthritis (see box 2).

Rheumatoid cervical spine changes are usually treated conservatively. The indications for surgery are well established in the symptomatic patient. Severe pain may be alleviated and neurological deterioration relieved. Controversy still exists, however, over the role of prophylactic procedures in asymptomatic patients.[5] Surgery has been recommended in cases with extensive subluxation or gross instability even without neurological deficit to avoid development of myelopathy.

The commonest surgical procedure is posterior C1–C2 fusion and wiring[5] with additional halovest stabilisation or C1–C2 transarticular screw fixation. Atlantoaxial subluxation that is not reducible may require removal of the posterior arch of atlas for cord decompression followed by occiput to axis fusion. An AADI of >7–10 mm or a posterior space (PADI) of less than 13 mm is a relative contraindication to surgery in other areas and the spine should be stabilised first.

Final diagnosis

Anterior atlantoaxial subluxation of the cervical spine in rheumatoid arthritis.

References

1 **Puttlitz CM**, Goel VK, Clark CR, *et al*. Biomechanical rationale for the pathology of rheumatoid arthritis in the craniovertebral junction. *Spine* 2000;**25**:1607–16.
2 **Neva MH**, Kaarela K, Kauppi M. Prevalence of radiological changes in the cervical spine—a cross sectional study after 20 years from presentation of rheumatoid arthritis. *J Rheumatol* 2000;**27**:90–3.
3 **Tucker SK**, Taylor BA. Spinal canal capacity in simulated displacements of the atlanto-axial segment—a skeletal study. *J Bone Joint Surg Br* 1998;**80**:1073–8.
4 **Halla JT**, Hardin Jr JG. The spectrum of atlantoaxial facet joint involvement in rheumatoid arthritis. *Arthritis Rheum* 1990;**33**:325–9.
5 **Eyers KS**, Gray DH, Robertson P. Posterior surgical treatment for the rheumatoid cervical spine. *Br J Rheumatol* 1998;**37**:756–9.

ECHO

QRS duration: a simple marker for predicting cardiac mortality in ICD patients with heart failure

L Bode-Schnurbus, D Böcker, M Block, R Gradaus, A Heinecke, G Breithardt, M Borggrefe

Please visit the *Postgraduate Medical Journal* website [www.postgradmedj.com] for a link to the full text of this article.

Background: Patients resuscitated from ventricular tachyarrhythmias benefit from implantable cardioverter-defibrillators (ICDs) as opposed to medical treatment. Patients with increased QRS duration receiving an ICD in the presence of heart failure are at greatest risk of cardiac death and benefit most from ICD therapy.
Objective: To determine whether an increased QRS duration predicts cardiac mortality in ICD recipients.
Design: Consecutive patients with heart failure in New York Heart Association functional class III were grouped according to QRS duration ($<$ 150 ms, n = 139, group 1; *v* $\geqslant$150 ms, n = 26, group 2) and followed up for (mean (SD)) 23 (20) months.
Patients: 165 patients were studied (80% men, 20% women); 73% had coronary artery disease and 18% had dilated cardiomyopathy. Their mean age was 62 (10) years and mean ejection fraction (EF) was 33 (14)%. They presented either with ventricular tachycardia (VT) or ventricular fibrillation (VF).
Main outcome measures: Overall and cardiac mortality; recurrence rates of VT, fast VT, or VF.
Results: Mean left ventricular EF did not differ between group 1 (33 (13)%) and group 2 (31 (15)%). Forty patients died (34 cardiac deaths). There was no difference in survival between patients with EF $>$ 35% and $\leq$35%. Cardiac mortality was significantly higher in group 2 than in group 1 (31.3% at 12 months and 46.6% at 24 months, *v* 9.5% at 12 months and 18.2% at 24 months, respectively; p = 0.04). The recurrence rate of VT was similar in both groups.
Conclusions: Within subgroups at highest risk of cardiac death, QRS duration—a simple non-invasive index—predicts outcome in ICD recipients in the presence of heart failure.

▲ *Heart* 2003;**89**:1157–1162.