Diagnosis of ruptured abdominal aortic aneurysm: a multicentre cohort study

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Objective **The aim of this study was to describe the presentation of patients with ruptured abdominal aortic aneurysm (rAAA) and identify factors contributing toward misdiagnosis.**

Methods **This was an observational study of cases with a final diagnosis of rAAA assessed at nine Emergency Departments and managed at one of two regional vascular centres in the UK.**

Results **Eighty-five consecutive cases were included. Seventeen [20.0%, 95% confidence interval (CI) 11.5–28.5%] patients reported important symptoms up to 3 weeks before index presentation. In the Emergency Department, most patients complained of abdominal and/or back pain, seven (8.2%, 95% CI 2.4–14.0%) additionally reported atypical pain and ten (11.8%, 95% CI 4.9–18.7%) denied pain altogether. Hypotension (36.5%, 95% CI 26.3–46.7%), tachycardia (18.8%, 95% CI 10.5–27.1%) and syncope (36.5%, 95% CI 26.3–46.7%) were documented in a minority of cases. Distracting symptoms were present in 33 (38.8%, 95% CI 28.4–49.2%) patients. The median time to diagnosis was 17.5 min (range immediate–12 days), and 21 (25.6%, 95% CI 16.3–34.9%) patients were misdiagnosed during clinical assessment.**

Conclusion **The classical signs and symptoms or rAAA are not always present and patients frequently show additional features that may confound the diagnosis. A high level of suspicion should be adopted for rAAA alongside a low threshold for immediate computed tomography. Further research is required to develop an objective clinical risk score or predictive tool for characterizing patients at risk.** ***European Journal of Emergency Medicine*** **00:000–000**

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Introduction

The prevalence of abdominal aortic aneurysm (AAA) in European men aged more than 65 is around 1.5% [1,2]. Although operative mortality may be as low as 37% [3], it is likely that many patients die before reaching definitive surgical intervention. Randomized trials (IMPROVE and AJAX) have not shown a definitive benefit to endovascular as opposed to open surgical strategy [4–6], but have reported the importance of logistics for patients with ruptured abdominal aortic aneurysm (rAAA) including efficient out-of-hours working, the threshold for permissive hypotension [7], the availability of endovascular aneurysm repair under local anaesthesia and efficient identification and transport of cases with rAAA to specialist centres [8].

The clinical presentation of rAAA is classically described as a triad of abdominal and/or back pain, hypotension and an expansile abdominal mass [9]. However, patients with rAAA may also present atypically with isolated symptoms commonly encountered in the Emergency Department (ED), for example, lower back pain or syncope [10]. An expansile abdominal mass is frequently not detected, with one analysis of pooled datasets suggesting that the positive predictive value of clinical examination for identifying AAAs was only 43% [11]. One study of 243 patients with known AAAs found that only 23% were palpable, even when the assessing clinician knew the diagnosis [12]. AAAs are particularly less likely to be detected on clinical examination in obese patients [13]. Atypical rAAA presentations reported in the published literature include transient lower limb paralysis [14,15], unilateral leg swelling [16], testicular ecchymosis [17,18], iliofemoral venous thrombosis [19], inguinoscrotal mass [20], phlegmasia cerulea dolens (lower limb pain, swelling and cyanosis) [21] and even obstructive jaundice [22]. The infrequent and varied presentation of rAAA may lead to misdiagnosis in-between 16 and 62% of cases [23–28]. Misdiagnosis of rAAA might contribute towards treatment delay or influence operative survival, and more research is required to describe the factors associated with accurate clinical diagnosis of this surgical emergency. Data from the AJAX Trial (Amsterdam Acute Aneurysm Trial) suggest that improvements in rAAA logistics, including centralization and preoperative planning, are key to optimizing outcomes [4].

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This two-network retrospective observational study aimed to describe the spectrum of rAAA presentation in the UK and to identify factors contributing towards misdiagnosis.

Methods

Setting

Cases were identified from two UK regional vascular centres that received transfers from seven general hospitals as well as referrals through their own EDs. The two vascular centres were the University Hospital Coventry & Warwickshire in Coventry and St George's Hospital in London. University Hospital Coventry & Warwickshire has 1116 beds [29] and acts as the vascular hub for Warwick Hospital (Warwick) and the George Eliot Hospital (Nuneaton). St George's Hospital has 914 beds [29] and provides vascular services for Kingston Hospital (Surrey), Croydon University Hospital (Croydon), St Helier Hospital (Carshalton) and East Surrey Hospital (Redhill). Both vascular centres had computed tomographic scanning facilities within their EDs during the study period.

Case selection

Inclusion criteria were any patient assessed between 1 December 2011 and 31 June 2012 and coded at discharge with a primary ICD-10 diagnosis of I71.3 (AAA, ruptured). Final diagnosis was made on the basis of radiological imaging, intraoperative and/or post-mortem findings. Data were extracted and recorded onto a structured proforma from case notes, imaging software and electronic patient records. Extracted fields included demographic details (age, sex, known AAA), signs and symptoms (pain distribution, vital signs, palpable AAA), mode of transport, details of index and subsequent clinical assessments (time, grade and specialty of doctor, investigations requested, provisional diagnosis) and outcomes (perioperative mortality, in-hospital mortality, 30 and 60-day mortality).

Data extraction

ED notes from both the initial presenting hospital and the vascular referral centre were accessed for all patients. A small pilot sample was used to design a standard proforma that recorded details that were consistently available for most cases in the cohort. Three authors (D.M., K.S. and S.T.) then independently extracted data onto a standard proforma and disagreements were resolved through discussion.

Definitions

Patients had a 'known AAA' if an AAA had been identified previously (e.g. incidentally or on screening), whether or not this was apparent to the assessing clinician. 'Altered consciousness' was defined as a recent history of syncope or reduced Glasgow Coma Score. Hypotension was defined as systolic blood pressure 90 mmHg or less (as this was commonly used as the threshold for cardiovascular 'shock' [30]) and tachycardia as a heart rate at least 100 beats per minute. BMI was calculated using height and weight from recent hospital episodes or from observations recorded on the ward in the day immediately following surgery. A palpable AAA was assumed to be present if documented following a clinical examination by a clinician of any grade or specialty.

The 'first clinician' was defined as the doctor performing a full clinical assessment of the patient at index presentation, in most cases the ED doctor. Brief triage assessments by nursing staff or paramedics were excluded. The diagnosis of rAAA was made when documented either as the primary diagnosis or as a differential to be excluded, for example, with ultrasound or computed tomographic scanning. In the binary logistic regression equations, cardiovascular instability was defined as hypotension and/or tachycardia and/or loss of consciousness (LOC), and nonspecialist clinician as anyone not occupying a consultant or a senior training grade (e.g. registrar) in vascular surgery or emergency medicine.

Statistical analysis

Continuous data were described as a mean with SD and 95% confidence intervals (CIs) if normally distributed and otherwise as a median with range and interquartile ranges (IQR). Missing data were managed by pairwise deletion. Binary logistic regression models were created to predict the correct initial diagnosis. Backward selection was used and inclusion in the final model required a significance level of $\alpha = 0.1$. Significant results were reported at $\alpha = 0.05$. All statistical analyses were carried out using SPSS, v22 (IBM Corp., Armonk, New York, USA).

Results

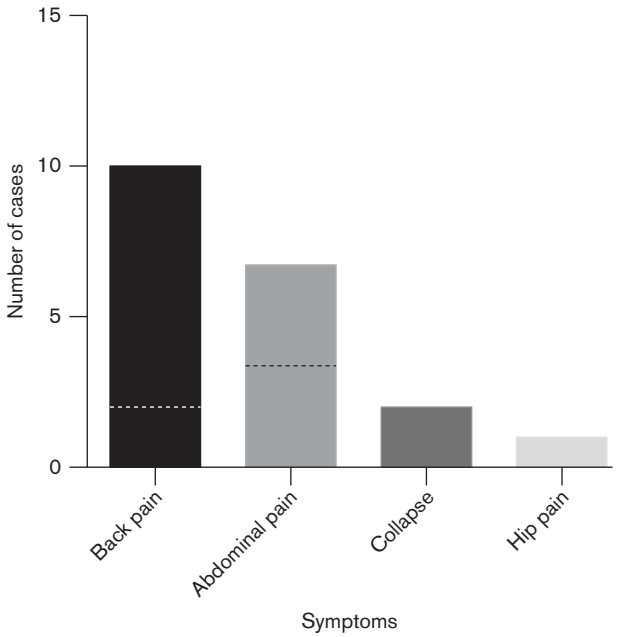
Eighty-five patients were identified as having an rAAA during the study period. The median age of the patients was 76.0 years (range 60.0–97.0, IQR 13.0), the majority of patients (82.4%, 95% CI 74.3–90.5%) were men and the median BMI was 27.0 kg/m^2 (range 18.0–37.0, IQR 5.8).

All case notes included an assessment of history, examination findings, vital signs, provisional diagnosis and management plan. It was possible to follow up all patients using discharge summaries, medical notes and bereavement office records.

Presentation

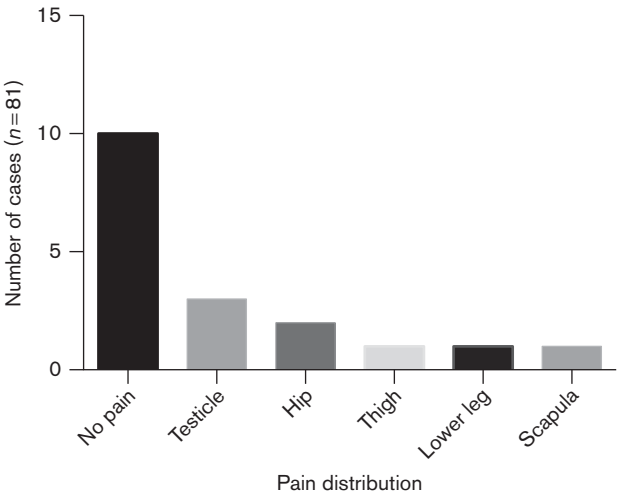
Seventeen (20.0%, 95% CI 11.5–28.5%) patients explicitly reported symptoms potentially attributable to AAA at least 1 day before their index presentation, which are presented in Fig. 1. Symptom duration ranged from 1 day to 3 weeks (mean 5 days), with five patients seeking medical attention during this time. Four saw their GP and one presented to both an ED and a fracture clinic after injuring their ankle during an unwitnessed collapse. In this group, five (29.4%,

Fig. 1



A bar chart showing the number of cases presenting with features likely attributable to a rAAA at least 1 day before final presentation. Hatched lines indicate the proportion of patients in each group who were previously known to have a AAA. AAA, abdominal aortic aneurysm; rAAA, ruptured abdominal aortic aneurysm.

Fig. 2



A bar chart showing the number of cases without pain or pain in an atypical distribution.

95% CI 19.7–39.1%) patients had a previously diagnosed AAA.

At ultimate presentation to the ED, 52 (61.2%, 95% CI 50.8–71.6%) patients complained of abdominal pain, 46 (54.1%, 95% CI 43.5–64.7%) complained of back pain, ten (11.8%, 95% CI 4.9–18.7%) complained of groin pain, and four (4.7%, 95% CI 0.2–9.2%) complained of loin pain. The combination of abdominal and back pain was only reported in 25 (29.4%, 95% CI 19.7–39.1%) cases. Although seven (8.2%, 95% CI 2.4–14.0%) patients complained of atypically distributed pain (Fig. 2), none complained of atypically distributed pain in isolation – all had abdominal and/or back pain in addition. Ten (11.8%, 95% CI 4.9–18.7%) patients denied having any pain at all. Although these patients were all able to communicate during clinical assessment, seven (70.0%, 95% CI 41.6–98.4%) had a recent history of syncope and/or LOC in the ED, which was much higher than the equivalent proportion in those reporting pain (32.0%, 95% CI 21.4–42.6%, Fisher's exact test $P = 0.005$). Only 21% (95% CI 12.3–29.7%) presented with the complete triad of back or abdominal pain, hypotension and a palpable mass.

LOC was recorded in 31 (36.5%, 95% CI 26.3–46.7%) cases and did not feature at any stage in 54 (63.5%, 95% CI 53.3–73.7) cases. Hypotension was present in 32 (37.6%, 95% CI 27.3–47.9%) cases and absent in 53 (62.4%, 95% CI 52.1–72.7%) cases. Similarly, only 16 (18.8%, 95% CI 10.5–27.1%) patients were tachycardic, with 69 (81.2%, 95% CI 72.9–89.5%) patients maintaining a normal heart rate. In total, 30 (35.3%, 95% CI 25.1–45.5%) patients showed no features of cardiovascular instability (tachycardia, hypotension or LOC) at any stage.

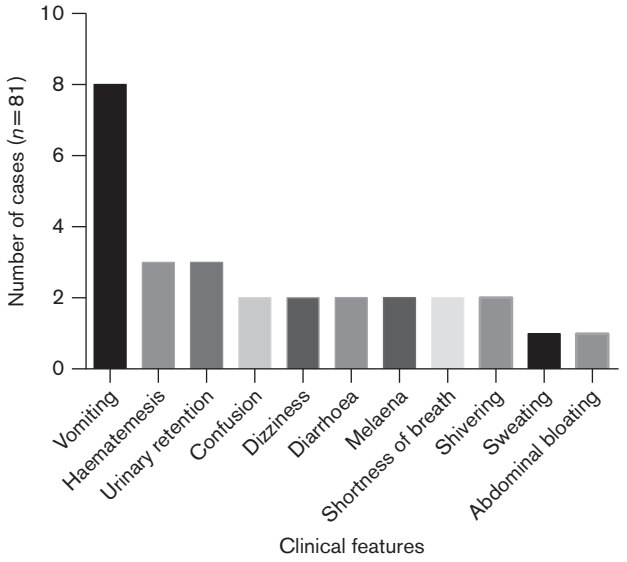
Potentially distracting symptoms were present in 33 (38.8%, 95% CI 28.4–49.2%) cases. Fifteen (17.6%, 95% CI 9.5–25.7%) had gastrointestinal symptoms that risked directing clinicians erroneously towards an alternative cause for abdominal/back pain. These symptoms are shown in Fig. 3. A palpable AAA was documented in 56 (70.0%, 95% CI 60.3–79.8%) cases.

Diagnosis

The median time to diagnosis from arrival in the ED was 17.5 min (range 0 min–12 days, IQR 126 min). Twenty-one (25.6%, 95% CI 16.3–34.9%) cases were not initially recognized as rAAA by the first clinician performing a full assessment. Alternative provisional diagnoses included diverticulitis, renal colic, nonspecific abdominal pain, appendicitis, intestinal obstruction, testicular torsion, pancreatitis, pneumonia, upper gastrointestinal bleed, vasovagal collapse, urinary tract infection and sepsis of unknown source. Diverticulitis and renal colic each featured twice as misdiagnoses.

In univariate analyses, the likelihood of the first assessing clinician accurately diagnosing rAAA was not significantly associated with collapse ($P = 0.203$), cardiovascular instability ($P = 0.998$) or known AAA ($P = 0.388$). Similarly, no particular distribution of pain (e.g. back pain) was significantly associated with making a correct

Fig. 3



A bar chart showing the frequency with which patients complained of features that might indicate a non-rAAA cause for their presentation. rAAA, ruptured abdominal aortic aneurysm.

initial diagnosis. The presence of a palpable AAA was associated with the correct initial diagnosis ($P = 0.037$). A logistic regression analysis showed that only the presence of a palpable AAA (odds ratio 3.3, 95% CI 1.1–9.4, $P = 0.029$) and collapse (odds ratio 3.2, 95% CI 1.0–10.0, $P = 0.042$) were independent predictors of the correct diagnosis of rAAA being made by the first assessor.

Discussion

This study confirms previous findings that rAAA is commonly misdiagnosed across a range of settings [23–28]. rAAAs were mistaken for 12 different diseases in this series. Over a quarter of cases were initially misdiagnosed and one patient was admitted with an incorrect diagnosis for 12 days. However, this level of misdiagnosis is comparable with ED data from the USA, where 30% of rAAA patients are not recognized on initial assessment [24].

A small subgroup (20%, 95% CI 11.5–28.5%) had symptoms potentially attributable to AAA in the days before their index presentation. The only other study to report interval between symptom onset and presentation suggested that 43% of rAAA patients develop potentially relevant features at least 9 h before admission [26]. Although clinicians are taught that rAAA presents as a triad of abdominal/back pain with hypotension and an expansile abdominal mass [9], this study showed that many rAAAs presented nonclassically. In our study, the only variables associated with correct eventual diagnosis were collapse on admission or the presence of a palpable AAA. Importantly, 38.8% (95% CI 28.4–49.2%) showed additional features that potentially confounded the diagnosis. These varied clinical presentations are a feature of rAAA pathoanatomy. Misdiagnosis can often arise from the variable compressive effect of a retroperitoneal haematoma. For example, testicular pain might be caused by compression of the ilioinguinal and/or genitofemoral nerves [31]. Although some patients reported no pain, this group had a significantly higher rate of LOC than those with pain (70.0 vs. 32.0%, Fisher's exact test $P = 0.005$), which raises the possibility of cognitive impairment caused by hypovolaemic compromise in this group. Other potentially distracting symptoms (e.g. vomiting and hyperventilation) were also consistent with haemodynamic shock. This complex clinical picture is particularly important in the context of the recent IMPROVE trial, which showed that most rAAAs initially present to nonvascular centres (60.3%) and outside of normal working hours (64.9%) [6]. These findings suggest that most rAAAs will first be assessed by a relatively inexperienced clinician in a nonspecialist centre.

No significant association was found between initial misdiagnosis and patient mortality. Despite this potentially being because of a limited cohort size, the finding is consistent with other retrospective series from the USA [24] and Europe [26]. Selection bias may underlie this observation because rAAA becomes more obvious in deteriorating patients, leading to the selection of more stable patients for retrospective in-hospital studies. This is a particular limitation of our study because patients who died in peripheral hospitals (some presumably undiagnosed) would not have been transferred to a specialist centre and so are absent from our cohort. Although markers of cardiovascular instability (tachycardia and hypotension) did not influence diagnosis in our series, others have shown that haemodynamically stable rAAAs are less likely to be recognized promptly [23]. It is likely that prompt diagnosis and intervention could improve outcomes across the population of patients with rAAA.

The demographic profile of patients in this series is consistent with the risk factors identified elsewhere [6,10]. Although smoking status and family history were not routinely documented, 82.4% (95% CI 74.3–90.5%) of patients were men older than 60 years of age. In addition, our study found a higher prevalence of palpable AAAs (70%, 95% CI 60.3–79.7%) than has been reported previously [11]. These findings emphasize that AAA should be considered in all older men presenting acutely with a wide range of symptoms, from hip pain to melaena. The frequency with which bedside ultrasound is used to identify AAAs might be increased by greater involvement of trained ED doctors in this noninvasive and readily available imaging technique. However, the critical conclusion is that EDs should adopt a low threshold for immediate computed tomography, which remains the gold standard for the diagnosis of rAAA.

Conclusion

rAAA is a frequently fatal surgical emergency that results in mortality if it is missed or misdiagnosed. This multicentre retrospective cohort study confirmed that the cardinal triad of features was only reported by a minority (21%, 95% CI 12.3–29.7%) of patients presenting with rAAA. Clinicians should therefore adopt a low threshold of suspicion for rAAA and urgent diagnostic imaging. Further research is necessary to characterize the relationship between misdiagnosis and patient outcomes and to develop an objective clinical risk score for improving the diagnosis of this important emergency.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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