Summary of cases of aortic dissection associated with giant cell arteritis reported in the literature

<table>
<thead>
<tr>
<th>Source</th>
<th>Number of cases</th>
<th>Age (yr)</th>
<th>Sex</th>
<th>Hypertension</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Broch 1947</td>
<td>1</td>
<td>68</td>
<td>F</td>
<td>+</td>
<td>First recorded case (case report) Series of 248 cases: 93 sudden deaths; diagnosed at postmortem examination</td>
</tr>
<tr>
<td>Klein 1953</td>
<td>3</td>
<td>66</td>
<td>F</td>
<td>NK</td>
<td>Postmortem series of 161 acute dissections; ‘healed aortitis’ found in both Case report</td>
</tr>
<tr>
<td>Larson 1984</td>
<td>2</td>
<td>NK</td>
<td>F</td>
<td>NK</td>
<td>Known GCA. Sudden death (case report) Postmortem series of 9 sudden deaths in GCA</td>
</tr>
<tr>
<td>Leonard 1979</td>
<td>2</td>
<td>62</td>
<td>F</td>
<td>FK</td>
<td>Known GCA. Sudden death (case report) Postmortem series of 171 acute dissections</td>
</tr>
<tr>
<td>BMJ 1960</td>
<td>1</td>
<td>70</td>
<td>M</td>
<td>+</td>
<td>Sudden death (case report) Postmortem examination (case report)</td>
</tr>
<tr>
<td>Save-Soderberg 1982</td>
<td>2</td>
<td>85</td>
<td>F</td>
<td>+</td>
<td>Sudden death (case report) Postmortem examination (case report)</td>
</tr>
<tr>
<td>Nordt 1989</td>
<td>1</td>
<td>70</td>
<td>F</td>
<td>-</td>
<td>Acute myocardial infarction caused by dissection; died (case report)</td>
</tr>
<tr>
<td>N Y State J Med 1985</td>
<td>1</td>
<td>77</td>
<td>F</td>
<td>+</td>
<td>Sudden death (case report) Postmortem examination (case report)</td>
</tr>
<tr>
<td>Ainsworth 1961</td>
<td>1</td>
<td>83</td>
<td>M</td>
<td>-</td>
<td>Survival following aortic graft (case report)</td>
</tr>
<tr>
<td>Harris 1968</td>
<td>2</td>
<td>70</td>
<td>F</td>
<td>+</td>
<td>Survival following aortic graft 14 years (case report) 99 postmortem examinations; 2 dissections among 15 GCA cases found</td>
</tr>
<tr>
<td>Ostberg 1972</td>
<td>2</td>
<td>NK</td>
<td>F</td>
<td>+</td>
<td>Both died; case reports</td>
</tr>
<tr>
<td>Muri 1980</td>
<td>2</td>
<td>-</td>
<td>F</td>
<td>NK</td>
<td>Survival following aortic dissection 14 years (case report) 99 postmortem examinations; 2 dissections among 15 GCA cases found</td>
</tr>
<tr>
<td>Evans 1994</td>
<td>16</td>
<td>M</td>
<td>LHV</td>
<td></td>
<td>Survival on steroids alone for 2 years</td>
</tr>
<tr>
<td>This paper</td>
<td>2</td>
<td>68</td>
<td>M</td>
<td>LHV</td>
<td>Survival on steroids alone for 3 years</td>
</tr>
<tr>
<td>Total cases</td>
<td>43</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

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Discussion

Giant cell arteritis is distinguished from Takayasu’s disease on clinical and pathologic grounds. Medical specialties are usually affected, although 15% of a large series were found to have large vessel involvement. Despite the characteristic histopathological findings of mononuclear cell infiltration, disruption of the internal elastic lamina and the presence of giant cells which would be expected to reduce the elasticity and tissue strength of large arteries, aortic dissection is an unusual complication (table). The first case was reported from Norway in 1947. A postmortem study of 111 cases of dissecting aortic aneurysm from western Japan did not implicate giant cell arteritis as a cause in any case, and 161 postmortem cases of dissecting aortic aneurysm from the Mayo Clinic also failed to implicate giant cell arteritis with certainty, though two had ‘healed aortitis’. Klein describes three cases of aortic dissection in giant cell arteritis that were fatal, one in the descending and two in the ascending aorta. Of 125 postmortem cases seen in Manchester, England, two had evidence of giant cell arteritis. The usual cause of death is aortic rupture, though, old, healed dissection may be an incidental finding. In a postmortem series of nine deaths related to giant cell arteritis, Save-Soderberg et al mentioned two elderly hypertensive females with aortic dissection, at least one of whom appears to have had a descending aortic lesion.

The first reported case diagnosed and who survived for some months was described in Ipswich. One previous case from our hospital did well after aortic resection and is still alive and symptom free 14 years later. In two other reports, two of 16 postmortem cases of giant cell arteritis died of aortic dissection, and two further fatal cases in women have been described from Japan.

Most of these patients showed evidence of hypertension in life or at postmortem examination. Twenty two of 27 reported cases were women – disproportionally 1:2 male:female ratio in giant cell arteritis. The gender of 16 cases with aortic dissection in the large series reported by Evans et al was not specified.

The diagnosis of acute aortic dissection may be made by four different imaging techniques. Retrograde aortography has a sensitivity of only 81–91%, though specificity may be more than 90%.11 It can demonstrate involvement of branch vessels and coronary arteries. CT has a sensitivity of 83–100% and a specificity of 90–100%. MRI has been shown to be up to 100% sensitive and specific in some studies. TOE may be 97–100% sensitive, but specificity as low as 68% has been reported, probably reflecting operator dependence.

The cases described had clinical evidence of giant cell arteritis, biopsy proven in two cases, and aortic dissection. The two presentations with fever and chest pain did not immediately suggest the diagnosis. Clinicians should be aware of potential life threatening large vessel disease in giant cell arteritis, particularly in female patients with hypertension. The outcome may be more favourable than previously believed, if the arteritis is adequately controlled with steroids. The diagnosis of giant cell arteritis should be considered in patients presenting with acute dissection of the aorta.

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Antibodies to collagens in sera from patients receiving bovine cartilage graft.

The production of autoantibodies against different collagen types has been described in association with several rheumatic diseases and experimental pathological entities. Antibodies to collagen type II have been demonstrated in relapsing polyarthritis1 and in rheumatoid arthritis.2

Cartilage grafts for tissue reconstruction have been used in many surgical specialties, especially in otorhinolaryngology and maxillofacial surgery. Autoantibody cartilage has the tendency to bracken. Furthermore, harvest of the material requires an additional operative procedure. Recently, we observed humoral reactivities against cartilage temporomandibular joint cartilage, a nasal contour reconstruction with allo- or autologous cartilage grafts.3 These


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patients showed resorption or rejection of the transplanted cartilage graft. Serum antibodies against collagen type IX and XI were found. However, there is no clear evidence for a causal role of these antibodies for the failure of the procedure.

More than 40 years ago, chemically preserved bovine cartilage was first used as an implant material, but absorption was a major problem; the combination of radiation and chemical preservation of bovine cartilage allowed Ersek al subsequently to obtain better results in animals. In 1988, the same authors reported their clinical experience with 53 patients using glutaraldehyde processed irradiated bovine cartilage for reconstruction. In 94% of the patients the results were successful, with no change in graft size or shape during an average follow up period of four years. They concluded that these processed cartilage grafts remained immunologically inert.

We have tested the presence of autoantibodies against collagen in a group of 44 patients who had received a graft of bovine cartilage that had been processed with both radiation and glutaraldehyde. These patients had undergone a maxillofacial reconstruction procedure between 1987 and 1992 in the Department of Oral and Maxillofacial Surgery of the University of Erlangen. There were 17 women with an average age of 31 years (range: 15–58), and 27 men with an average age of 37 years (range 13–72). None of the patients who underwent transplant surgery showed rejection or absorption problems. Sera were obtained at the end of a variable follow up period that lasted between one and six years.

Enzyme linked immunosorbent assays using collagen types I, II, III, IX, and XI as antigens were performed as described previously. Sera of the 44 patients and seven healthy donors were tested in serial dilutions. The optical densities were plotted in a coordinate system. A logarithmic regression curve through the values obtained for each patient was extrapolated to the y axis. As shown in the figure, the reactivity to all collagen types was significantly stronger in the patients compared with healthy donors; however, the difference was most pronounced with collagen type IX. When the titres for type IX collagen were compared with the titres of any other anticollagen reactions within the transplanted patient group, the significance of the difference was p < 0.05.

It may be concluded that production of autoantibodies, at least to collagen type IX, takes place in patients receiving chemically processed, irradiated bovine cartilage. These results and earlier studies with autologous and allogeneic grafts suggest that collagen type IX is the major antigen in humoral immune responses against cartilage grafts. Collagen type IX is known to be important for collagen fibril growth in cartilage, and is covalently linked to the surface of collagen type II fibrils; it is therefore probably far more exposed to the immune responses than the major cartilage collagen, type II.

As the surgical procedures in all the cases presented were successful, we believe that humoral immunity alone does not explain the rejections seen in inflammatory cartilage lesions. One well studied problem with xenogenic implants is the hyperacute rejection that results from the recipient's natural antibodies reacting against the donor's endothelial cells. The observed tolerance could be explained in part by the fact that these grafts have no cells at all, and that the radiochemical treatment of the cartilage matrix destroys some key epitopes, or at least renders them inaccessible. The lack of graft rejection despite a strong humoral immune response indicates that cellular immunity may have a major role in causing the unsuccessful cases of cartilage grafting in other disorders.

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