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, arthralgia, and loss of 6–8 kg of body weight. She was hypertensive. Physical examination revealed the absence of a bilateral radial pulse and arterial pressure in the upper extremities. Subclavian and axillary artery bruits were audible. Redness of the forearms and hands with trophic changes to the nails were seen. Retinopathy and temporal arterial tenderness or nodularity were not seen. Laboratory data showed an erythrocyte sedimentation rate of 101 mm/hour, and a C reactive protein of 61.9 mg/litre (normal range, 0–3). Doppler ultrasonography of the renal arteries and an echocardiogram were normal. Magnetic resonance angiography of the aortic arch and its branches (fig 1) showed total proximal stenosis of both the subclavian arteries and a significant filiform stenosis of the right vertebral artery, which had an inverted flow. Bilateral temporal arterial biopsy results were negative. Treatment with 0.5 mg/kg/day of prednisone was started, and significant clinical and laboratory improvement was seen. The corticosteroid dose was progressively reduced, and the patient was well one year later.

The differential diagnosis of Takayasu arteritis includes other causes of aortitis and large vessel arteritis but, because of the age of our patient, giant cell arteritis is the most likely. These disorders are not mutually exclusive, but are overlapping, because they are histologically similar and share the same pathogenicity. In addition, giant cell arteritis involves the aorta and its large branches in up to 15% of cases, and approximately 50% of these 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.

E Antón

Department of Internal Medicine, Hospital of Zumárraga, 20700-Zumárraga, Guipúzcoa, Spain; ejanaranda@hotmail.com

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...
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 epiphysal 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. One recent series suggests that the term malignant is not warranted.

In summary, the average age of occurrence of this tumour in previous published series is calculated to be 14 years and 10 months. The youngest previously recorded cases were in 9 year old patients. Therefore, our patient, who is more than 7 years younger than these individuals, is the youngest patient to be recorded with this diagnosis.

The term fibrocartilagenous mesenchymoma with low grade malignancy was first described by Dahlin et al for a rare bone 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 epiphysal 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. One recent series suggests that the term malignant is not warranted.

In summary, the average age of occurrence of this tumour in previous published series is calculated to be 14 years and 10 months. The youngest previously recorded cases were in 9 year old patients. Therefore, our patient, who is more than 7 years younger than these individuals, is the youngest patient to be recorded with this diagnosis.

The term fibrocartilagenous mesenchymoma with low grade malignancy was first described by Dahlin et al for a rare bone
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></td>
<td>0.245</td>
<td>0.036</td>
<td>0.004</td>
<td>0.889</td>
</tr>
<tr>
<td>(p&lt;1&lt;.02)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Paired t test</td>
<td></td>
<td>0.755</td>
<td>0.402</td>
<td>0.810</td>
<td>0.093</td>
</tr>
<tr>
<td>(p&lt;1&lt;.03)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

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

Acknowledgements

This research was supported by FAPESP Fundação de Amparo à Pesquisa do Estado de São Paulo, Proc. 02/03562.

P de J Ribeiro, J L Narciso, C F de Toledo, D R Borges
Gastroenterology Division, Department of Medicine, Universidade Federal de São Paulo, São Paulo, 04023-900 Brazil, durval@gastro.epm.br

References


BOOK REVIEW

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—for example, 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.

The juxtaposition of macroscopic photos with the ultrasound images is helpful for interpretation and correlation of the findings. A strange concept provided under congenital tumours (page 548) is that Castleman disease and Takayasu disease are malformation syndromes with haemangiomias.

In addition, several typographical errors were noted.

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

CORRECTION

Aafzali 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 Aafzali B.

CALENDAR OF EVENTS

Practical Pulmonary Pathology
26–29 July 2005, Royal Brompton Hospital, London, UK
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
Further details: Wolfson Conference Centre, Hammersmith Hospital, Du Cane Road, London W12 ONN, UK. (Tel +44 (0)20 8383 3117/3227/3245; Fax +44 (0)20 8383 2428; e-mail wcc@ic.ac.uk)