

Inflammatory Abdominal Aortic Aneurysm (IAAA)



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PURPOSE: *The aim of this study is to report our experience about the inflammatory abdominal aortic aneurysm (IAAA).*

METHODS: *Between January 1999 and January 2008 we treated 8 cases of IAAA. Two patients underwent surgery in emergency. The preoperative diagnostic procedure were ultrasound (US), computed tomography (CT) and intravenous urography (IVU). In 6 elective patients the diagnosis of IAAA was obtained preoperatively. In one case a left hydronephrosis was demonstrated by intravenous urography (IVU). All patients underwent open surgery with mid-line incision and transperitoneal access.*

RESULTS: *No 30-days mortality occurred. A case of pancreatitis was treated with conservative therapy. All patients had 60-days corticosteroid therapy.*

CONCLUSIONS: *Our data suggest that because IAAA have the same rate of rupture of AAA, they need the same preventive treatment as non inflammatory abdominal aortic aneurysm (AAA). The kind of approach OPEN-EVAR should be chosen with the same criteria as AAA, even if EVAR treatment doesn't allow us to obtain the biopsy. Furthermore there are no sufficient evidences about regression of retroperitoneal fibrosis after EVAR treatment. Also the premature onset should be considered in the choice of treatment.*

KEY WORDS: Aneurysm; Inflammatory; Open repair.

Introduction

In 1972 Walker defined the Inflammatory Abdominal Aortic Aneurysm (IAAA) as: 'thick, firm, smooth wall with a shiny white appearance' where 'dense fibrosis extended to involve adjacent structures' ¹.

The aim of this study is to report our experience of 8 cases of IAAA observed between January 1999 and January 2008. Currently the IAAA is identified "by the presence of a thickened aneurysm wall, marked perianeurysmal and retroperitoneal fibrosis and dense adhesions of adjacent abdominal organs" ²⁻⁴. The IAAAs rep-

resent the 3-10% of all Abdominal Aortic Aneurysms (AAAs) ¹⁻¹¹. Autopsic study confirmed its presence in 1-6%¹². The EUROSTAR registry database reports a prevalence of 1.4% in 3665 AAA ¹³.

The Registry of the Italian Vascular and Endovascular Surgery Society (SICVEREG) reports 2.3% IAAA in 3578 AAA in the Italian population ¹⁴.

Materials and Methods

Between January 1999 and January 2008 we treated eight cases of IAAA (representing 3.1 % of the AAA observed in the same period). Six were in election and two in emergency. Mean age was 73 (66-84) years, seven patients were male and one female. Mean transverse diameter was 6.8 cm (min 5.2 e max 9.0). (Tab I)

The preoperative diagnostic procedures were ultrasound (US), computed tomography (CT) and intravenous urog-

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Fig. 1

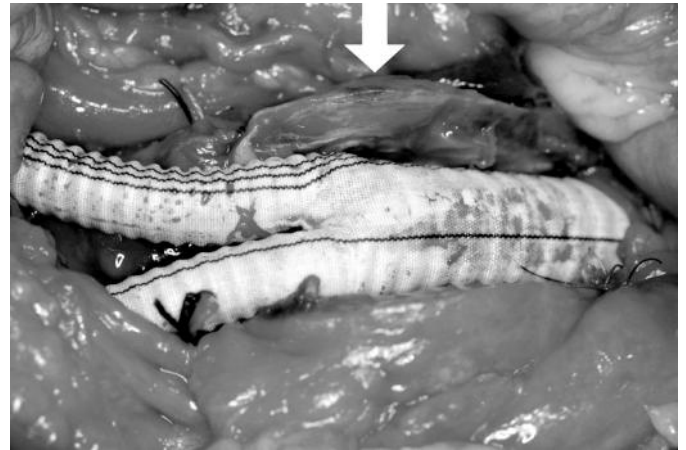


Fig. 3

TABLE I - Preoperative Data

Patients n.	1	2	3	4	5	6
E/U	E	E	E	E	U	U
CT	+	+	+	+	-	-
Abd. Pain	+	+	++	+	++	+++
Fever	+	-	+	-	+	+
ESR	+	+	+	+	+	+

E: election; U:urgency; CT: computed tomography; Abd Pain: abdominal pain: +/++/+++ mild/moderate/severe; ESR: erythrocyte sedimentation rate.



Fig. 2

raphy (IVU). CT-scan was specific for elective cases showing a periaortic inflammatory cuff. (Fig. 1) IVU demonstrated a left hydronephrosis. (Fig. 2) Preoperative findings were abdominal pain in all patients (two patients had severe pain); fever and an elevated erythrocyte sedimentation rate (ESR) were detected in all patients. (Tab. I)



Fig. 4

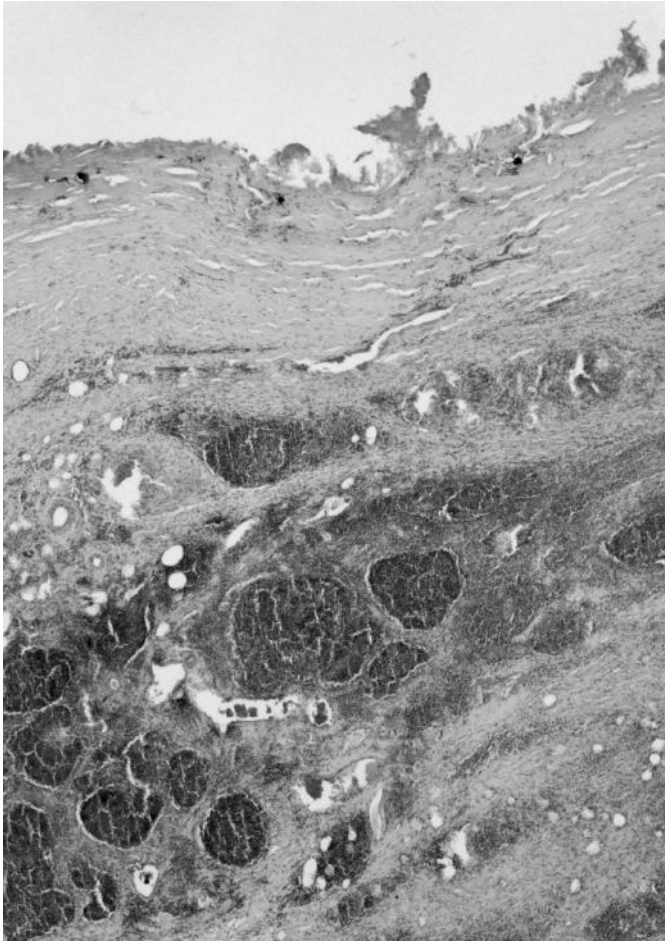


Fig. 5

Results

Macroscopic findings of IAAA were a thick, white/gray, inflammatory wall involving fourth duodenal portion (Fig. 4). In all cases was present an inflammatory component of retroperitoneal tissue that in one case involved the left ureter.

No 30-days mortality occurred. One patient experienced acute oedematous pancreatitis, treated with conservative therapy; no other major complication occurred. Istological examination of the aneurysmatic wall demonstrated the presence of lymphocytic and monocytic infiltration (Fig 5).

Hydroureteronephrosis was treated with conservative therapy. Complete resolution of abdominal pain, fever and ESR was seen in all cases (Tab. II).

We had a case of late mortality for chronic renal failure not linked to surgery.

Discussion

Controversy exists about the real pathogenesis of IAAA but it shows a predilection for male,^{2,3,8,15,16} and current smoke⁹. It present also a familial tendency⁹ and a

TABLE II - Outcomes 12 months

Patients n.	1	2	3	4	5	6
Death	—	—	—	—	—	—
Abd. Pain	—	—	—	—	—	—
Fever	—	—	—	—	—	—
ESR	—	—	—	—	—	—

Abd Pain: abdominal pain; ESR: erythro sedimentation rate.

premature incidence (5-10 years) in comparison with non inflammatory AAA¹⁷.

Lymphocytes (T and B), macrophages and a relevant fibrous component are the characteristics of intra and extra-parietal flogistic infiltrate¹⁸⁻²⁰.

Actually still remain different hypothesis for IAAA development. In particular these are vasculitic²¹, infective (herpes simplex, citomegalovirus, chlamydia, mycoplasma, Salmonella)²²⁻²⁴, auto-immune (local and systemic)²⁵ and genetic¹⁷ factors are considered.

Current understanding favours for an immuno-mediated hypothesis²⁶, based on the theory that Elastin Degradation Products (EDP), produced by metalloproteinases, can cause an immune reaction subsequently alimanted first by macrophages and then by lymphocytes activity²⁷⁻²⁹.

Recently the presence of a similar parietal phlogistic infiltration both in AAA and IAAA has been demonstrated. Several Authors have hypothesized the possibility of a single pathogenetic process with different modality of expression¹⁸.

The parietal flogistic process can involve adjacent structures, in particular ureters, duodenum and major retroperitoneal veins^{30,31}.

Clinical findings contribute to differentiate IAAA from non-inflammatory AAA. Abdominal back pain, fever, weight loss, are more frequent symptoms (93%)⁹ then those linked to hydroureteronephrosis, aorto-enteric fistula or deep venous thrombosis (9%)^{31,32}.

In our experience emergency surgery was required in 25% of cases (two cases). Literature reports rupture rate of IAAA similar to AAA (3-14%), so IAAA needs the same preventive treatment of AAA^{3,4,9,11,31,33-35}.

Helpful preoperative diagnostic procedure are US, CT, magnetic resonance imaging (MRI), intravenous urography (IVU) and Positron Emission Tomography (PET). Walker CT triad represented by inflammatory cuff around the aorta, the parietal thockening and the infiltration of adjacent structure, allow preoperative diagnosis. CT has a sensibility of 83%, specificity 99% and overall accuracy 93.7%^{36,37}.

More complex is the diagnosis in course of aneurysmatic rupture where the retroperitoneal tissue planes become poorly defined and can make the identification of an inflammatory component difficult.

Other diagnostic pitfall is retroperitoneal sarcomatous

tumor^{11,38,39}. Metabolic activity of IAAA allow the application of PET^{40,41}.

Although the IAAA presents a challenge to the surgeon, recent series show that mortality and morbidity rates are similar to AAA; these remarks are valid also for late survival^{3,9,11,12,33,34}. We did not register mortality at 12 months follow-up.

Open surgery produces regression of retroperitoneal fibrosis in 68.3-75% of cases^{8,42}.

Because of technical difficulty of open IAAA repair, endovascular aneurysm repair (EVAR) was proposed as an attractive option¹¹.

EUROSTAR data demonstrate the reliability of EVAR repair, in terms of short outcomes, with results similar to AAA. Nevertheless this approach doesn't allow the biopsy which may confirm the diagnosis^{11,13,43-47}.

A further limit of EVAR is the actual absence of consistent data concerning the regression of the fibrosis^{11,13}.

Ureters involvement ranges from 24.6 to 32% of cases of IAAA with bilateral presentation in 50%.

Although different treatment are described (stenting, ureterolysis, nephrostomy) a conservative approach should be maintained wherever possibile, in consideration of favourable evolution of retroperitoneal fibrosis after IAAA open treatment^{30,31}.

There are no evidences concerning the potential efficacy of the antiphlogistic therapy in the management of IAAA. In our experience we delivered empiric steroid therapy at low dose for at least 60 days. Other experience with immunosuppressant or antibiotics showed no significant results^{9,11,48}.

Conclusions

The IAAA is a relatively frequent entity. There are no evidence about its real pathogenesis. Currently AAA and IAAA are considered as a single pathogenetic process with different modality of expression. IAAA, having the same rupture rate of AAA, needs the same preventive treatment. Open repair or EVAR should be chosen with the same criteria of AAA even if there are no data about regression of retroperitoneal fibrosis after EVAR.

Riassunto

OBIETTIVO: Lo scopo di questo studio è di riportare la nostra esperienza circa gli aneurismi infiammatori dell'aorta addominale (IAAA).

MATERIALI E METODI: Nel periodo compreso tra gennaio 1999 e gennaio 2008 sono stati trattati otto casi di IAAA. Due pazienti sono stati sottoposti ad intervento chirurgico in urgenza. Le procedure diagnostiche preoperatorie sono state gli ultrasuoni (US), tomografia computerizzata (CT) ed urografia intravenosa (IVU). In sei pazienti (trattati in elezione) è stato possibile porre diagnosi di

IAAA. In un caso è stata diagnosticata idroureteronefrosi sinistra con urografia intravenosa. Tutti i pazienti sono stati trattati con chirurgia tradizionale open con incisione mediana ed accesso trans-peritoneale.

RISULTATI: Non è stata registrata mortalità perioperatoria. Il follow-up ha evidenziato un caso di pancreatite edematosa trattata con terapia medica. Tutti i pazienti sono stati trattati con terapia corticosteroidica empirica per un periodo di 60 giorni.

CONCLUSIONI: I nostri dati suggeriscono che l'IAAA, avendo lo stesso rischio di rottura degli aneurismi dell'aorta addominale aterosclerotici, necessita dello stesso trattamento preventivo. La scelta circa l'approccio chirurgico tradizionale od endovascolare (EVAR) dovrebbe essere scelto con gli stessi criteri che guidano la scelta nel trattamento degli aneurismi aterosclerotici. La scelta del trattamento EVAR comunque non permette alcun prelievo biotico e quindi la diagnosi di certezza. Inoltre non ci sono evidenze circa la regressione della fibrosi retroperitoneale associata all'IAAA dopo trattamento EVAR. Anche la precoce insorgenza dell'IAAA rispetto all'aneurisma aterosclerotico dovrebbe rappresentare un parametro significativo nella scelta del trattamento.

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