VALENTINO’S SYNDROME IN AN ADOLESCENT BOY WITH PEPTIC ULCER PERFORATION SIMULATING ACUTE APPENDICITIS

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Abstract
We wish to report a 14-year-old male adolescent who presented with sudden onset epigastric followed by right sided abdominal pain, vomiting and fever. He has right sided abdominal guarding, rigidity and tenderness simulating appendicitis initially. He had strong family history of peptic ulcer perforations. The abdominal radiograph showed football sign and upright chest radiograph confirmed pneumoperitoneum. At laparoscopy duodenal ulcer perforation was diagnosed. Patient underwent primary repair of duodenal perforation with omental patch and peritoneal toilet uneventfully. The presentation, treatment, and outcome of this case requiring surgery for complication of pediatric peptic ulcer disease (PPUD) in the post-histamine 2 -blocker and proton pump inhibitor era has been highlighted.

Keywords: duodenal ulcer, acid peptic disease, peptic perforation, adolescent, appendicitis, peritonitis, Valentino’s syndrome.

Introduction
Gastrointestinal perforation (GIP) in general and peptic ulcer perforation (PUP) in particular is a potentially life threatening pediatric emergency requiring an appropriate diagnostic and therapeutic approach. A PPUD with complications generally present in toddlers and teenagers [1-11]. We present our case which was referred as appendicitis which was excluded immediately after examination and initial investigations. This is a case of Valentino’s syndrome not very rare in surgical practice but a common clinical trick especially for the young surgeon, named after Rodolph Valentino who presented with the right lower quadrant abdominal pain caused by perforated peptic ulcer and subsequently died due to post-operative sepsis [2]. However, one should be aware about existence of very rare dual pathology and anecdotal case in which duodenal ulcer perforation with coexistence of acute appendicitis which has been reported in the literature [3].

Case report
A 14-year-old previously healthy male adolescent under foster care presented to us at night via accident and emergency department with sudden onset epigastric and then right sided abdominal pain, vomiting and fever. Abdomen was guarded, tender and rigid on right side. Initial accident and emergency department diagnosis was acute appendicitis with perforation. He had on and off abdominal pain for the past one year and has been seen by his general practitioner and diagnosed musculoskeletal and growth pain and received paracetamol and ibuprofen as analgesic and non-steroidal anti-inflammatory medications. There was significant family history of peptic ulcer perforation of his father at the age of 36 years and one of the aunts in thirties.

On examination, patient was unwell, pale, febrile, dehydrated and had tachycardia. Abdominal examination revealed guarding, rigidity and tenderness in the right side of the abdomen. The white cell count was elevated at 20.2×10⁹/L and a C-reactive protein of <0.3 mg/L. An abdominal radiograph revealed intraperitoneal gas suggestive of a football sign (Fig. 1) and a chest radiograph identified free air under both hemi diaphragms (Fig. 2). He required fluid resuscitation on admission and...
started on triple intravenous antibiotic therapy which appeared to help initially. He underwent diagnostic laparoscopy which showed evidence of peptic perforation in the anterior aspect of first part of duodenum and four quadrant peritonitis. Perforated duodenal ulcer was repaired primarily and augmented by means of an omental patch and peritoneal toilet was carried out for peritonitis.

Post-operatively, a battery of tests to rule out Zollinger-Ellison syndrome (ZES) was carried out and was found to be within normal levels. Chest X ray was normal (Fig. 3). His duodenal biopsies were consistent with inflammation and ulceration of the mucosa and revealed no evidence of Helicobacter infection.

Following the above, he made a good recovery, tolerated feeds and was discharged home. At 12 month follow up, he is well and asymptomatic.

Discussion

Although the overall incidence of peptic ulcer disease is increasing in children, peptic ulcer perforation is rare in children. Our case illustrates that appendicitis is the most common cause of acute surgical abdomen in adolescent children and although peptic perforation is uncommon, alternate diagnoses must be borne in mind in children presenting with sudden abdominal pain and peritoneal irritation signs. Perforated chronic duodenal ulcer in children with coexistence of acute appendicitis has been reported so dual pathology rarely may co-exist together [2].

In a large series with 20 years of experience, Hua et al. [3] found mean age of presentation to be 14 years with an age range of 2-18 years, 90% were adolescent, 80% being male, 10% had coexisting clinical events before the perforated peptic ulcer disease perforation. Acute abdominal pain was seen in all patients, and over 94% had peritoneal signs. Radiography showed sub diaphragmatic free air in over 82%; this was concluded to be the most important tool for establishing diagnosis in this series of 52 patients. Diagnostic laparoscopy is a useful tool in children with visceral perforation as it avoids treatment delays and exposure to excess radiation of a CT scan [1].

Severe abdominal pain in a significantly sick adolescent associated with rigid abdomen and signs of shock should prompt one to think of possible peptic perforation. Atypical and puzzling presentations have been reported [4-5]. The differential diagnosis of peptic perforation in adolescents is determined by age family history and severity of symptoms. In adolescents mucosal irritation secondary to gastritis, alcohol consumption, steroid and non-steroidal anti-inflammatory drug medications, H Pylori infection, blood group ‘O’, Zollison-Ellison syndrome (ZES) and physiological stress ulcers in burns, head injury and mucosal ischemia may be responsible for acute exacerbation followed by frank perforation.

The fundamental aim in an adolescent with PPUD is resuscitation and stabilization followed by a diagnostic laparoscopy with a view to proceed with repair. Effective proton pump prophylaxis of children receiving steroids and non-steroidal anti-inflammatory drugs (NSAIDs) may play a role in decreasing the risk of PPUD. Our patient had recurrent upper abdominal

![Figure 1: Abdominal radiograph demonstrating football sign](image1)

![Figure 2: Upright chest radiograph showing pneumoperitoneum under both domes of diaphragm](image2)
pain interpreted to be musculoskeletal in origin and received NSAIDs without PPI cover which may have increased the risk. H. pylori infection is common in the adolescent requiring triple therapy to eradicate it and in view of strong family history; ZES or other endocrine cause should be ruled out.

Current concepts of management regarding pediatric peptic ulcer perforation include simple suture, primary suturing and Graham patch omentoplasty. Pyloroplasty with or without vagotomy and antrectomy similar to Billroth I procedure are other options in severe or recurrent case [1-11]. Laparoscopic primary repair of the perforation and Graham patch omentoplasty is adequate in most pediatric cases. Significant postoperative complications with morbidity of over 17% and mortality of nearly 4% were observed. Female sex, more coexisting clinical events, no evidence of chronic ulcer, and treatment by simple suture were associated with significant poor prognosis on univariate analysis.

However, in multivariate analysis, female sex and simple suture were the only variables statistically significant. Although delay in surgery of over 12 hours was not significantly related to complications, there was a greater tendency toward the development of complications [3]. Delayed diagnosis or mistreated perforations may carry high morbidity and mortality [8].

Conclusion
Peptic ulcer perforation can mimic appendicitis clinically and high index of suspicion based on sudden severe abdominal pain, board like rigidity and guarding, sick patient with signs of shock may help establish diagnosis early. Abdominal and chest radiographs are helpful and laparoscopy helps for early diagnosis and appropriate treatment. Primary repair with omental serosal patch (Graham patch) may be sufficient treatment in children.

Peptic ulcer perforation (PUP) should be suspected in adolescents who present with acute abdominal pain together with prominent peritoneal signs. Children with PUP have a more favourable prognosis than adults.

REFERENCES


Figure 3: Post-operative chest radiograph showing anaesthetic central line and no pneumoperitoneum.