Large vessel arteritis: a diagnostic challenge in the elderly

Takayasu arteritis is a rare form of chronic inflammatory arteritis affecting large vessels, predominantly the aorta and its main branches. Those at highest risk are adolescent girls and women in their 2nd and 3rd decade of life, and this disease is most frequently seen in Japan, Southeast Asia, India, and Mexico. An elderly woman with Takayasu arteritis of late onset, an extremely unusual disorder in the Western world, is described. A 67 year old woman presented with a two month history of bilateral forearm pain associated with coldness, numbness, and stiffness. She had claudication symptoms in the upper extremities, asthenia, and loss of up to 15% of cases, and approximately 50% of cases have a negative temporal artery biopsy result. Normally both diseases can be differentiated on clinical grounds. The most

discriminatory factors that led to the correct diagnosis in 95% of patients are age of 40 years at disease onset, ethnic background, clinical signs of upper limb vascular insufficiency, shoulder stiffness, and scalp tenderness. The diagnosis is made by vascular imaging findings (proximal stenotic lesions predominate and tend to be bilateral) and American College of Rheumatology criteria (table 1). This patient fulfilled five of the six American College of Rheumatology specific diagnostic criteria. Vascular imaging is crucial in making the correct diagnosis in patients with large vessel giant cell arteritis. Aortitis does not lead to arterial obstruction, but leads to arterial dilatation and aneurysm formation, and most often affects the distal subclavian artery. A real diagnostic dilemma appears when, as in this patient, the clinical presentation is similar (aortic arch syndrome without cranial symptoms), laboratory findings are non-specific, temporal biopsy results are negative, and age is a confusing feature. In such cases, giant cell arteritis with large artery involvement and Takayasu arteritis of late onset have to be considered.

This case suggests that Takayasu arteritis may be underdiagnosed in the elderly in the Western world.

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References

Fibrocartilagenous mesenchymoma of bone: the youngest reported case in a patient aged 1 year and 7 months

We report the youngest recorded example of a fibrocartilagenous mesenchymoma of bone, with an age at presentation of 1 year and 7 months. The youngest previously recorded cases have been in 9 year old patients. A male infant aged 1 year and 10 months originally presented to the orthopaedic clinic with a three month history of limping and pain in the left leg. There was no history of trauma and there were no localising signs in the left leg. An x ray showed a lucent area with surrounding sclerosis involving the proximal left tibial metaphysis. He had originally been brought to the accident and emergency unit two days after the initial

Table 1 American College of Rheumatology criteria for the classification of Takayasu arteritis (1990)

<table>
<thead>
<tr>
<th>Criteria</th>
<th>Requirement</th>
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<tr>
<td>Age at disease onset &lt; 40 years</td>
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<tr>
<td>Claudication of extremities</td>
<td></td>
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<tr>
<td>Decreased brachial artery pulse</td>
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<tr>
<td>Blood pressure difference &gt; 10 mm Hg</td>
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<tr>
<td>Bruit over subclavian arteries or aorta</td>
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<tr>
<td>Arteriogram abnormality</td>
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</table>

At least three of the six criteria are required for the diagnosis of Takayasu arteritis.

Figure 1 Magnetic resonance angiography of the aortic arch showing total stenosis of both subclavian arteries (small arrows), and a significant filiform stenosis of the right vertebral artery, which had an inverted flow (large arrow).

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tumour that had a specific clinical behaviour, radiological features, and morphological features. The differential diagnosis of this lesion involves several entities, including chest wall hamartoma (mesenchymoma) and dedifferentiated chondrosarcoma. The most important differential diagnosis is fibrous dysplasia with cartilaginous differentiation. The tumour usually occurs in the metaphyses of the long bones and commonly presents with pain. Other sites have been documented, such as the metatarsal bones, vertebrae, and the pubis.

Radiologically, this tumour often abuts an open growth plate. These tumours are predominantly radiolucent, although some mineralisation is often seen. Cortical destruction is an additional common finding.

This lesion is generally treated by surgical resection. Local recurrences have been recorded. On review of the current limited literature, no distant spread has been recorded and there is no associated mortality. This entity is thought to represent a true neoplasm rather than a hamartoma.

There have been four previously published series describing fibrocartilagenous mesenchymoma of bone, comprising, in total, 20 patients. Our patient is more than 7 years younger than the youngest previously recorded case. The light microscopic features together with the clinical presentation and radiological appearance are characteristic. The most important histological feature is cartilage with the appearance of epiphysial plates.

Some cases recur locally, but no metastasis or death was caused by these lesions. Dahlin thought that this lesion was of low grade malignancy. A more recent series suggests that the term malignant is not warranted.

In summary, the average age of occurrence was 14 years. Therefore, our patient, who is more than 7 years younger than these individuals, is the youngest patient to be recorded with this diagnosis.

References

\[\gamma\text{-Glutamyltransferase decreases in patients with the chronic form of schistosomiasis mansoni treated with ursodeoxycholic acid}\]

Schistosomiasis mansoni is a chronic parasitic liver disease, characterised by a relative preservation of hepatic function, but there are early alterations in laboratory test results. The rise in serum \(\gamma\text{-glutamyltransferase (\(\gamma\text{GT}\))}\) activity in patients with schistosomiasis does not correlate with either parasite load or ultrasonographic alterations.

Eleven adults (nine men) with raised serum \(\gamma\text{GT}\) activity and the "pure" form of schistosomiasis participated in the prospective study (eutrophic patients; body mass index > 20 and < 30 kg/m\(^2\); abstemious or with alcohol ingestion less than 160 g/week; non-users of hepatoxic and/or cholestatic drugs; and hepatitis B surface antigen, anti-hepatitis C core antigen, and anti-hepatitis B core antibody negative). All patients were previously treated for the parasitosis. Echographic examinations were interpreted as described previously. Upper digestive endoscopy revealed the presence of oesophageal and/or oesophagogastroduodenal varices and/or hypertensive gastropathy. The patients were treated with ursodeoxycholic acid at a dose of 10 mg/kg/day, divided into two daily portions for a period of six weeks. Before treatment, during treatment, and 30–45 days after drug discontinuation, the patients underwent clinical and laboratory evaluation.

At the beginning of the treatment, \(\gamma\text{GT}\) was 3.1 (SEM, 0.4) times the upper normal limit, whereas alkaline phosphatase was normal (mean, 1.1 times the upper normal limit; SEM, 0.2). There was a progressive and significant decrease in \(\gamma\text{GT}\) to near normal values at the end of treatment and an increase again after drug discontinuation, whereas alkaline phosphatase remained unaltered. Aminotransferases also decreased during treatment and increased again after drug discontinuation (table 1). Prothrombin index (mean international normalised ratio at the beginning, 1.2; SEM, 0.1) and platelet count (mean, 96 × 10\(^3\)/litre at the beginning; SEM, 23) remained unaltered.

Ursodeoxycholic acid is widely used for the treatment of primary biliary cirrhosis and other chronic cholestatic liver diseases. We now report that the increased serum \(\gamma\text{GT}\) activity seen in patients with the chronic form of schistosomiasis is ursodeoxycholic acid sensitive. The mechanism of this increase in \(\gamma\text{GT}\) seems to be different from that which occurs in alcoholism, and is compatible with the existence of changes in the biliary tree.

Further studies are required to understand the role of ursodeoxycholic acid in the molecular and cellular mechanisms of bile secretion and its ability to reduce \(\gamma\text{GT}\) activity in schistosomiasis mansoni (for example whether it acts on the evolutionary stage of the disease), and to determine whether \(\gamma\text{GT}\) could be used as a prognostic marker for this disease.
Acknowledgements
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References

Table 1: Laboratory parameters in patients with schistosomiasis mansoni at the beginning and end of treatment with ursodeoxycholic acid and 30–45 days after discontinuation of the drug.

<table>
<thead>
<tr>
<th>Treatment period</th>
<th>Total bilirubin (mg/l)</th>
<th>AST (U/l)</th>
<th>ALT (U/l)</th>
<th>AP (U/l)</th>
<th>γGT (U/l)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Before</td>
<td>13 (1)</td>
<td>46 (5)</td>
<td>44 (4)</td>
<td>290 (47)</td>
<td>155 (19)</td>
</tr>
<tr>
<td>End</td>
<td>12 (1)</td>
<td>34 (2)</td>
<td>30 (4)</td>
<td>302 (46)</td>
<td>66 (8)</td>
</tr>
<tr>
<td>After</td>
<td>13 (1)</td>
<td>41 (6)</td>
<td>38 (4)</td>
<td>240 (42)</td>
<td>157 (27)</td>
</tr>
<tr>
<td>Paired t test</td>
<td>0.245</td>
<td>0.036</td>
<td>0.004</td>
<td>0.889</td>
<td>0.0003</td>
</tr>
<tr>
<td>(p &lt; 0.01)</td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Paired t test</td>
<td>0.755</td>
<td>0.402</td>
<td>0.810</td>
<td>0.093</td>
<td>0.649</td>
</tr>
<tr>
<td>(p &lt; 0.01)</td>
<td></td>
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</table>

Values are mean (SEM). ALT, alanine aminotransferase; AP, alkaline phosphatase; AST, aspartate aminotransferase; γGT, γ-glutamyltransferase.

Embryo and Fetal Pathology. Color Atlas with Ultrasound Correlation


This is a comprehensive and extremely well illustrated atlas with good quality colour photographs. The information provided is up to date and well presented. It has an excellent well illustrated section on embryonic development and good photos of growth disorganised embryos. Dating and staging of embryos should be straightforward using this atlas. There are helpful tables throughout the book that make for easy quick reference. Errors of morphogenesis are followed by malformation syndromes, dysplasia, and disruption. The chapter on malformation syndromes is well set out with practical tables of syndromes that refer to a specific malformation—such as, unilateral renal agenesis. Both dysplasias and disruptions are well discussed and illustrated.

A novel addition is the provision of colour diagrams of each operative procedure performed for the correction of congenital heart disease. A good list of likely complications for each operation is provided.

A strange concept provided under congenital tumours (page 548) is that Castleman disease and Takayasu disease are malformation syndromes with haemangiomases. In addition, several typographical errors were noted.

This book adds a new dimension to the currently available fetal and neonatal pathology books.

H C Wainwright

Arafzali B, Goldsmith DJ. Beneficial effects of statins on the kidney. J Clin Pathol 2004;57:673–4. The first author's name was misprinted as Af azali B when it should have been Arafzali B.

BOOK REVIEW

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Further details: Professor B Corrin, Brompton Hospital, London SW3 6NP, UK. (Fax +44 (0)20 7 351 8293; e-mail b.corrin@ic.ac.uk)

Breast Diagnostic Histopathology
Update
22–23 September 2005, Hammersmith Hospital and Imperial College, London, UK
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